SEC Patient Discussions

October 18, 2009

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Case #1

AM

Cutaneous Hodgkin’s lymphoma

CASE # 1
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	AM
Area of interest:	L inner thigh, L upper arm and axilla
History:	47 y/o male presented in 2007 with a slow-growing lesion on his left inner thigh. He reported the lesion to be present for 20 yrs +. The lesion was asymptomatic, but was biopsied by a physician in Ohio a few years prior. Pt. reported the biopsy to be inconclusive so a repeat biopsy was warranted, but he refused. Upon examination a new lesion was noted in his left axilla.
PMH:	Hodgkins lymphoma- Nodular sclerosing, stage IIB with initial inguinal and retroperitoneal lymph node (+) dx in 2004 s/p 6 cycles of ABVD. Tonsillary hypertrophy s/p tonsillectomy (benign). Emphysema, Smoking, Obesity, DVT, ventral hernia s/p repair.
FH:	Father- laryngeal CA. Mother- Liver CA. Sister- Colon CA. Brother- lung CA s/p resection.
PE:	R inner thigh with violaceous/dusky pink soft nodules that coalesce to form large plaques. L inferior shoulder w/ firm, dome shaped discrete erythematous papules, some with kertatotic plugs. Left anterior axilla with collection of follicular based dome shaped almost translucent papules some with dark centers; numerous follicular cysts on back.
Pathology:	A dense dermal infiltrate of lymphocytes, plasma cells, eosinophils, large atypical cells with Reed-Sternberg/ variant cell morphology. These atypical cells do stain with CD30+, CD15+ (weakly), CD45(-). Scattered B cells were noted staining with CD20(+) and CD79a(+). Langerhans cells and dermal dendritic cells were CD1a(+). Histiocytes stained with CD68(+). The infiltrate was composed predominately of T-cells that also stained for CD2(+), CD3(+), CD5(+), CD7(+), CD4 partial (+), CD8 partial (+). PAX 5 positive. Findings c/w Classical Hodgkin lymphoma.
Laboratory:	PET/CT whole body negative since 2007. Slighty elevated WBC at 11.2, Hgb 18.3, Hct 54.3. Normal differential. Normal BMP and LFTs.

Diagnosis: Cutaneous Hodgkin lymphoma

Treatment and Course: Since this lesion was resistant to previous chemotherapy, radiation and repeat chemo was discussed. Radiation was deferred due to the large surface area. Oncology decided to observe the lesions and follow serial clinical exams. Repeat PET/CT scans are done annually along with CBC, CMP. Currently, the patient shows only cutaneous Hodgkin's disease with no systemic involvement.

Comments: Cutaneous Hodgkin's disease is a rare entity estimated with an incidence of 0.5%-3.4% of patients with Hodgkin's disease. Overall, the incidence has been decreasing due to the improvement in diagnosis and treatment. However, when lesions are present, it tends to be in the setting of advanced disease and is a poor prognostic sign (Stage IV). There are three ways to explain the spread of Hodgkin's disease to the skin: hematogenous dissemination, direct extension from an involved lymph node, and retrograde lymphatogenous spread to the area drained by an infected node (most common). There are five reported cases of isolated primary cutaneous Hodgkin's disease without systemic involvement, three had a benign course, two developed systemic Hodgkin's disease. Histopathologically, these lesions can be differentiated from lymphomatoid papulosis (LyP) by the presence of Reed-Sternberg cells that are CD30(+), CD15(+), CD45R (-); LyP lesions are usually CD15 (-), CD45R (+). They are further distinguished from anaplastic large cell lymphoma due to the polymorphous background of inflammatory cells. 3-50% of patients with Hodgkin's disease have nonspecific cutaneous manifestations such as Addison-like hyperpigmentation, pruritis, acquired icthyosis, herpes zoster, and alopecia.

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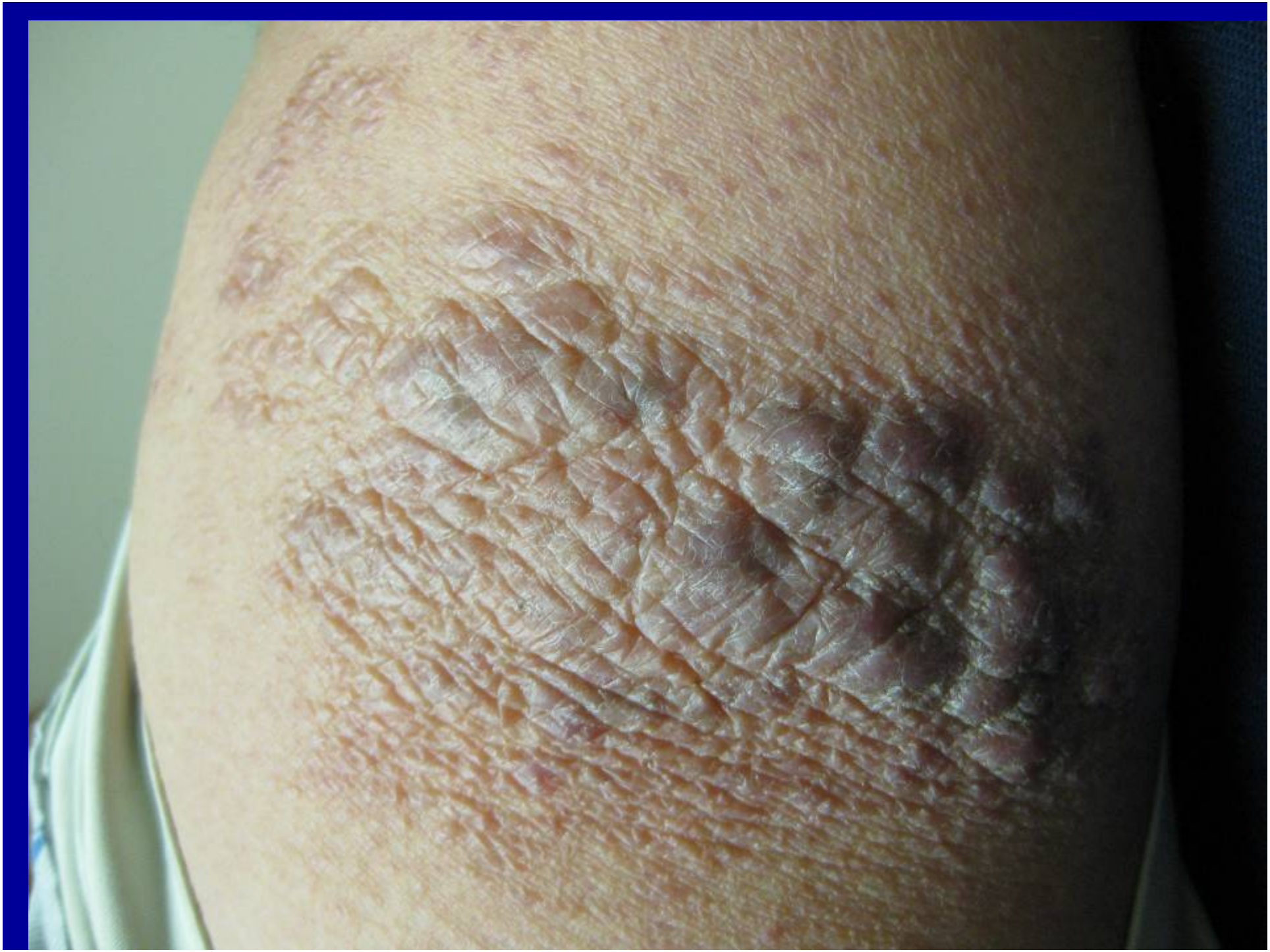

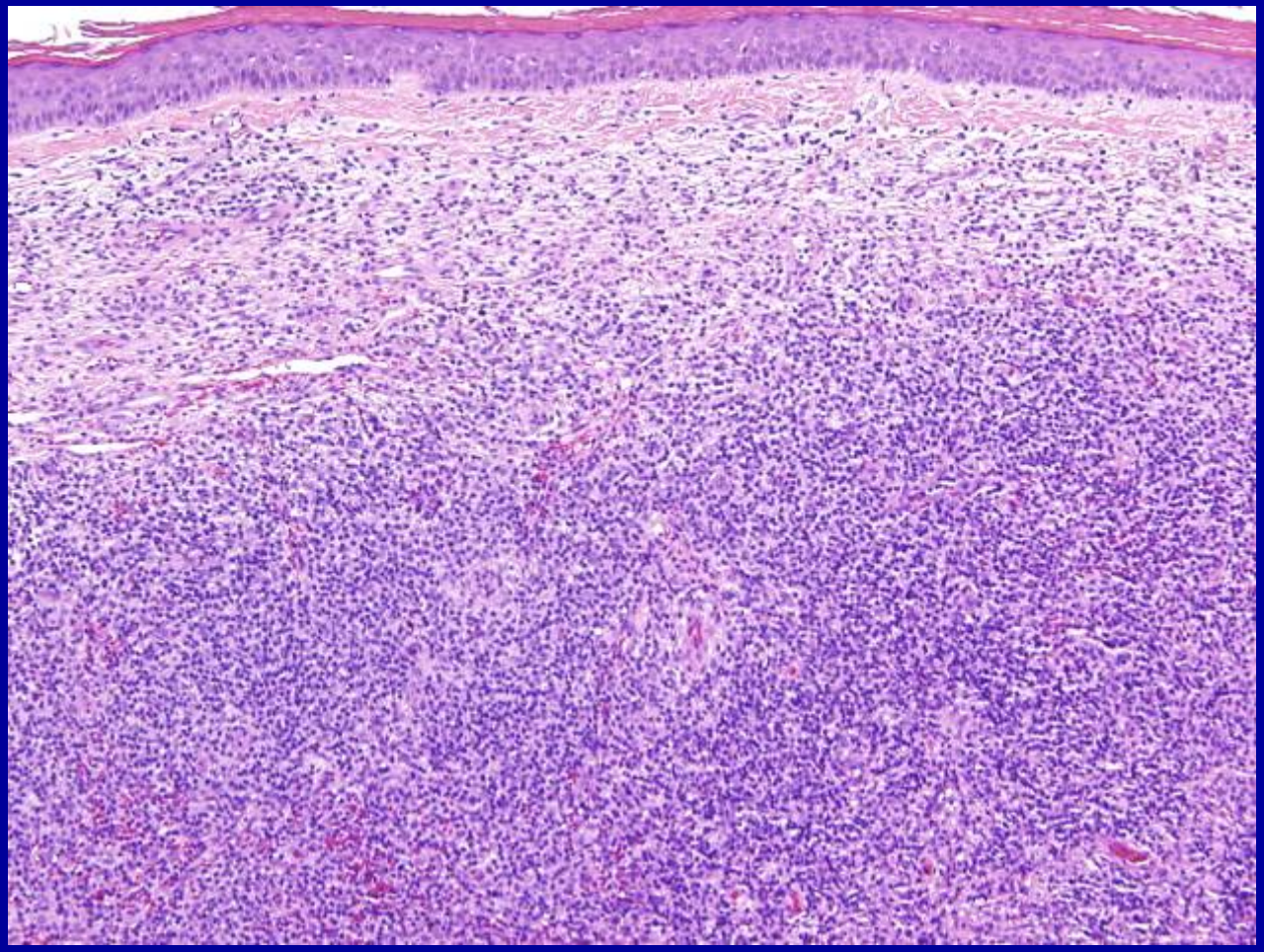

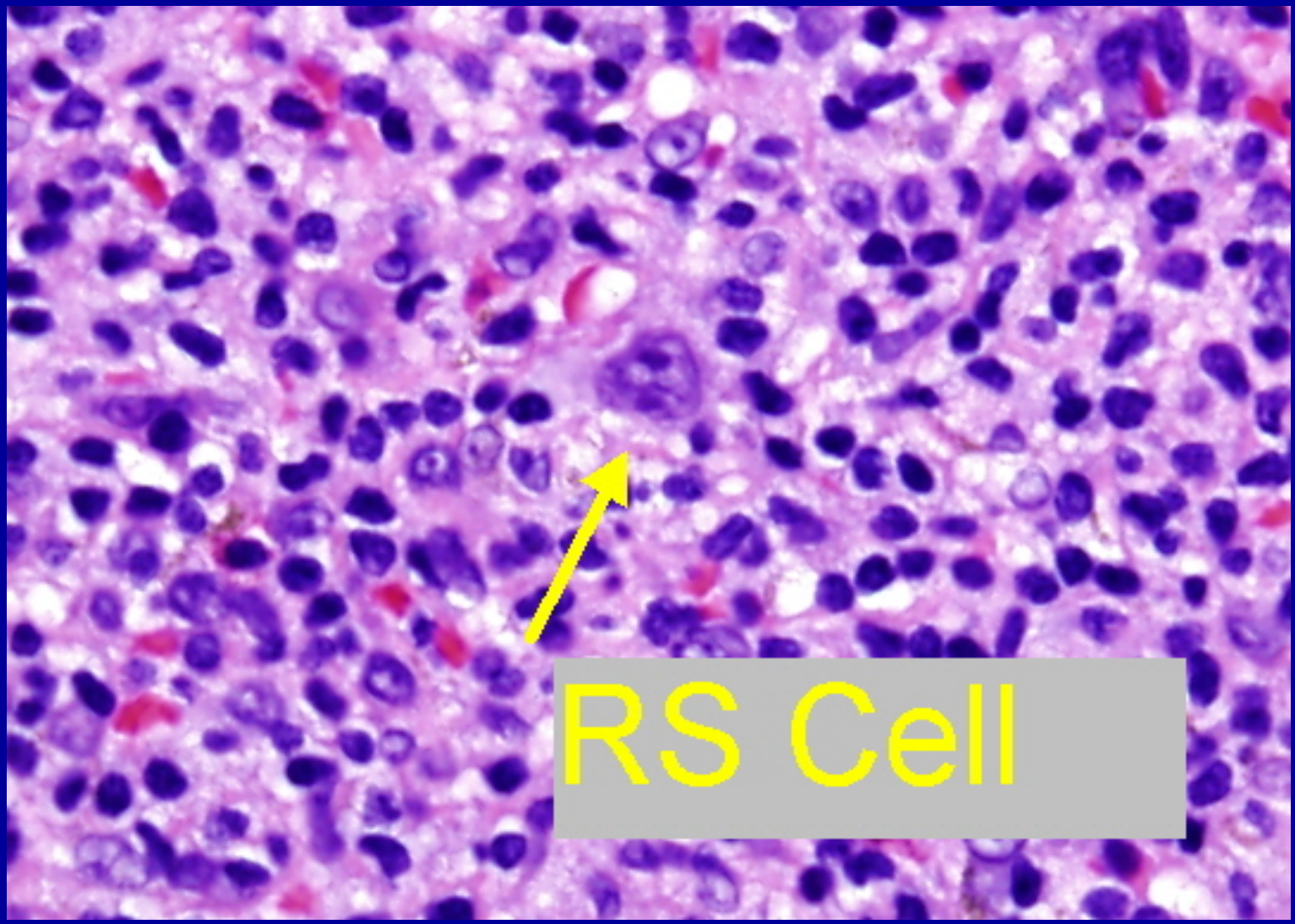
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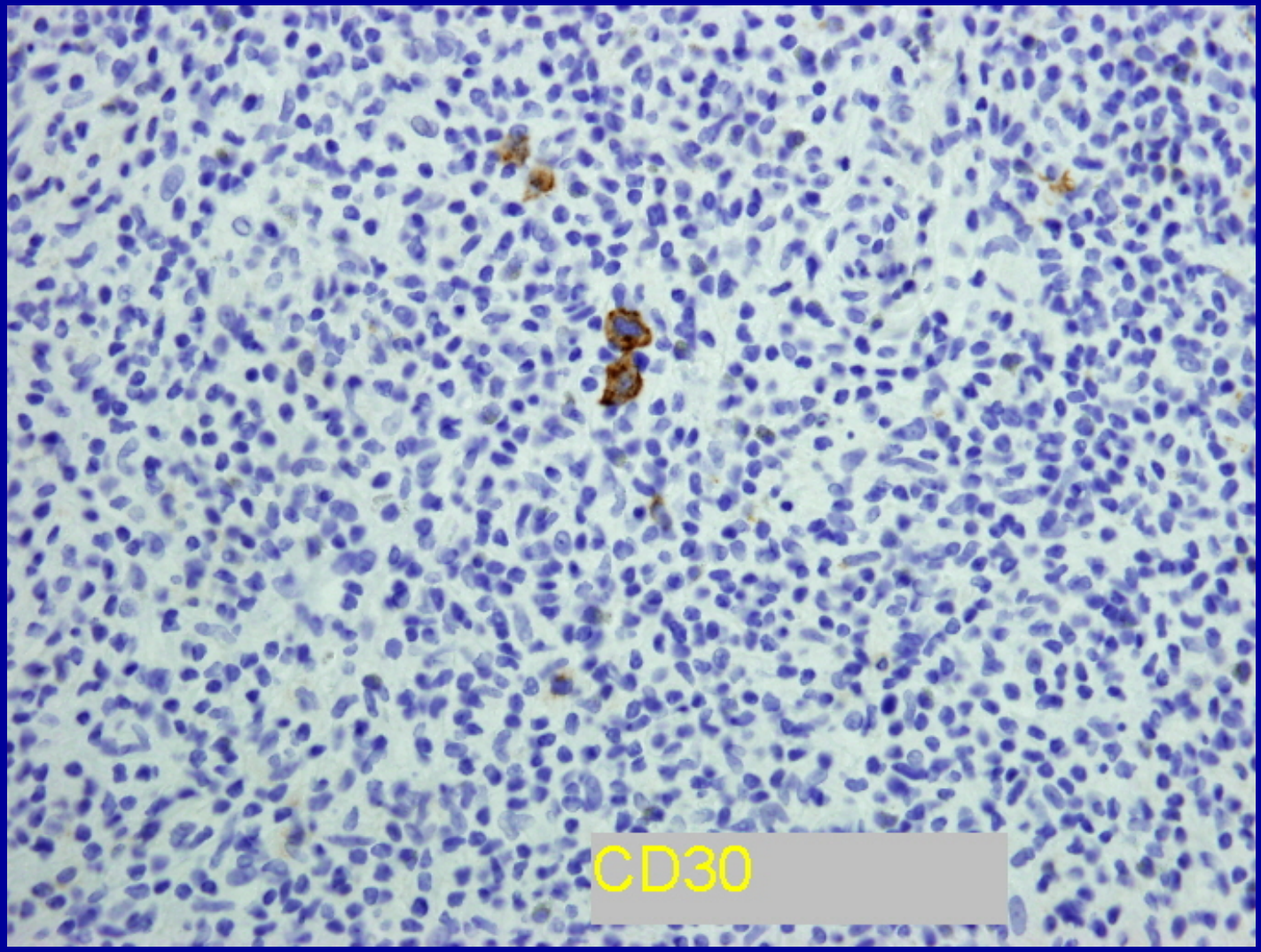
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Case #2

MG

Linear Focal Elastosis

CASE # 2
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	MG
Area of interest:	Back
History:	16 y/o African-American male with a 4 year h/o asymptomatic, linear plaques of the back. He denied any use of topical or systemic corticosteroid use, trauma, or significant change in weight.
PE:	Multiple hyperpigmented, horizontally-oriented linear plaques of the mid to low back. These palpable band-like lesions crossed the midline. Hypopigmented macular striae of the posterior axillary folds and bilateral hips were noted.
Pathology:	Two specimens were submitted for comparison- one was normal skin and the other affected skin. In comparison to the normal biopsy, the abnormal biopsy skin shows a variation in size and shape of reticular dermal collagen bundles. The overlying epidermis is unaltered. Special stained sections (Verhoeff Van Gieson stain) for elastic fibers show fragmented and aggregated elastic fibers in the reticular dermis.
Diagnosis:	Linear Focal Elastosis
Treatment and Course:	Patient declined further follow-up. However, usually these lesions are poorly responsive to treatment.
Comments:	Linear focal elastosis, also known as elastotic striae, is a rarely reported condition characterized by palpable, striae-like, linear plaques or bands. There have been over 20 cases reported in the literature. Lesions typically occur on the mid to lower back. However, they can also occur on the face and lower extremities. Linear focal elastosis is reported in men more often than women. Histologically, there are increased amounts of elastic fibers between hypertrophic collagen in the reticular dermis. These elastic fibers, staining with Verhoeff van Gieson, may appear wavy or clumped with split ends resembling a "paintbrush." The etiology is unknown but postulated to be an excessive repair reaction or an intrinsic abnormality of elastic metabolism/degradation. There have been no associations with systemic, mechanical, or iatrogenic triggers. There is no reported definitive treatment.

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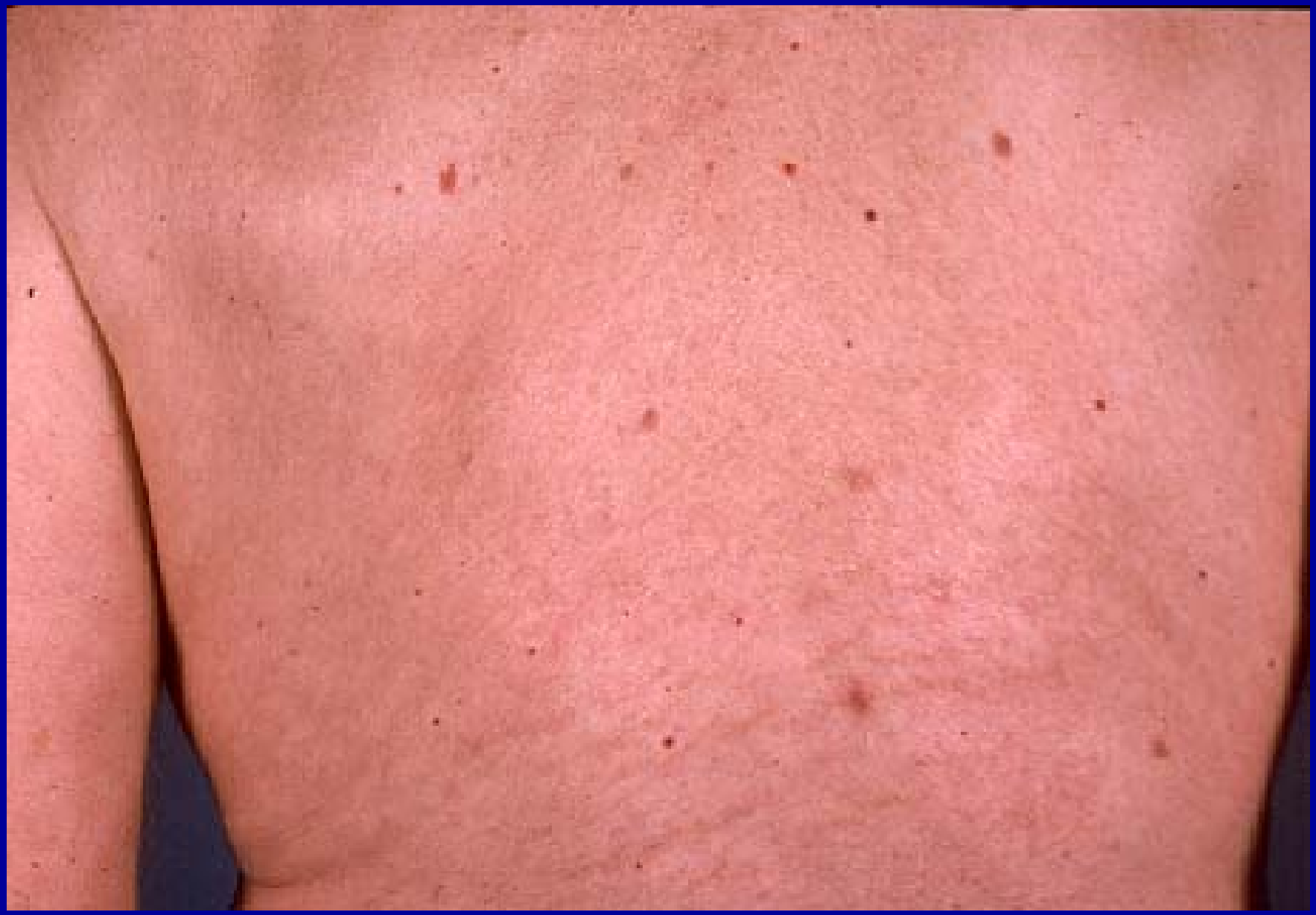

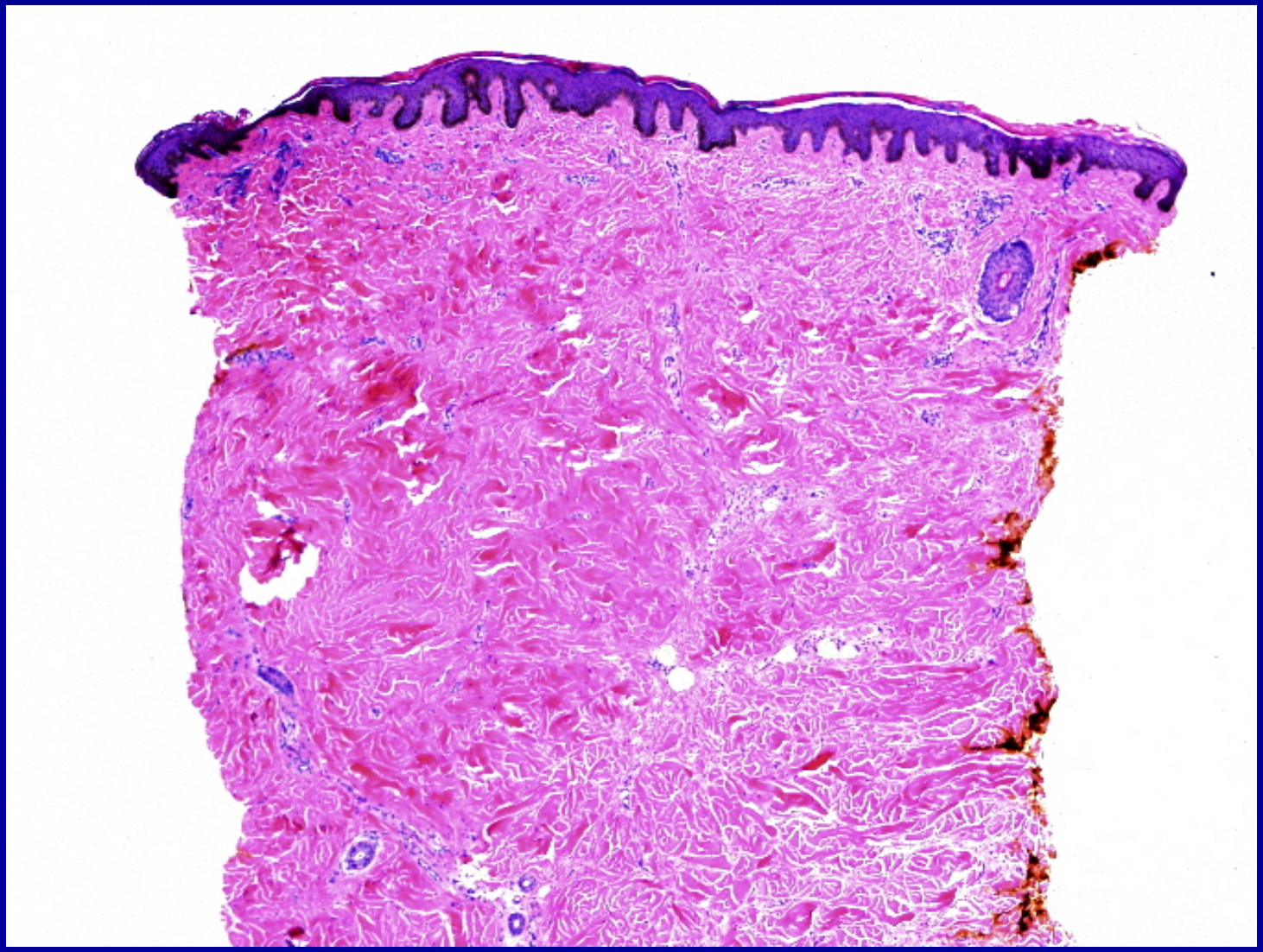

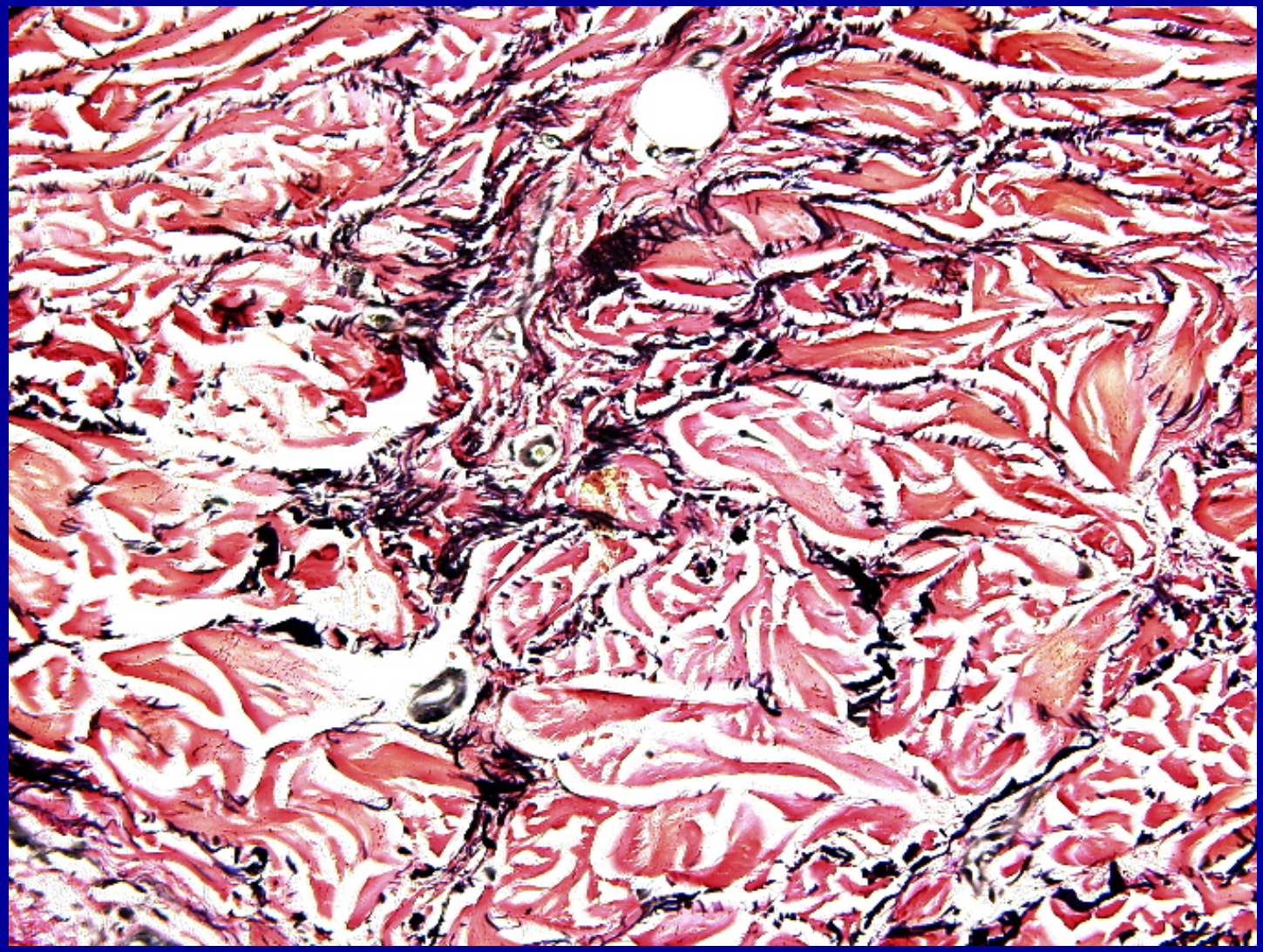

Case # 3

LR

Focal Dermal Hypoplasia
(Goltz Syndrome)

CASE # 3
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	LR
Area of interest:	Left leg
History:	1 day old Hispanic female, born to a 25 y/o G2P2 mother with no health problems, was born with lesions on her L lateral leg. Pregnancy, labor, and delivery were all uneventful. Dermatology was consulted for a vesicular, linear rash that was thought to be a herpes infection by the pediatrician.
Family History:	Normal phenotypic half brother. Normal phenotypic mother and father. No affected relatives. No history of miscarriages.
PE:	Left lateral leg following the lines of Blaschko are soft, erythematous shiny papules that extend from calf to the ankle. Initially, appeared to be vesicular in nature but no fluid was extruded upon unroofing a lesion. No externally visible congenital malformations. Normal digits noted on hands and feet. Baby was feeding and sleeping well.
Pathology:	Punch biopsy reveals a compact parakeratosis with a focal subcorneal pustule composed predominately of neutrophils. The vital epidermis is thinned with attenuation of the normal rete ridge pattern. The dermis is reduced in overall thickness. There is an abnormal distribution of adipose tissue within the dermis. Adipose tissue is focally present at the DEJ.
Laboratory:	PORCN gene mutation/deletion/duplication in exons 1-14 was negative. This is identified as positive in more than two-thirds of cases with Goltz syndrome by DNA sequencing of the PORCN gene by PCR. However, affected males and some females are mosaic. Patients with a low level of mosaicism for a PORCN mutation are difficult to identify and could be missed. It may be necessary to test DNA derived from more than one tissue.
Diagnosis:	Focal Dermal Hypoplasia (Goltz syndrome)
Treatment and Course:	DNA sequencing for large deletions is pending on this patient due to initial negative PORCN gene mutations. Referral to ophthalmologist and orthopedic surgeon upon diagnosis is recommended. X-ray findings of osteopathia striata can be a useful marker of disease. Medical

management is targeted toward the various soft-tissue, dental and skeletal anomalies, with the goal of achieving optimal functional and aesthetic results. Periorificial fibrovascular papillomas may continue to appear during adulthood; which may require repeated surgical intervention. Sketetal deformities may cause severe handicap but overall patients have a normal lifespan.

Comments:

Focal dermal hypoplasia (FDH), also known as Goltz's syndrome, is an uncommon genetic disorder. 200-300 cases worldwide have been reported, 30 of which are males. It is usually, but not always, X-linked dominant (lethal in males). Males born with this disorder frequently are post-zygotic mutations or XXY genotype. Approximately, 95% of females with focal dermal hypoplasia have a new gene mutation; 5% have inherited the mutation from a parent. Baby LR's mild phenotype (absence of limb anomalies, CNS or external ocular anomalies) suggests she may be a mosaic. Loss of function mutation in the X-linked PORCN gene impairs vital cellular signaling in tissues of ectodermal and mesodermal origin. Common dermatological findings include linear or Blaschkoid atrophic or papular lesions on the trunk and extremities which on histology reveal areas of "fat herniation" through a hypoplastic dermis. Another characteristic cutaneous finding is the presence of papillomatous lesions that involve mucocutaneous junctions, alopecia, dystrophic or hypoplastic nails. Oligodontia or small teeth with dysplastic enamel can also be seen. A salient feature of this disease is the presence of syndactyly involving the digits, also referred to as a "lobster claw" deformity. Other musculoskeletal findings include asymmetric limb shortening and hypoplastic digits. An associated, but not pathognomonic, radiologic finding is osteopathia striata, this manifests as hyperdense striations in the metaphyses of long bones. Patients may also present with otic, ophthalmic, neurologic, gastrointestinal or genitourinary involvement. A classic clinical feature described in association with Goltz syndrome patients is the linear crease along the nasal alar rim.

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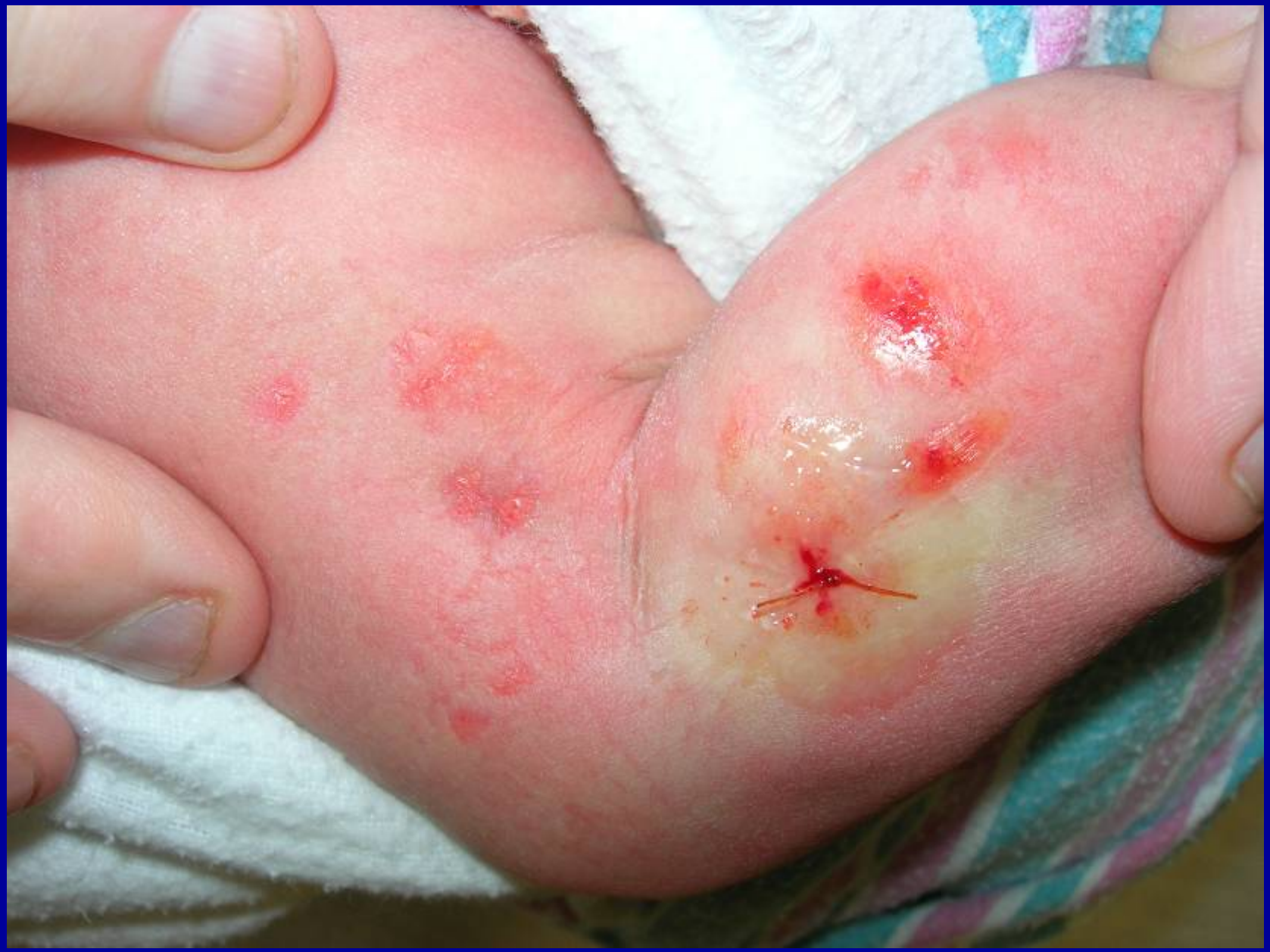

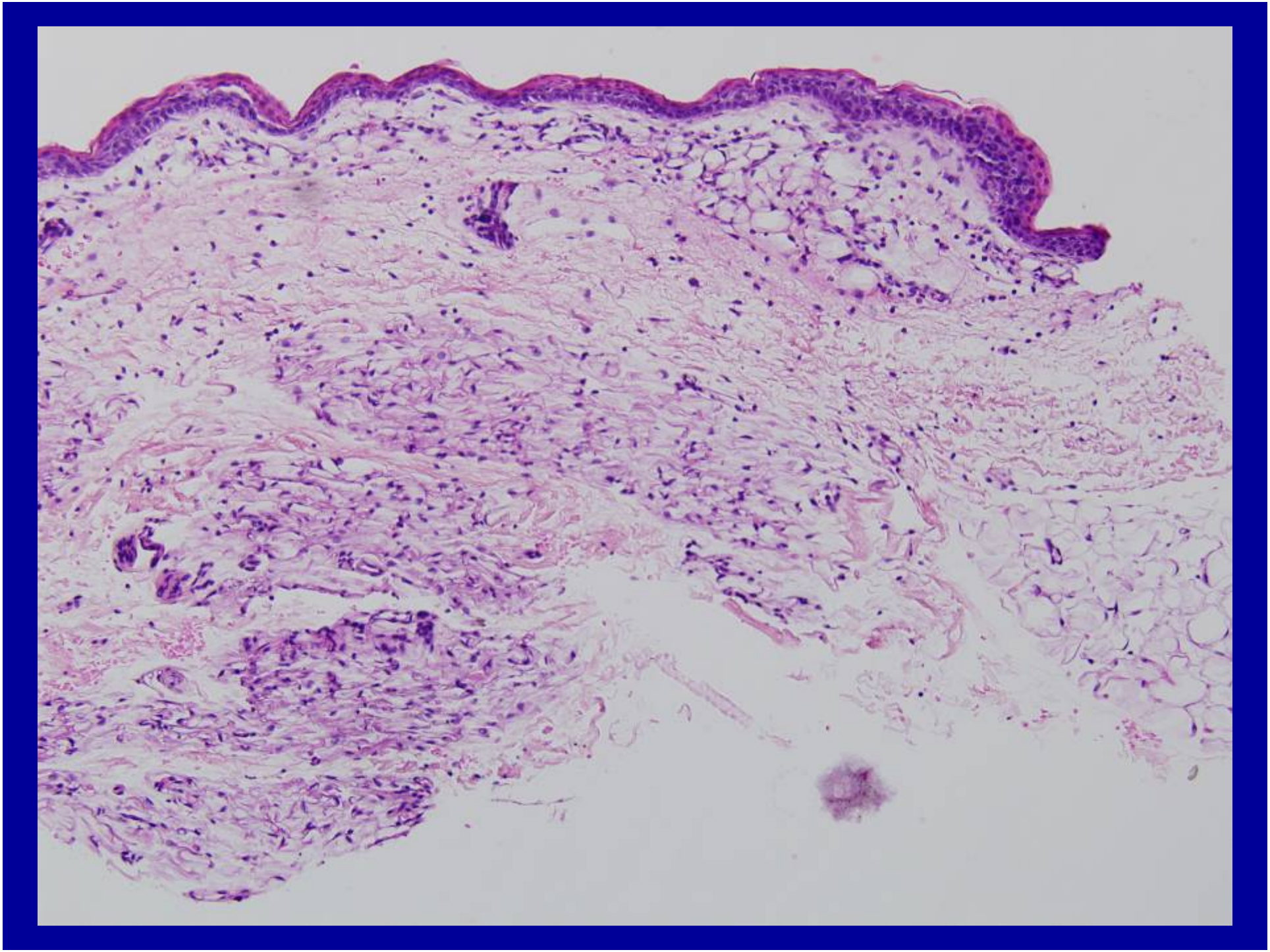

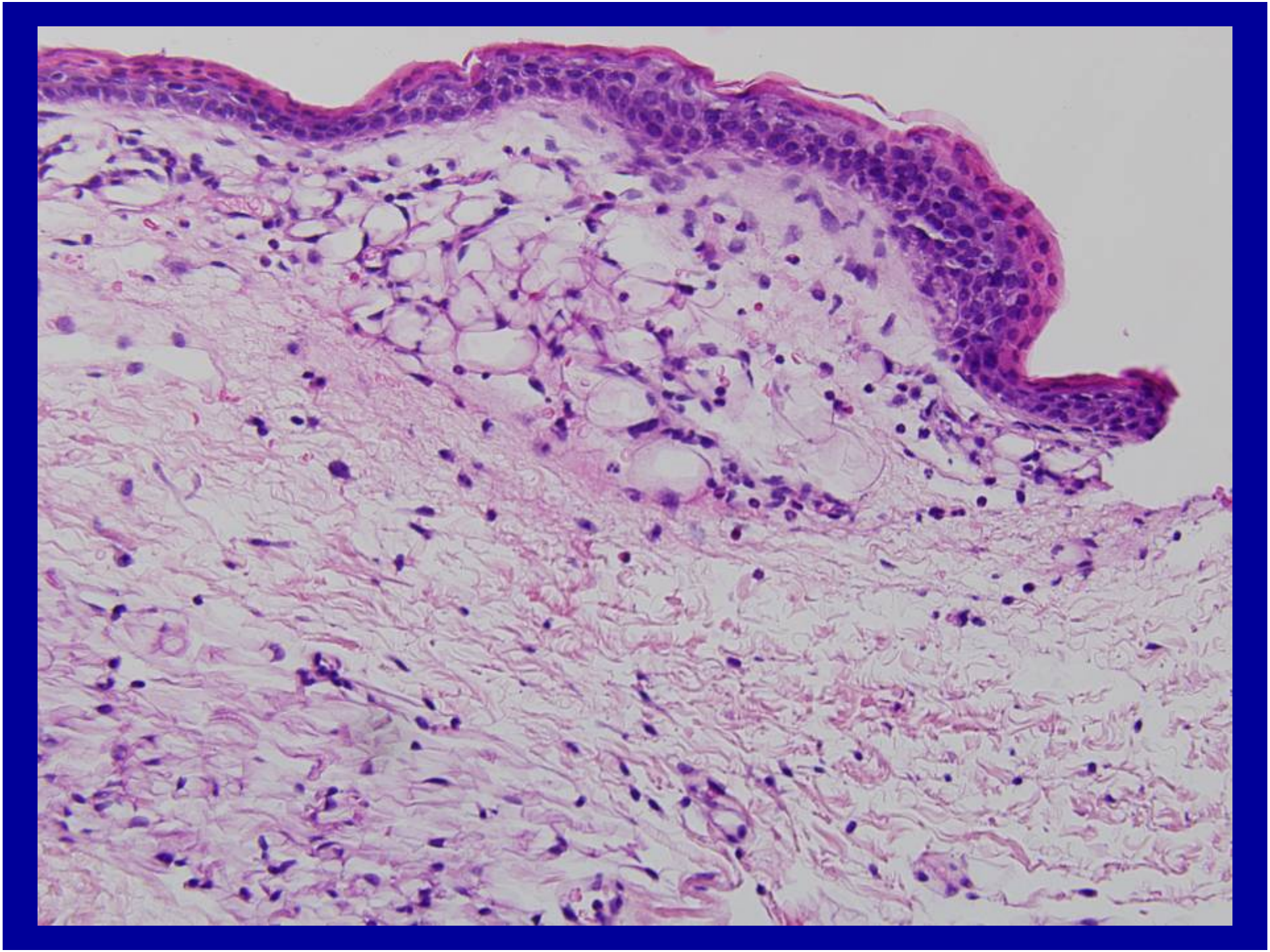

Case # 4

BR

Minocycline Hyperpigmentation
(Type II)

CASE # 4
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	BR
Area of Interest:	Arms, legs
History:	82 y/o woman with a 6 month history of darkening pigmentation on her arms and legs. She reports taking minocycline since January 2009 due to an MRSA infection after hip replacement.
Physical Exam:	Bluish-gray hyperpigmented patches on bilateral forearms and anterior shins.
Pathology:	R shin: The epidermis shows a flat rete ridge pattern with a basket-weave orthokeratotic horn. Within the dermis, there is abundant brown-black pigment in the cytoplasm of histiocytes. The pigment stains positive with both Fontana Masson staining for melanin pigment and Perls stain for iron pigment.
Diagnosis:	Minocycline-induced hyperpigmentation (Type II)
Treatment and course:	Pt. reported not being able to stop minocycline because of her MRSA infection. It is well known that after cessation of minocycline, lightening of the pigmentation can occur over months to years in Types I and II. However, Type III pigmentation tends to be more permanent.
Comment:	Minocycline is a crystalline material that turns black when it is oxidized. Drug-induced hyperpigmentation is a well known side effect of minocycline. Cutaneous and oral pigmentation from minocycline is not necessarily dose dependent. However, risk increases with a cumulative dose greater than 100 grams. Pigmentation can be seen in skin, nails, sclera, oral mucosa, thyroid, bones, and teeth. There are three types of minocycline induced hyperpigmentation. Type I is a blue-black discoloration that occurs in inflammatory lesions and scars, such as acne. Type II is a blue-gray macule or patch that appears in previously normal skin, common on the arms and anterior shins. Type III is a muddy brown discoloration that occurs in sun exposed sites. Histologically, Type I hyperpigmentation reveals both intra- and extracellular iron which stains with Perls (Prussian blue). This is likely secondary to hemosiderin or minocycline deposition with iron chelation. Type II lesions contain both iron and melanin pigment. Therefore, these lesions stain with Perls and Fontana Masson. Type III lesions show increased

melanin in the basal layer and dermal macrophages, therefore, only stains with Fontana Masson. Types I and II may improve with time and tend to be responsive to Q switched Alexandrite laser treatment.

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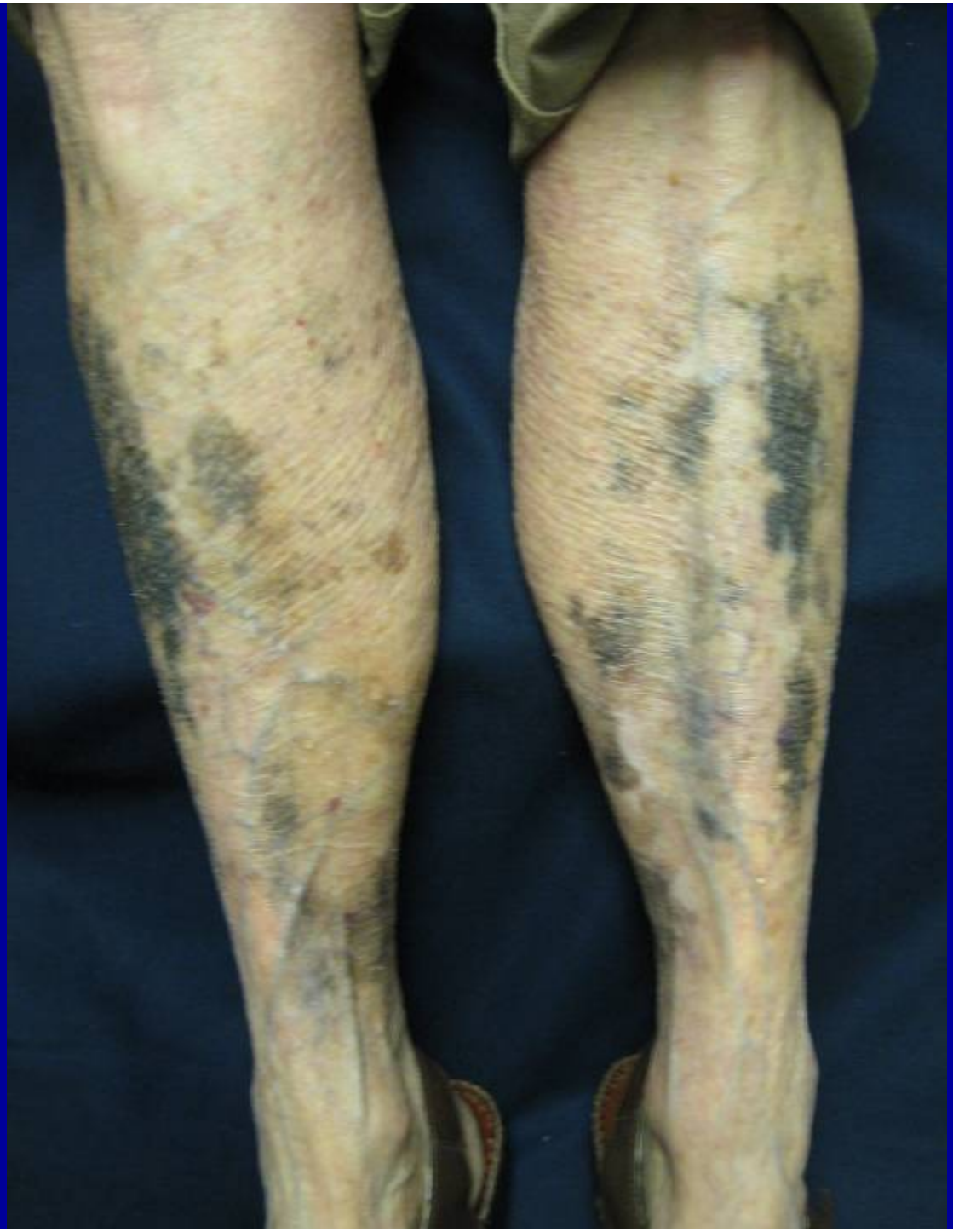

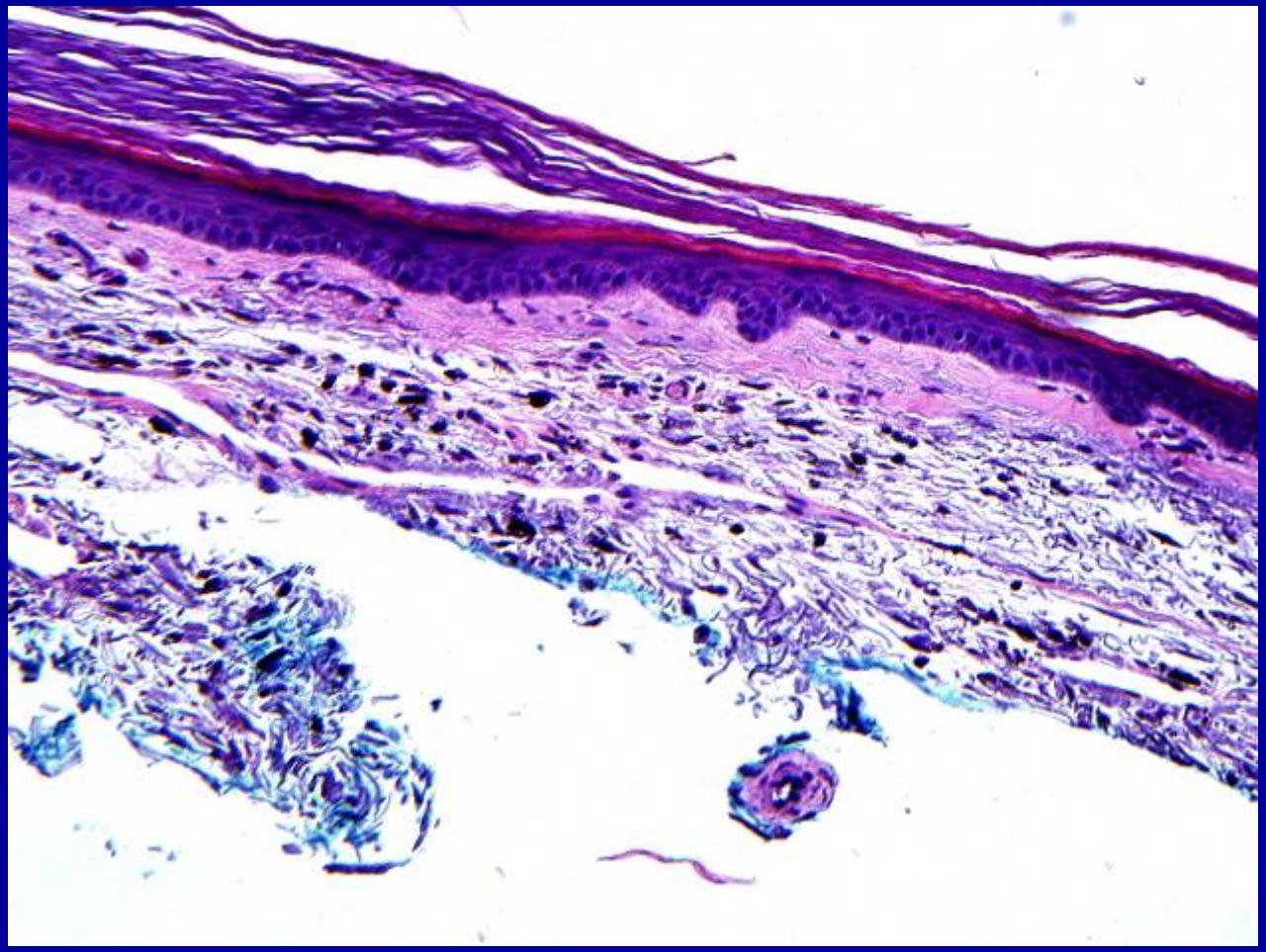

Case #5

HK

Familial presenile sebaceous gland hyperplasia

CASE # 5
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	HK
Area(s) of Interest:	Face
History:	40 year old Chinese woman with progressive worsening of multiple facial papules that began forming in her mid twenties. No known endogenous or exogenous exposures. No significant past medical history.
Family History:	Father with a history of oily skin
Physical Exam:	A myriad of confluent monomorphic yellowish 1-2 mm papules with extensive involvement of the entire face with conspicuous periocular, distal nasal tip, and upper lip sparing. Some the papules have erythema and telangiectasias as a component.
Social History:	Patient is married and currently attempting to conceive
Laboratory:	Testosterone, DHEA-S, Prolactin all normal
Pathology:	Two biopsies done with both biopsies showing within the dermis a pilosebaceous apparatus that opens to the surface through a patulous sebaceous duct that contains sebum and keratin. There is slight acanthosis of the epidermis adjacent to the opening of the follicle. Within the dermis there are enlarged sebaceous lobules with admixed perifollicular and perivascular mixed inflammatory infiltrate that contains eosinophils, lymphocytes, and histiocytes consistent with sebaceous hyperplasia with inflammation.
Diagnosis:	Familial Presenile Sebaceous Gland Hyperplasia
Treatment and Course:	Patient has undergone several topical acne treatments including retinoids and benzoyl peroxide with no benefit. Photodynamic therapy also did little to improve her condition. Oral isotretinoin therapy did improve her condition but the teratogenic effects of this medication preclude the use at this time.
Comment:	Familial presenile sebaceous gland hyperplasia, also known as premature sebaceous hyperplasia, presents with extensive sebaceous hyperplasia with onset usually at puberty and worsening with age. It has been described in the literature to follow these characteristics: (1) confluence of lesions sparing periorbital and perioral regions, (2) highly functional glandular hyperplasia resulting in

excessive sebaceous secretion, (3) absence of acne, (4) lesions on the face but also on the neck and upper chest, (5) unresponsiveness to conventional acne treatment, (6) histopathologic features similar to senile sebaceous hyperplasia, and (7) appearance during puberty or just afterwards with a slow progressive nature. Several of the described cases show a familial inheritance with a pedigree suggesting an autosomal dominant inheritance. No internal malignancies or hormonal abnormalities have been reported. Response to oral isotretinoin has been reported with relapse in the majority of cases.

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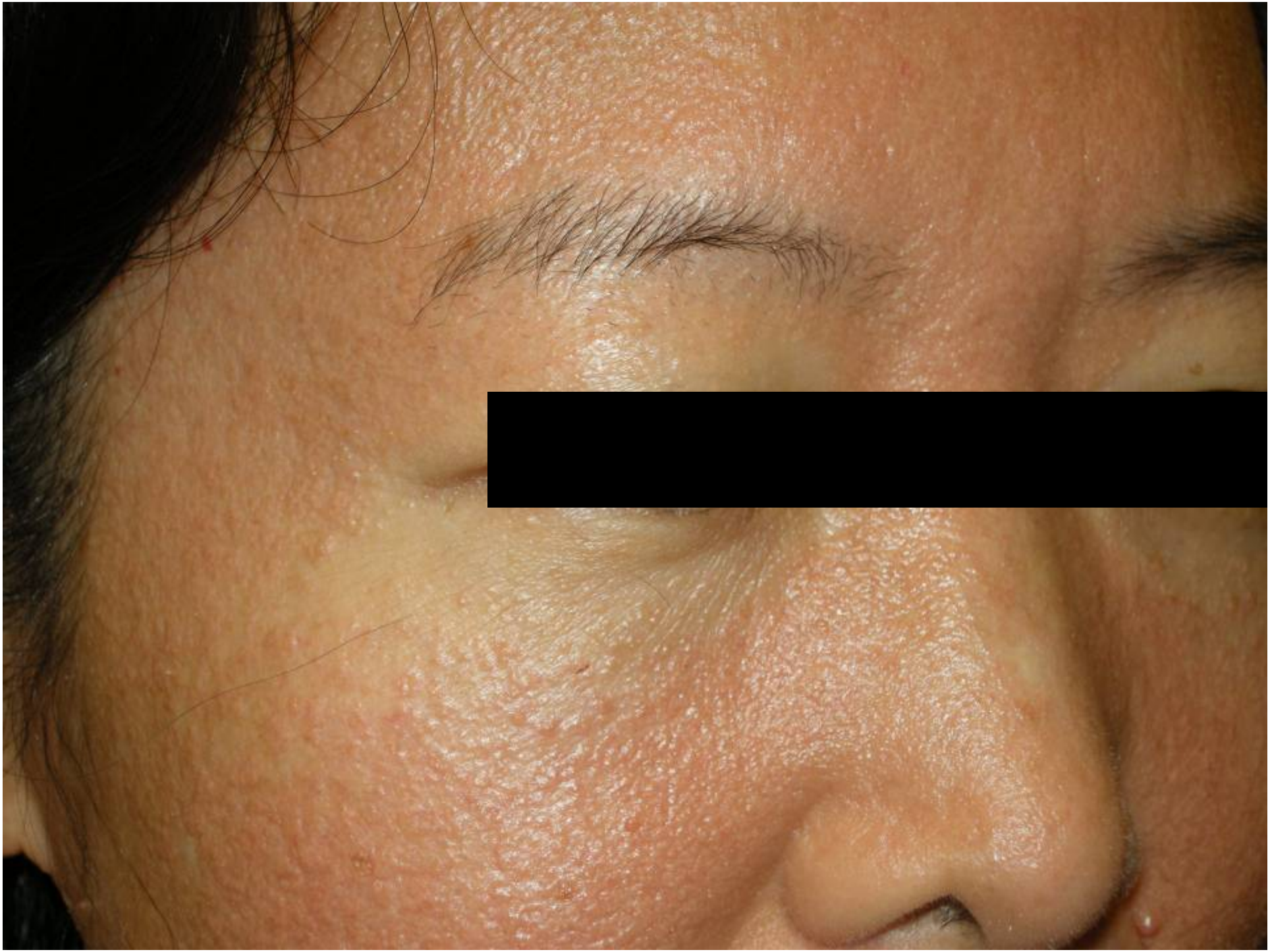

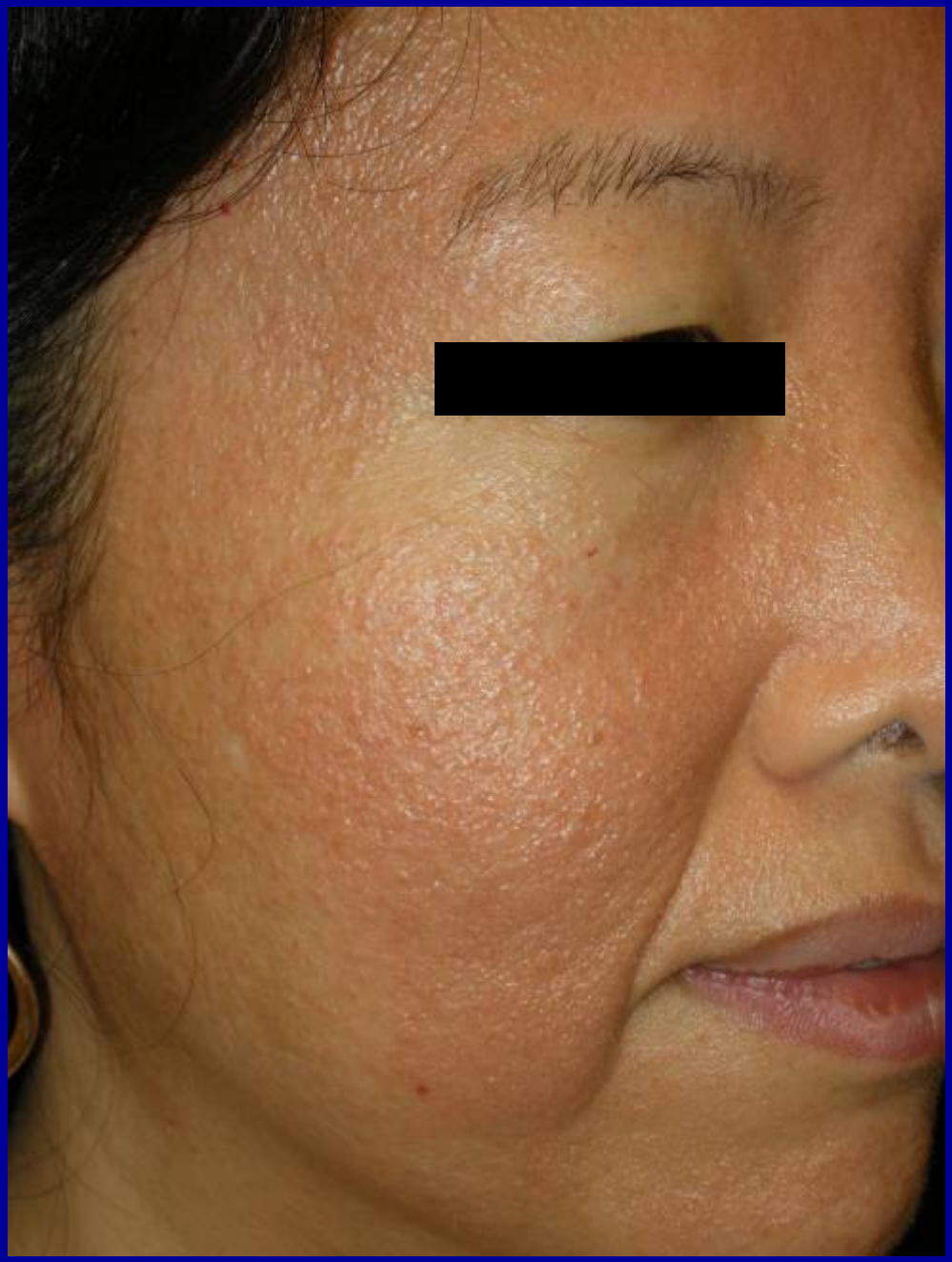

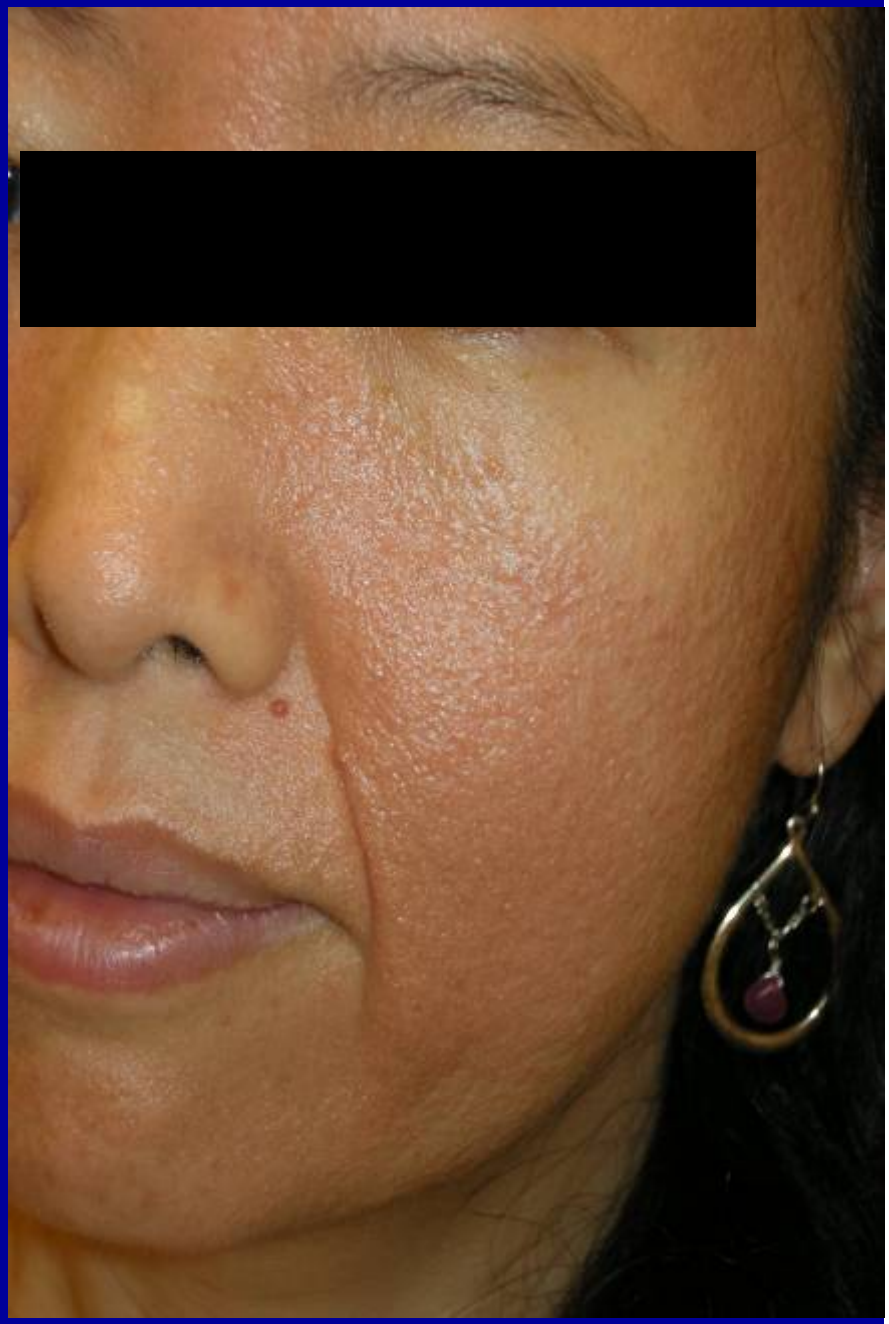

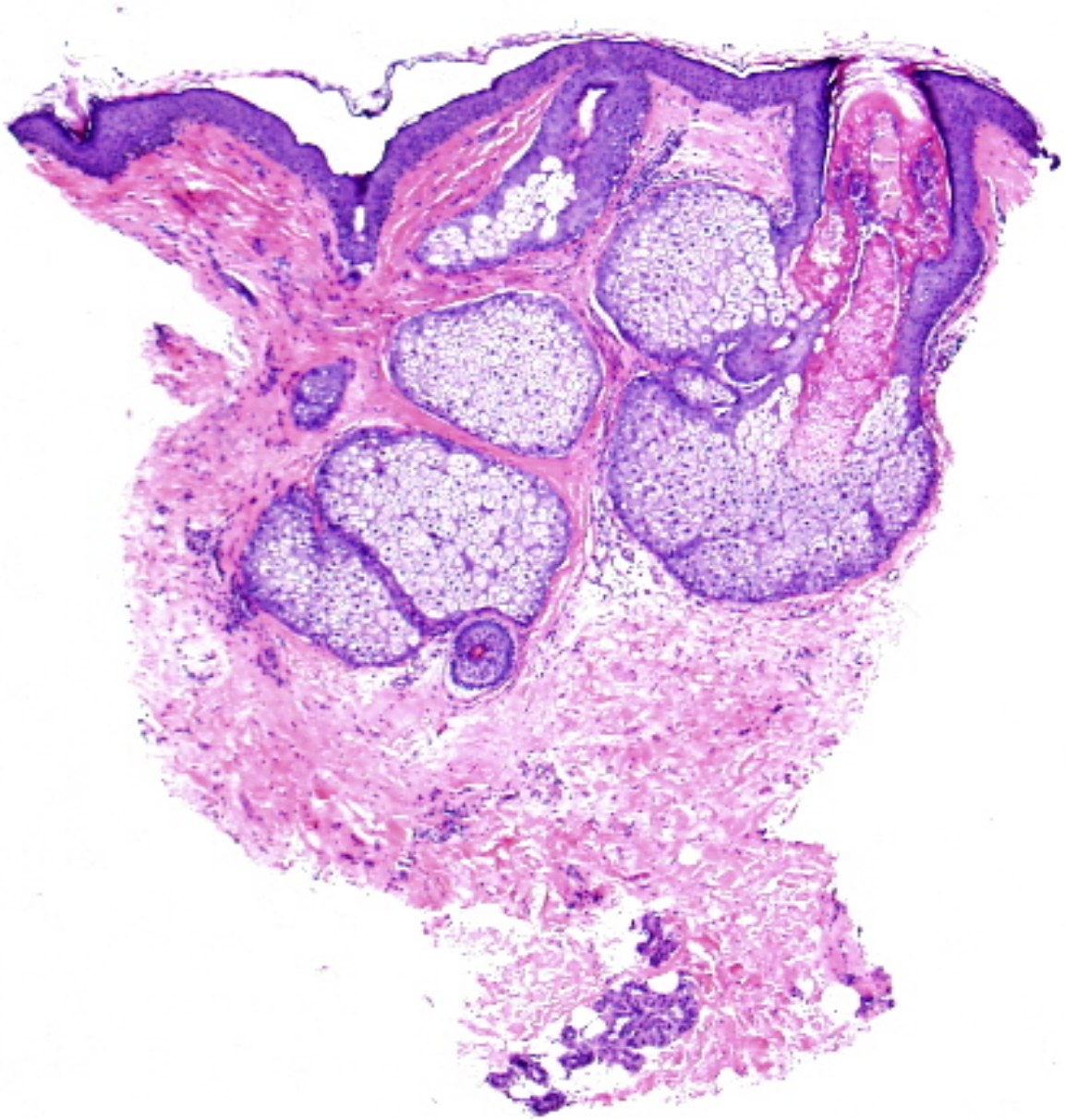

Case # 8

AS

Infantile Pustular Psoriasis

CASE # 8
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: AS

Area(s) of Interest: Scalp, face, trunk, upper and lower extremities, and nails

History: 5 year old Caucasian girl born after a full-term vaginal delivery with no complications. No skin lesions or disorders were noted at birth. At six months of age, she developed a diffuse cutaneous eruption over her entire body. Since then, she has had numerous cutaneous flares, which have subsided over the past 1-2 years with only persistent nail involvement.

Family History: No skin diseases in the family

Physical Examination: Pictures from infancy show extensive annular red plaques studded by small pustules covering her entire body. Currently, involvement is limited to her nails with thickening and dystrophy of all fingernails and toenails, sparing the 3rd right toenail.

Histopathology: The stratum corneum is hyperkeratotic with parakeratosis and serum crust. Mild irregular acanthosis is seen in the epidermis with neutrophils accumulating into subcorneal pustules. There are dilated papillary dermal blood vessels and a superficial perivascular infiltrate consisting of lymphocytes and neutrophils.

Laboratory Studies: Normal serum calcium

Diagnosis: Infantile Pustular Psoriasis

Treatment and Course: The patient presented to our clinic in 2009 at age 5 with nail disease only. Multiple photos from 6 months of age on showed full body involvement ofannular pustular psoriasis, which was unrelated to any systemic corticosteroid use. No family history of psoriasis was known. Associated findings during her flares were geographic tongue and anormal serum calcium. Though her psoriasis tended to flare when she was ill, she never tested positive for *Streptococcus*. Interestingly, she has not had a generalized flare since undergoing elective tonsillectomy at age 3. Prior to that, she continued to have recurrent flares despite systemic therapy including including methotrexate, cyclosporine, acitretin, and various topical steroids and Vitamin D analogues. She never received phototherapy or treatment with a TNF-α inhibitor. Since her tonsillectomy, she has been in clinical remission with only mild, localized

flares, which are quickly controlled with topical steroids. Her nail disease, however, has persisted.

Discussion:

Childhood psoriasis can be separated into congenital, infantile, and childhood types, though no strict diagnostic guidelines exist. Some authors use the following to classify them: congenital – presenting at birth; infantile – presenting within the first year of life; childhood – presenting after the first year of life. Regarding congenital and infantile psoriasis, there is some overlap with both seborrheic dermatitis and diaper dermatitis. Most cases of infantile psoriasis involve the diaper area, but when excluded, plaque type and scalp psoriasis predominate, with generalized pustular eruptions being very rare. As opposed to adult disease, facial involvement is common. Also, childhood pustular psoriasis is typically unassociated with systemic steroid use, a prior history of plaque type psoriasis, or serum calcium abnormalities. Family history is seen in 89% of infantile psoriasis, but interestingly, low incidences are seen amongst cases of congenital psoriasis. No correlation between early age of onset and severity of disease has been found. Topical emollients are effective in mild cases, though more extensive cases may require topical steroids, phototherapy, or immune modulating medications. TNF-alpha inhibitors are effective, though the long-term safety of these medications in children is unknown. The correlation between streptococcal infection and childhood guttate psoriasis has prompted the use of antibiotic therapy and tonsillectomy as treatment, leading to clearance rates of 30-50%. Topical tazarotene 0.05% may be a safe and effective treatment for psoriasis of the nails.

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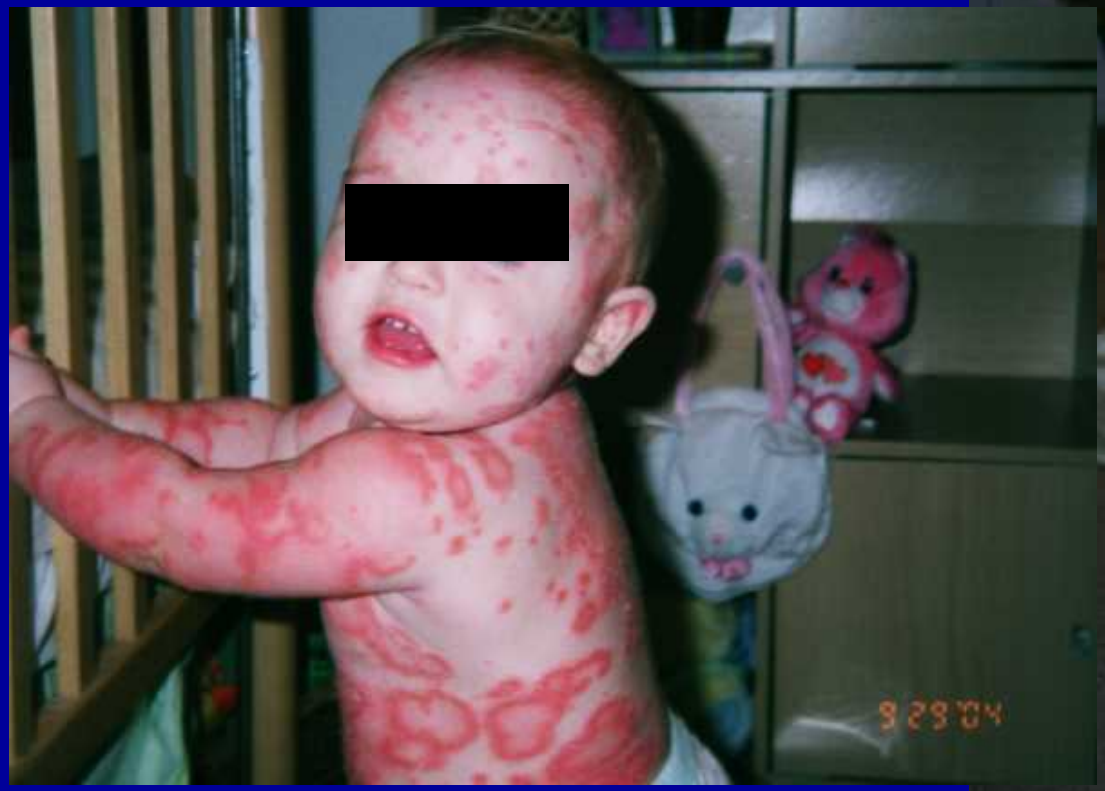
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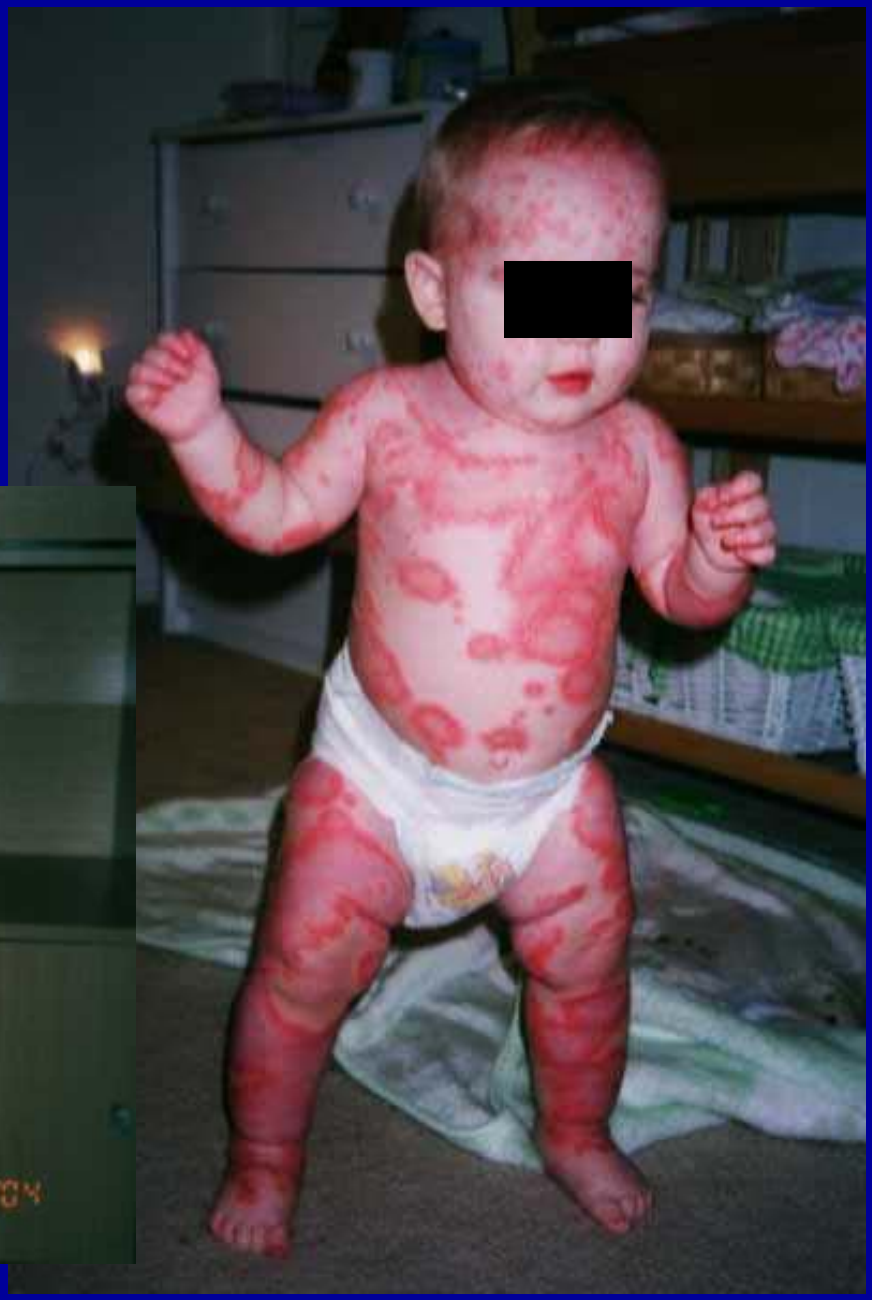

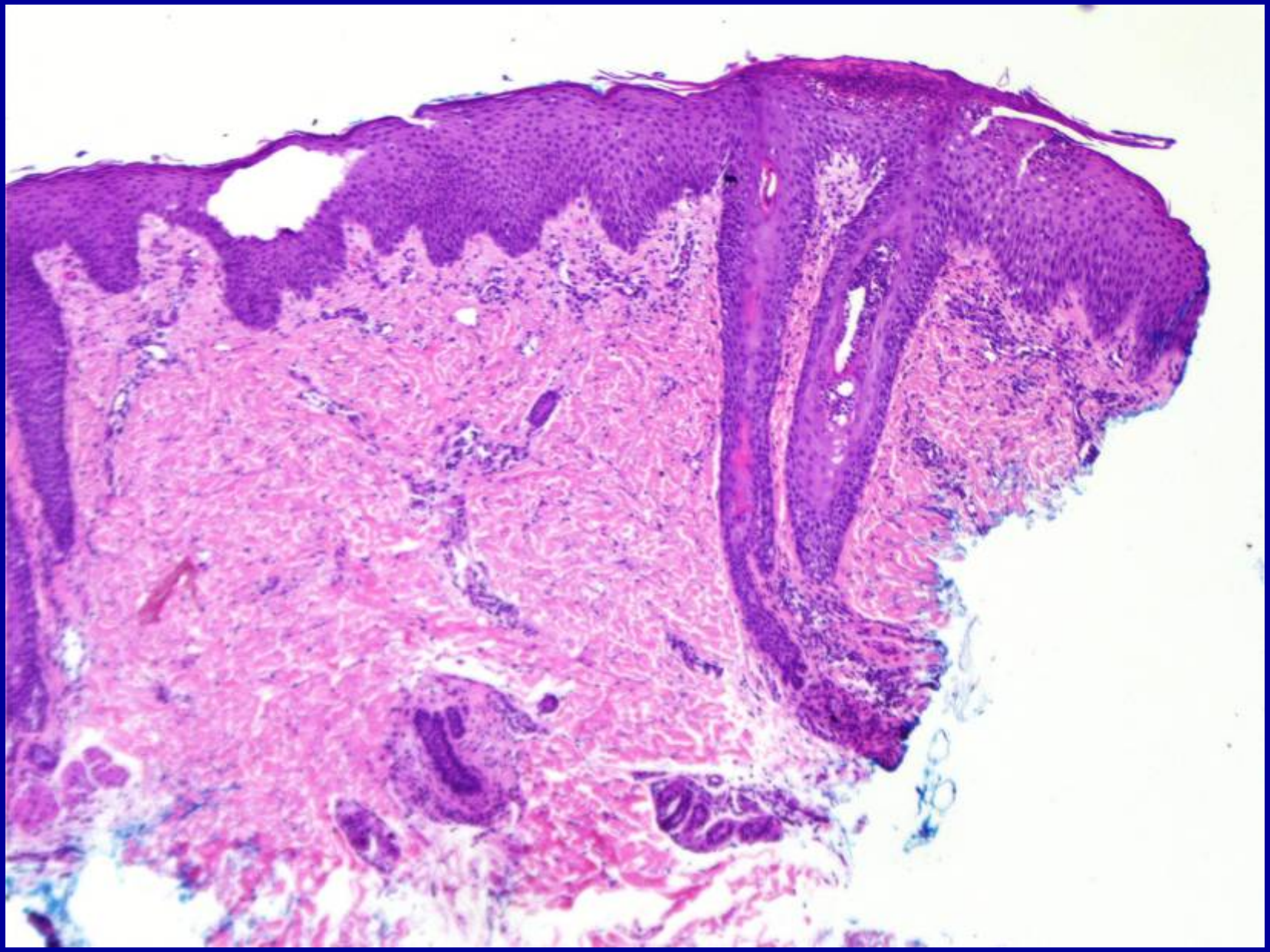

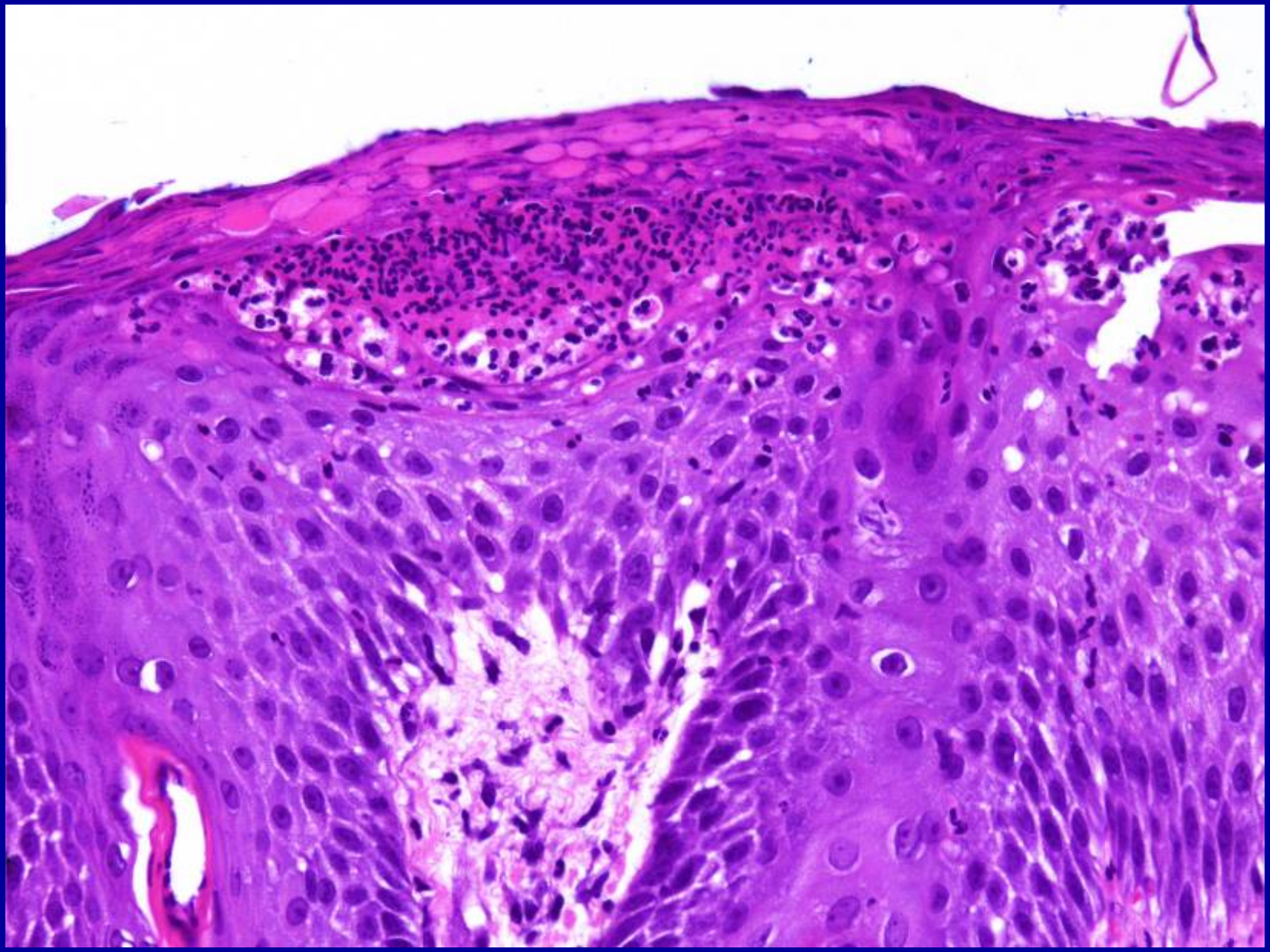

Case # 9

MK

Incontinentia Pigmenti (IP)

CASE # 9
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	MK
Area(s) of Interest:	Upper and lower extremities, trunk, scalp, and digits.
History:	6 day old Caucasian female born after a term, uncomplicated pregnancy. At birth, the skin was noted to have mild erythema and digital deformities on the left hand were noted. At 3 days of age, vesicles appeared in linear patterns.
Physical Examination:	Linearly distributed bands of erythema with numerous vesicles on the upper and lower extremities, trunk, scalp, and genitalia. Severe foreshortening of the second, third, fourth, and fifth digits was noted on the left hand. The left thumb was fully developed with nail intact.
Histolopathology:	Focal intercellular edema with exocystosis of eosinophils and dyskeratotic keratinocytes within the epidermis. Sparse interstitial dermal infiltrate consisting of eosinophils and occasional neutrophils.
Laboratory Studies:	Southern blot analysis of DNA revealed a deletion mutation of the NEMO gene at Xq28.
Diagnosis:	Incontinentia Pigmenti (IP)
Treatment and Course:	A complete ocular evaluation at 2 weeks of age revealed mild hyperopia and no evidence of retinal abnormalities. She has been closely observed for seizure activity or other evidence of CNS abnormality and none has been identified. Follow-up visits in the early months of life revealed resolution with crusting of the vesicles and diminution of diffused erythema but occasional recurrent crops of new vesicles most prominent on the extremities.
Discussion:	Incontinentia pigmenti (IP) is a rare X-linked dominant genodermatosis that affects mainly female patients as it is usually fatal for males in utero. The four stages of skin lesions are as follows: vesicular (birth to 1-2 weeks), verrucous (2-6 weeks), hyperpigmentation (3-6 months), and hypopigmentation (second to third decade). The lesions follow the lines of Blaschko. Patchy alopecia at the vertex of the scalp, onychodystrophy, subungal tumors with underlying lytic bone lesions, and palmoplantar hyperhidrosis can also be seen. Extracutaneous manifestations include abnormalities of the teeth (up to

90%), bones (40%), central nervous system (33%), and eyes (35%). Dental defects include anodontia, cone, or peg teeth. CNS findings include seizures, mental retardation, and spastic paralysis. Eye abnormalities include strabismus, cataracts, optic atrophy, and retinal vascular changes. Skeletal deformities include syndactyly, skull deformities, dwarfism, spina bifida, club foot, supernumerary ribs, hemiatrophy, and shortening of the legs and arms. To our knowledge there has been one previous report of unilateral acheiria in a girl with IP (4). That child had complete absence of the hand in contrast to our patient who had incomplete development of four digits.The defect underlying IP is well-defined as a loss of activity of the regulatory component encoded by the NEMO/IKKc gene. The NEMO protein activates NFκB, which is thought to protect from TNF-α driven apoptosis.

References:

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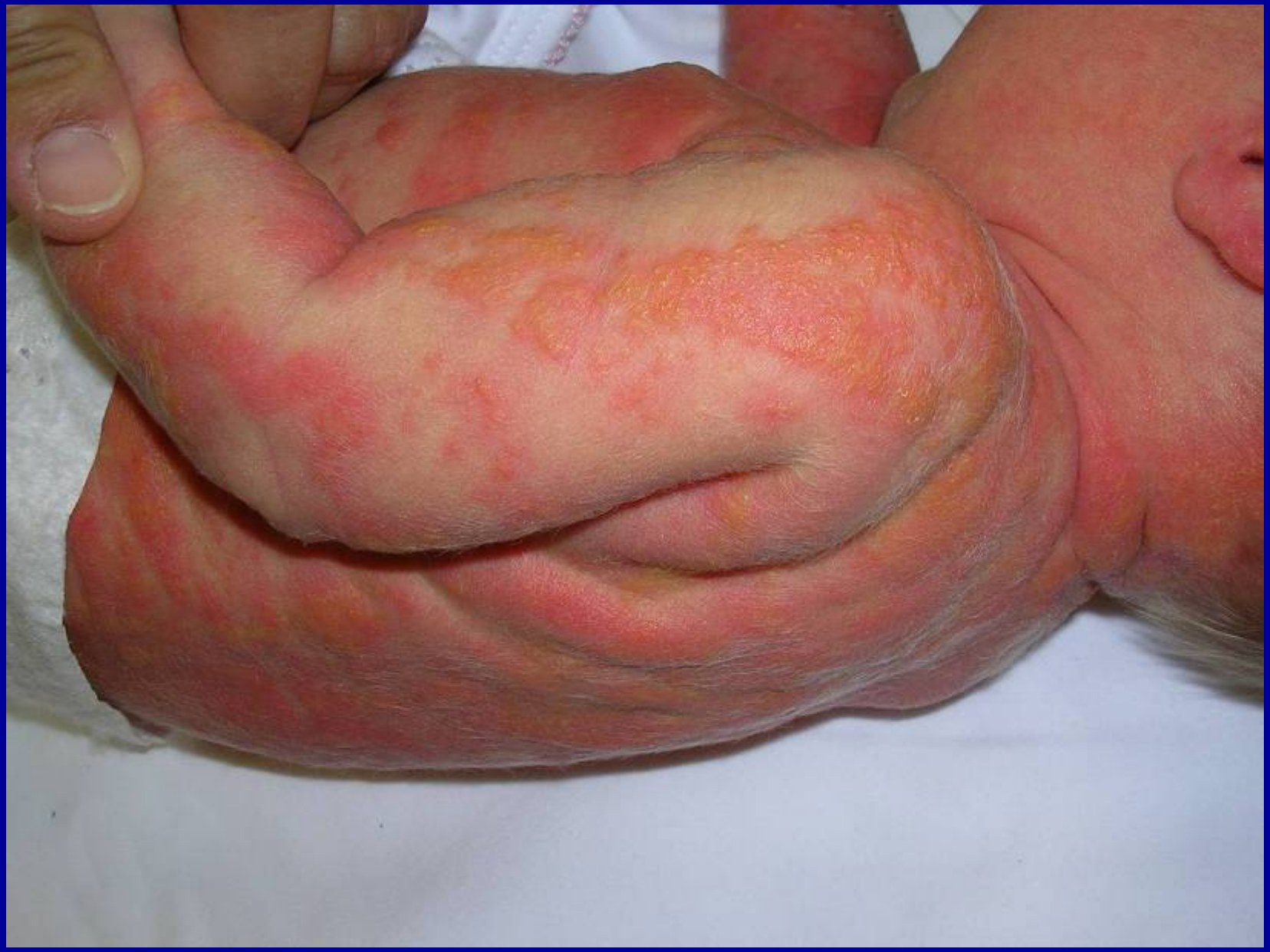

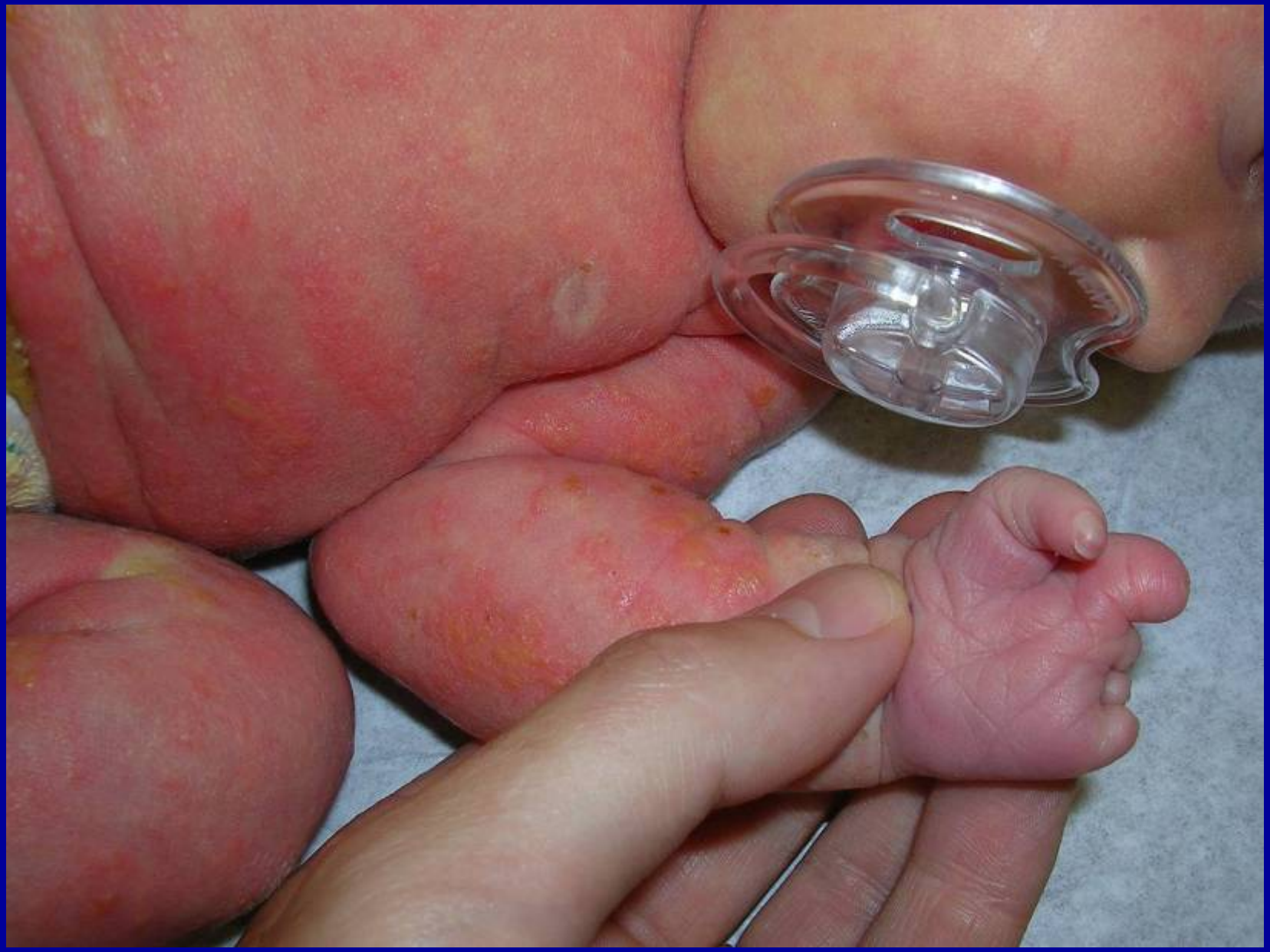

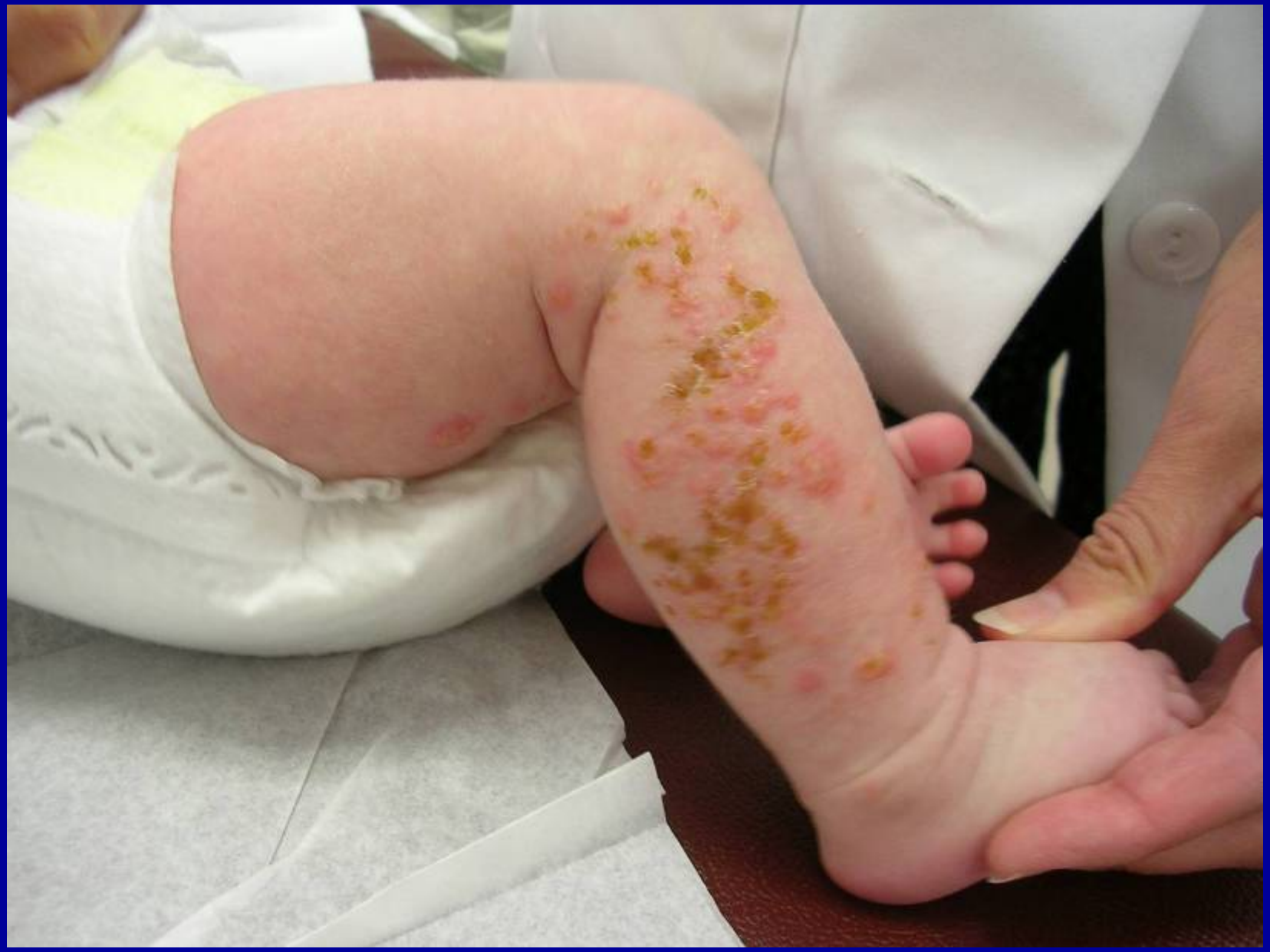

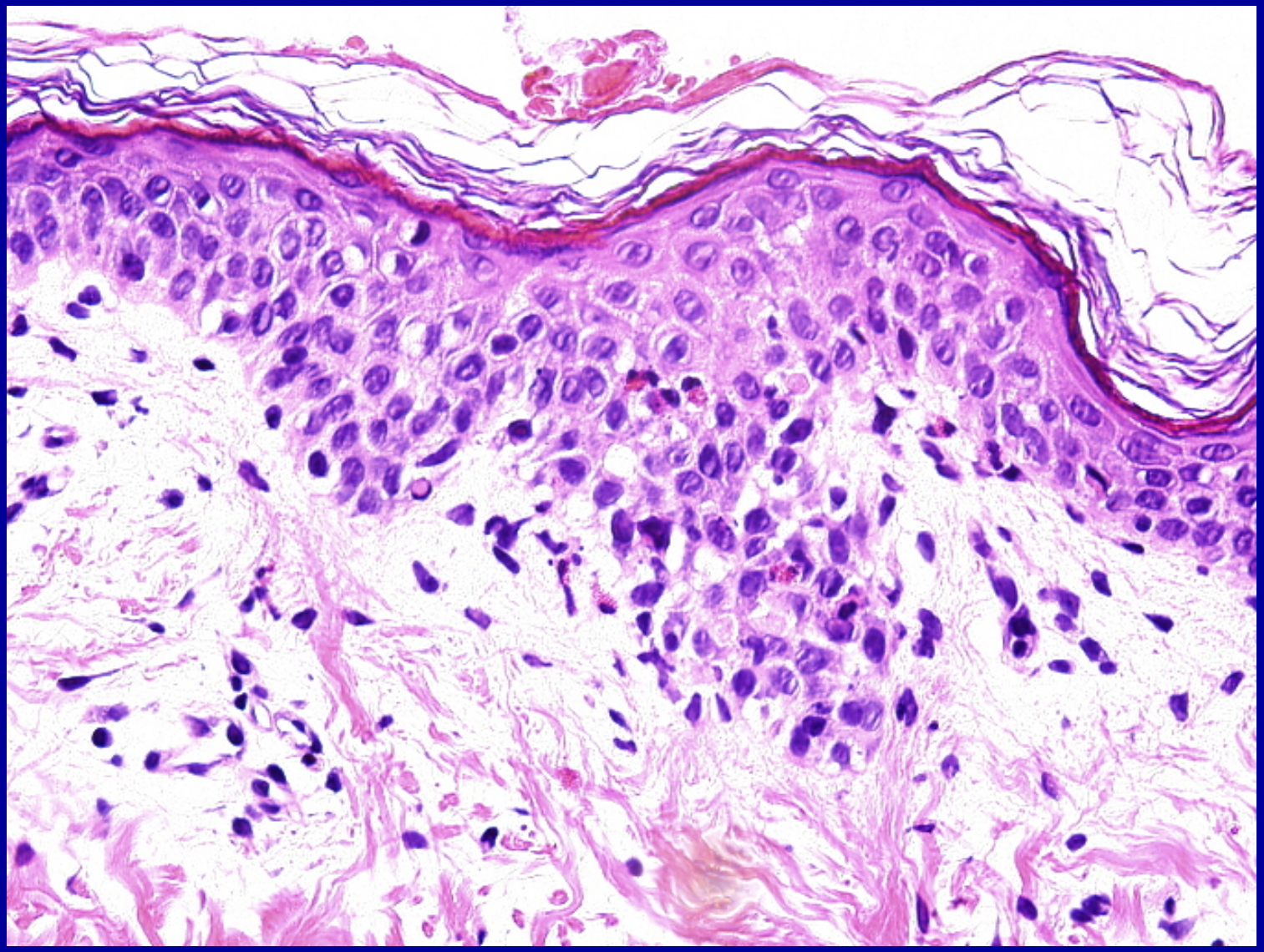

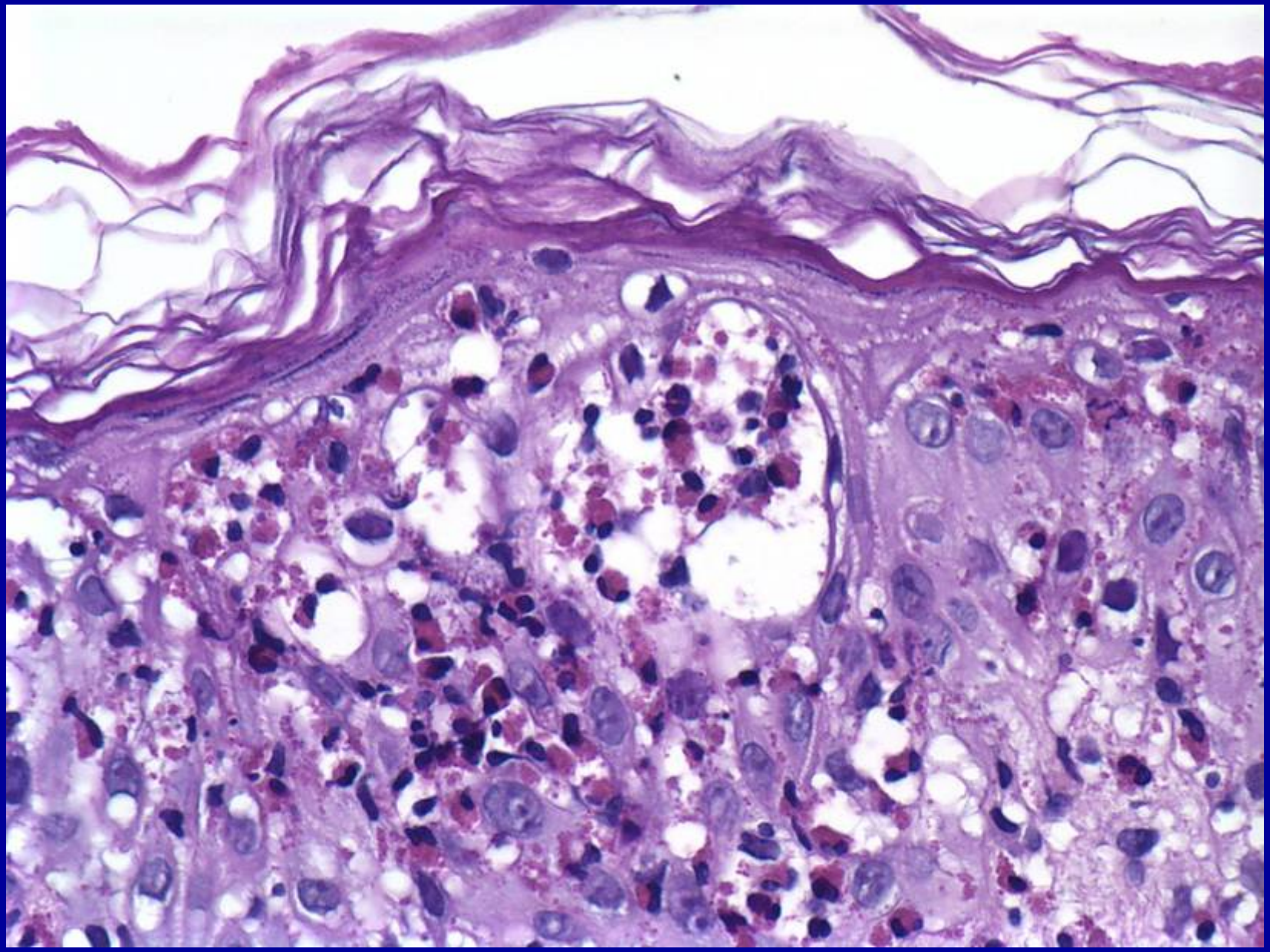

Case # 10

CE

Adult Mastocytosis – Urticaria Pigmentosa

CASE # 10
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: CE

Area(s) of Interest: Face, neck, trunk, and extremities

History: 38 yo WM presented in 2002 with a macular eruption over his sides and axillae associated with slight pruritus. Both the extent and symptomatology of the eruption increased with time leading to flares of wealing and intense pruritus.

Physical Examination: Over the face, trunk, and proximal extremities are scattered red-brown macules admixed with evenly pigmented tan-brown macules over sun-exposed areas, but also occurring in the axillae and other sun-protected areas.

Histopathology: A diffuse infiltrate is seen in the papillary dermis which consists of mononuclear cells with abundant, finely granular cytoplasm, and occasional eosinophils. There is vasodilation and interstitial edema. The mononuclear cells show strong cytoplasmic labeling when stained with tryptase.

Laboratory Studies: Tryptase 16.6 ng/L [0.4-10.9]

Diagnosis: Adult Mastocytosis – Urticaria Pigmentosa

Treatment and Course: Patient was placed on a leukotriene inhibitor as well as various antihistamines, including both H1 and H2 receptor antagonists to control the symptoms of histamine release. Despite avoidance of known triggers, he continues to have occasional urticarial flares. He carries an Epipen with him at all times, but has yet to experience any respiratory distress associated with these flares. He has a history of gastroesophageal reflux disease and irritable bowel syndrome which may or may not be related to his mast cell disease. Due to the clinical appearance of his lesions and the density of mast cells (four to five fold increase) seen on biopsy, he is best classified as adult urticaria pigmentosa. Mast cell density is greatest in confluent or nodular disease (diffuse or solitary mastocytoma) and least in telangiectasia macularis eruptiva perstans .

Discussion: Adult mastocytosis represents ~35% of mastocytosis cases and unlike the disease in childhood, is nearly always associated with an activation mutation in c-*kit*. Age of onset is between 20 and 40 years old. Organ systems involved include the skin, bone marrow, liver, spleen,

lymph nodes, and gastrointestinal tract, with skin being the most frequently involved organ system followed by the bone marrow. Cutaneous mastocytosis is divided into urticaria pigmentosa (most common), mastocytoma (solitary or multiple), diffuse cutaneous, and telangiectasia macularis eruptiva perstans (rare). Symptoms include wealing, pruritus, and flushing, especially upon stroking, which is Darier's sign. Systemic mastocytosis is defined by mast cell proliferation in extracutanous organ systems and should be considered in patients with a tryptase level greater than 20ng/mL. The spectrum of systemic disease spans from indolent to leukemic to sarcomatous. Symptoms include nausea, vomiting, diarrhea, palpitations, headache, dyspnea, and wheezing, though massive histamine release in cutaneous disease may also give rise to these symptoms. Skin involvement can be seen in the presence or absence of systemic involvement. Interestingly, it is rarely seen in cases of aggressive systemic disease. Treatment is aimed at avoiding triggers of mast cell degranulation and controlling symptoms of histamine release with antihistamines. Severe cutaneous disease responds to PUVA, but quickly recurs. Severe cases of systemic mastocytosis may require immunomodulatory agents or chemotherapy.

References:

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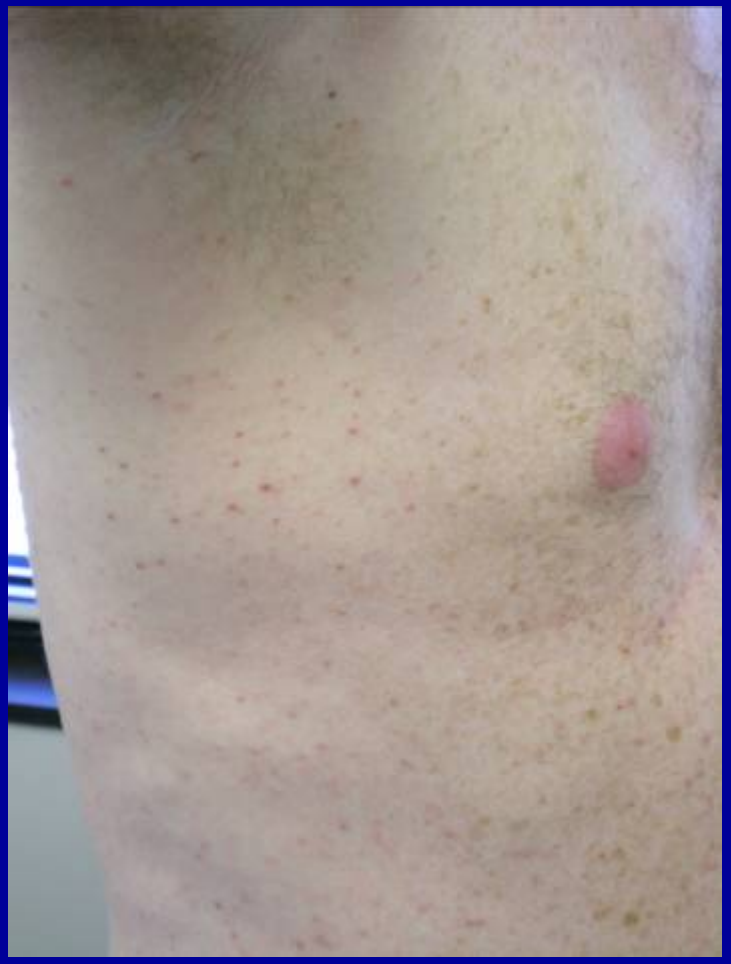
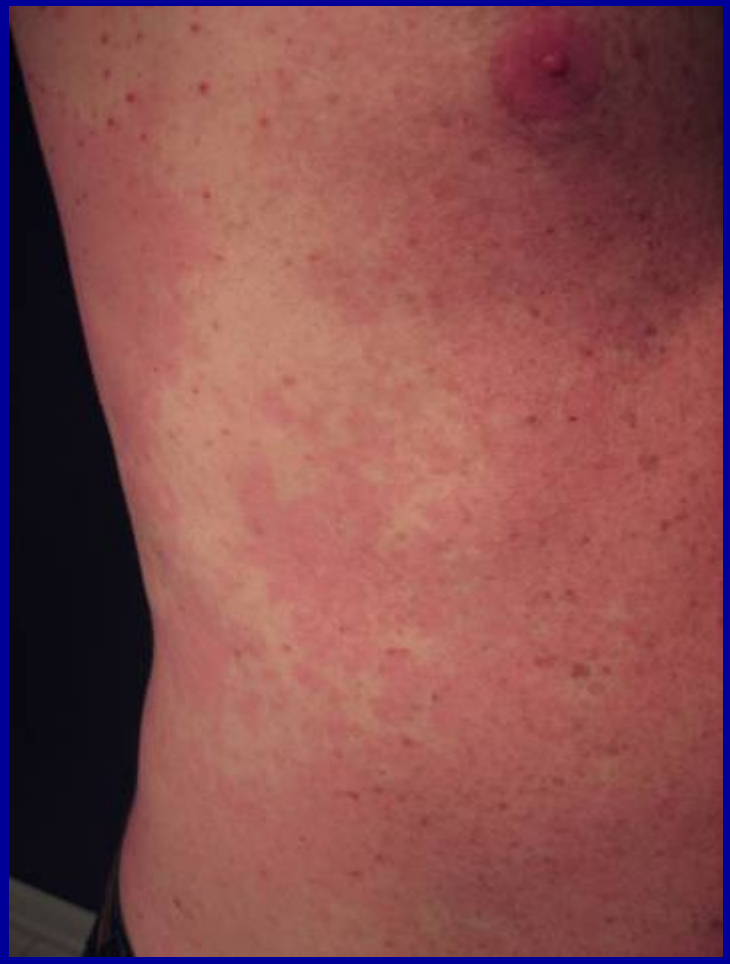
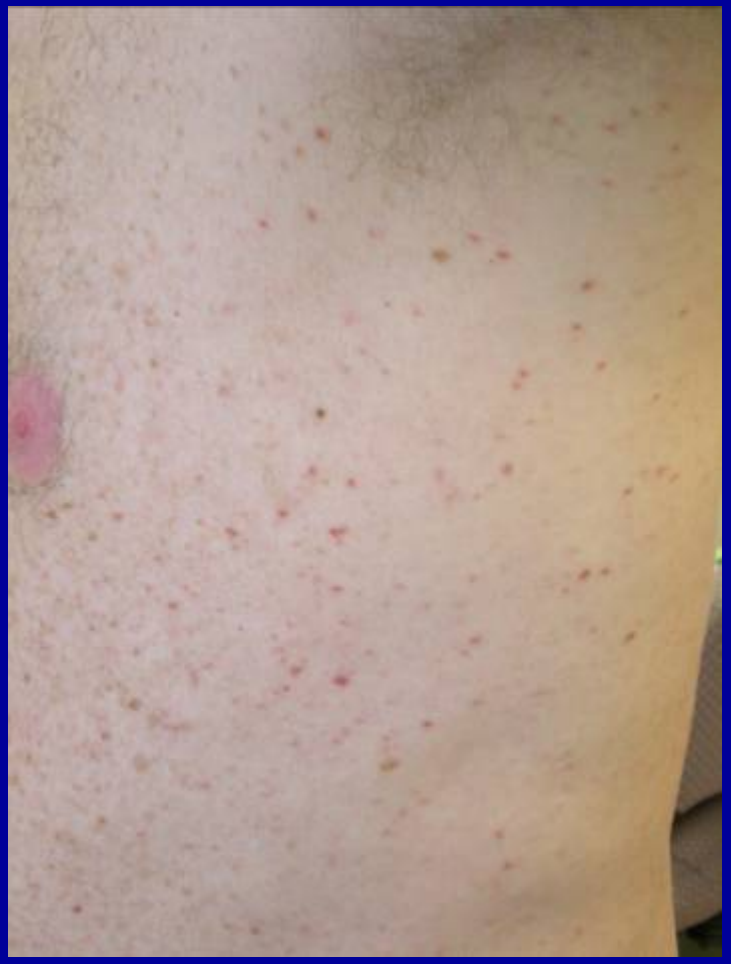
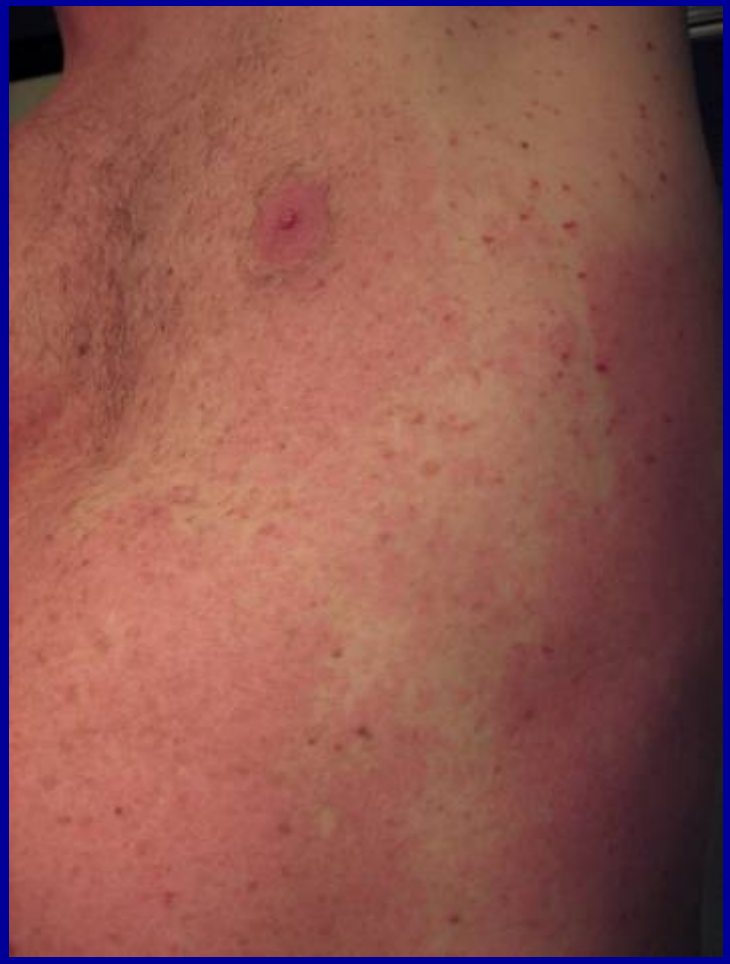
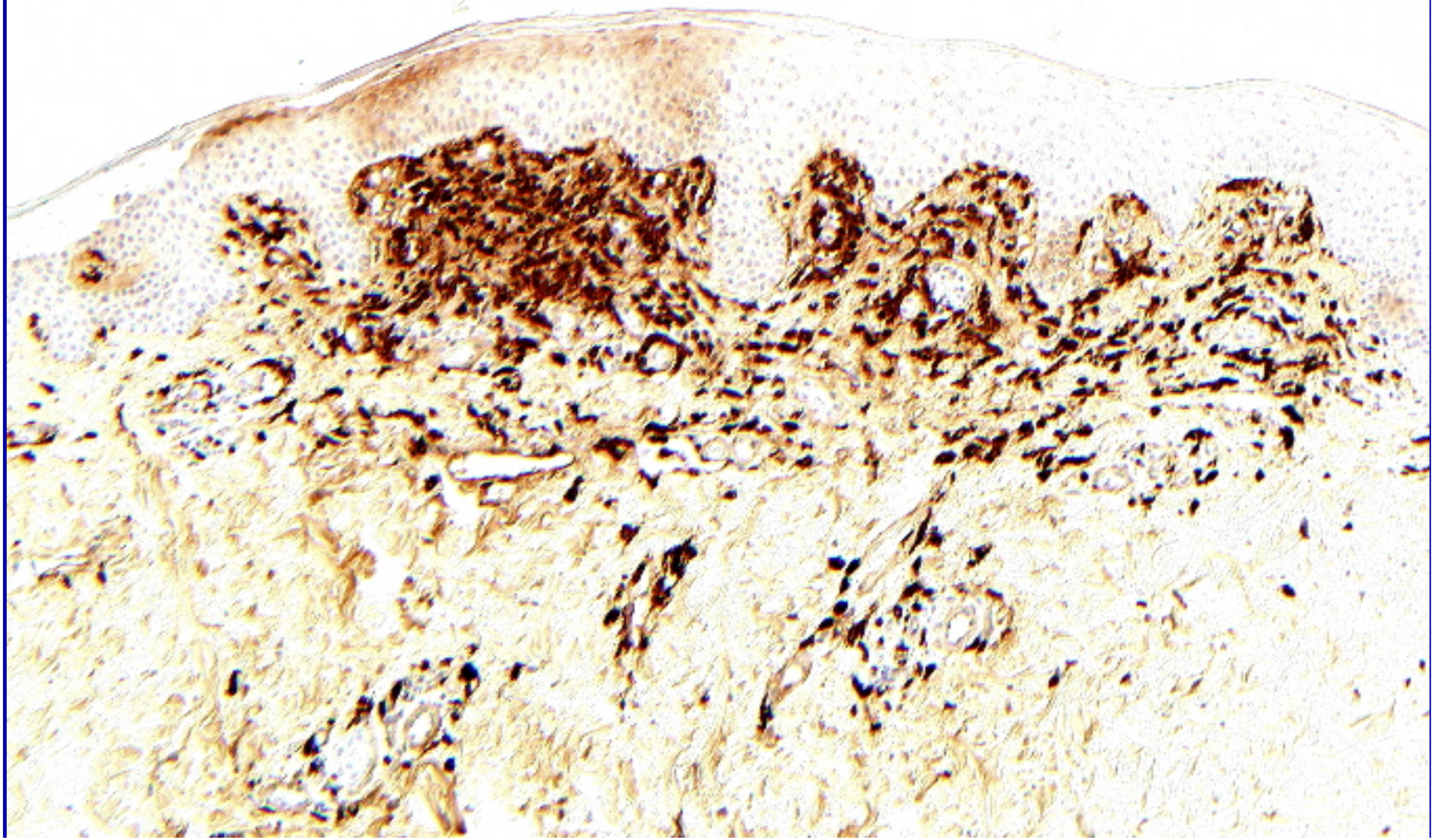








MAST CELL TRYPTASE STAIN



Case # 11

MD

Reticular Erythematous Mucinosis (REM)

CASE # 11
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: MD

Area(s) of Interest: Neck and chest

History: 43 year old Caucasian woman with a several year history of a persistent skin eruption over her neck and chest. It is exacerbated by heat and does not itch.

Physical Exam: Over the anterior neck and chest is an ill-defined, pink, reticulated pattern of papules and plaques. A background of actinic damage is seen with dyschromia, telangiectasias, and sub-mental sparing.

Histology: The epidermis is normal. There is a mild superficial and deep perivascular inflammatory infiltrate of lymphocytes. Mild solar elastosis is noted in the superficial dermis, but widely spaced collagen is seen in the reticular dermis. Colloidal iron stain shows increased dermal mucin.

Lab Studies: ANA, SSa, SSb, anti-Smith, and anti-ds DNA negative. C3, C4 levels WNL.

Diagnosis: Reticular Erythematous Mucinosis (REM)

Treatment and Course: The patient presented with a one year history of the eruption in 2004. Differential diagnosis at that time included LE and REM. A biopsy showed findings seen in both, but due to normal labwork, and the morphology and distribution of the eruption, a diagnosis of REM was favored. The patient was placed on Plaquenil 200mg BID with cessation of progression, but no clearance of the eruption. She has remained stable on Plaquenil for 5 years with worsening of the erythema upon trials of discontinuation. Various topical therapies including Westcort 0.2% cream, Elidel 1% cream, and Differin 0.1% gel have not provided much additional benefit. Most recently, we have initiated treatment with the Pulsed Dye Laser with moderate response, though we have only treated small areas at a time. Though some degree of the eruption has persisted clinically, a repeat biopsy in 2008 showed persistence of the perivascular infiltrate, but resolution of the mucin deposition.

Discussion: REM is a rare primary cutaneous mucinosis characterized by pink papules and plaques which often form a reticular or netlike pattern. It is seen most frequently over the chest and back of middle-aged women, though cases in men and children can be seen. It is exacerbated by sunlight,

heat, OCPs, pregnancy, and menopause. It is usually asymptomatic, but can be pruritic. Histologically, it can be indistinguishable from tumid lupus erythematosus and many consider these entities to exist on a spectrum. Lab abnormalities, circulating antibodies, or positive findings on direct immunofluorescence are rarely seen in REM. Antimalarials are typically effective in clearing the eruption in one to two months. Alternative treatment options include topical or systemic steroids, topical calcineurin inhibitors, tetracyclines, UVB, and pulsed-dye laser. REM has been documented to occur in patients with various other medical conditions including endocrine, autoimmune, infectious, and malignant disorders, though no direct correlation has been found. Reports of cases affecting siblings and twins suggest a possible genetic predisposition.

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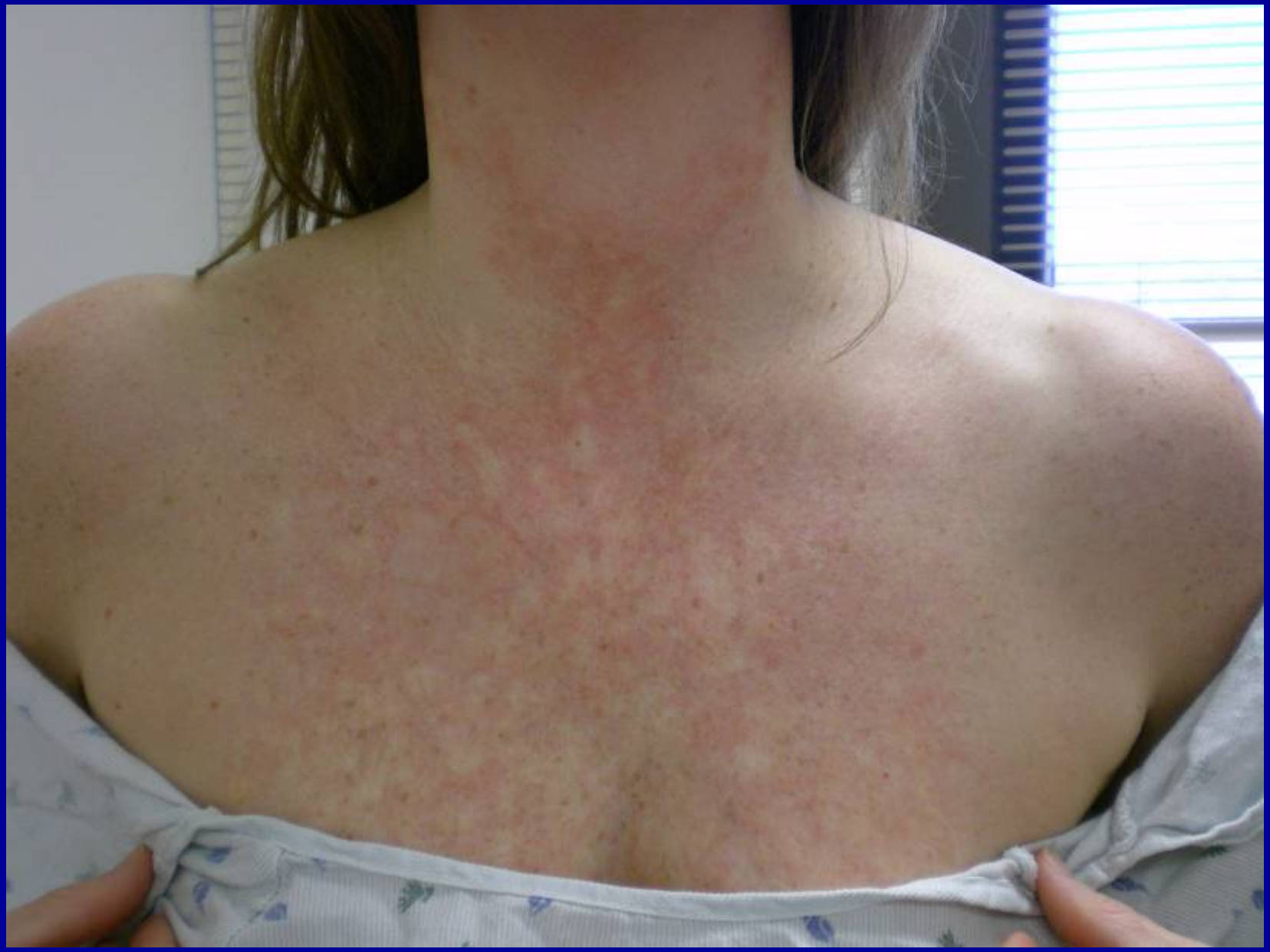

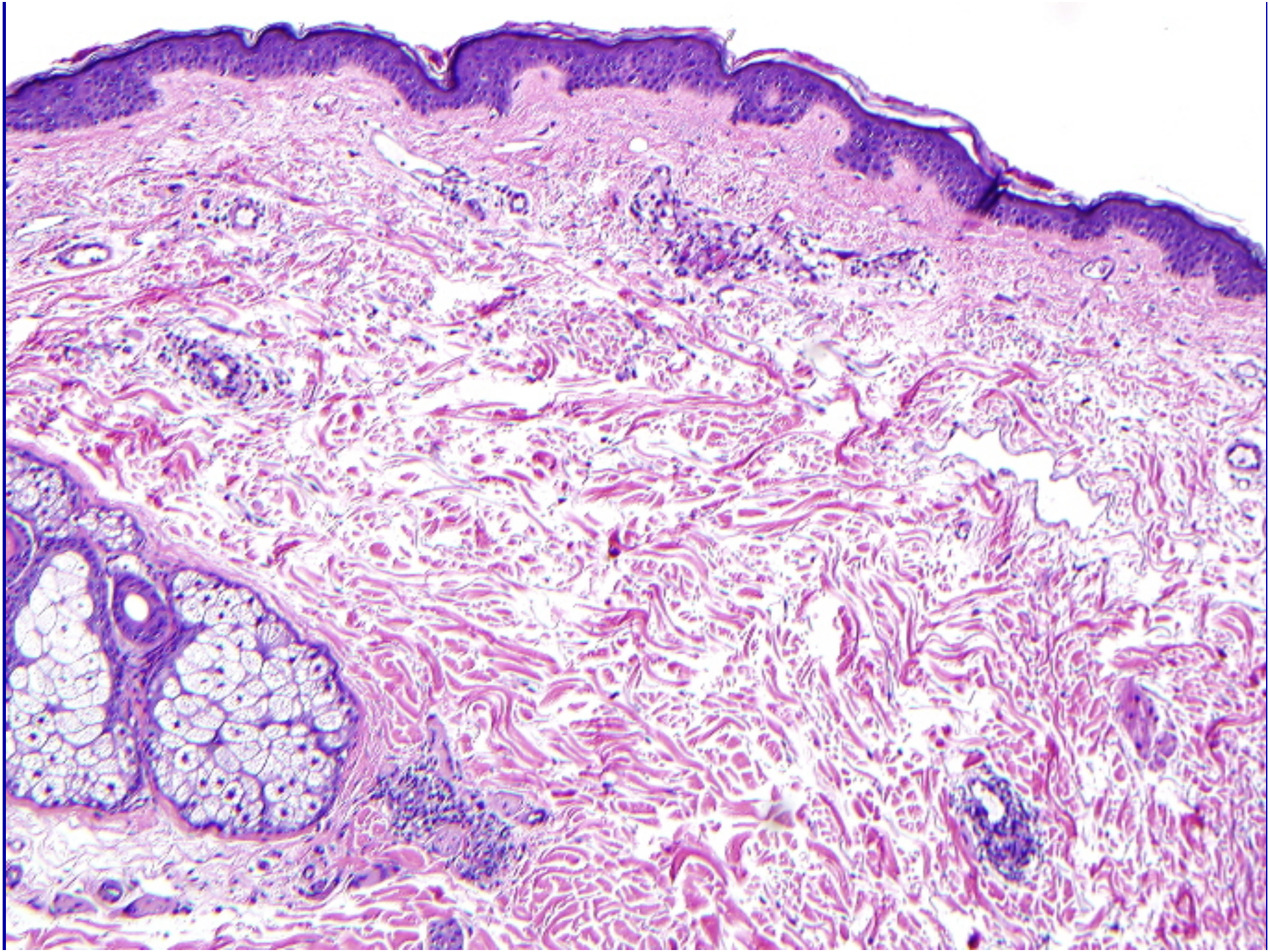

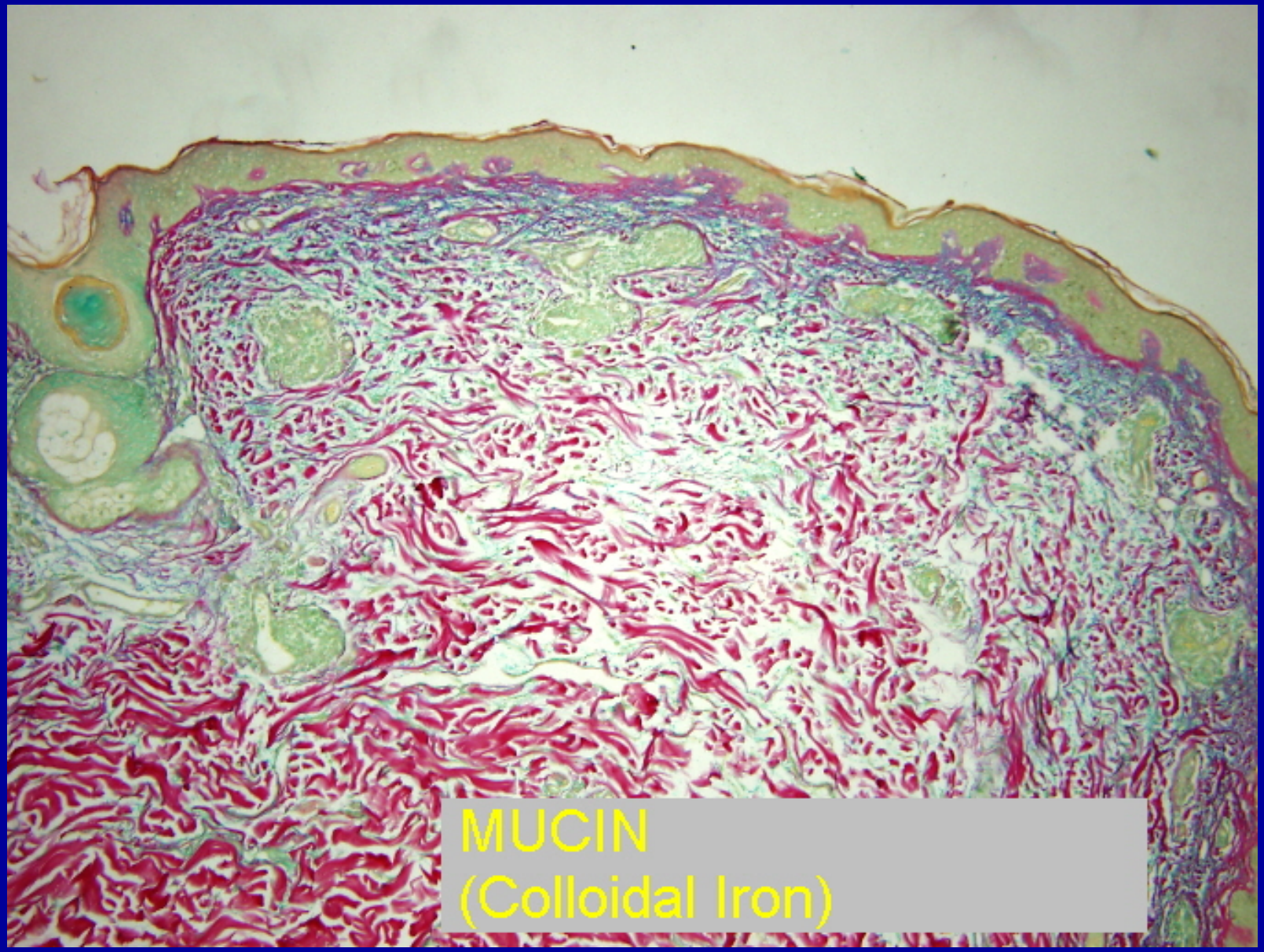
MUCIN
(Colloidal Iron)

Case #12

GC

Viral-associated Trichodysplasia of Immunosuppression

CASE # 12
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: GC

Areas of Interest: Face, trunk, and extremities

History: A 5-year-old male, with history hypoplastic left heart syndrome, underwent cardiac transplant secondary to intravenous inotropic dependence. Post-transplant, the patient was placed on mycophenolate mofetil, tacrolimus and prednisone. The patient developed donor-specific antibodies post-transplant. To combat against antibody mediated rejection, the patient received plasmapheresis, IVIG, rituximab and cylcophosphamide. Following this therapy, the patient was restarted on tacrolimus and prednisone. Approximately one year following the transplant, the patient developed an eruption on the face that progressed to involve the trunk and extremities.

Past Medical History: The patient has a history of a rare genetic disorder, Kabuki syndrome which is characterized by a distinctive faces of long palpebral fissures with eversion of outer third, arched eyebrows with sparse outer half, prominent eyelashes, prominent misshapen ears, and depressed nasal tip. These patients also have skeletal abnormalities, cognitive delay, and congenital heart defects

Family History: Noncontributary

Physical Exam: Numerous spiny, follicular papules involving primarily the central face, trunk and proximal thighs. Thickenings of the skin of the face, particularly the nose, chin and ears to leonine features

Histopathology: Histologic sections revealed numerous enlarged, bulbous anagen hairs. A thin layer of basophilic, germinative cells transitioned to inner root sheath-type cells containing numerous enlarged trichohyaline granules. The inner root sheath-like cells abruptly cornified without the presence of a granular layer. No hair shafts or papillae were identified. Outer root sheath-type epithelium was present only at the upper half of affected bulbs. The upper segment of the follicle showed a dilated, hyperkeratotic and shaftless infundibulum. Vacuolated keratinocytes with pyknotic nuclei and coarse keratohyaline granules findings consistent with viral cytopathic effects were seen in the upper layers of the perifollicular epithelium.

Diagnosis: Viral-associated trichodysplasia of immunosuppression (VATD)

Treatment and Course: The patient was placed on cidofovir 3% cream topically twice a day. This topical regimen had little effect on our patient's eruption. One case report describes the observation of discontinuing oral valganciclovir with the onset of VATD; empirically restarting valganciclovir brought about the resolution of the eruption.[8] Based on this report, out patient was placed on systemic valganciclovir. This systemic therapy has improved our patient's VATD, but did not provide total resolution. Our patient unfortunately developed bone marrow suppression secondary to systemic valganciclovir.

Comment: Originally termed "trichodysplasia spinulosa", VATD has been linked to various other immunosuppressive medications primarily in transplant patients. Most recently, VATD has been recognized in patients receiving chemotherapy for acute and chronic lymphocytic leukemia and non-Hodgkin's lymphoma. Electron microscopy of other reported cases demonstrate intranuclear, icosahedral viral particles morphologically suggestive of papovavirus family.

polyoma

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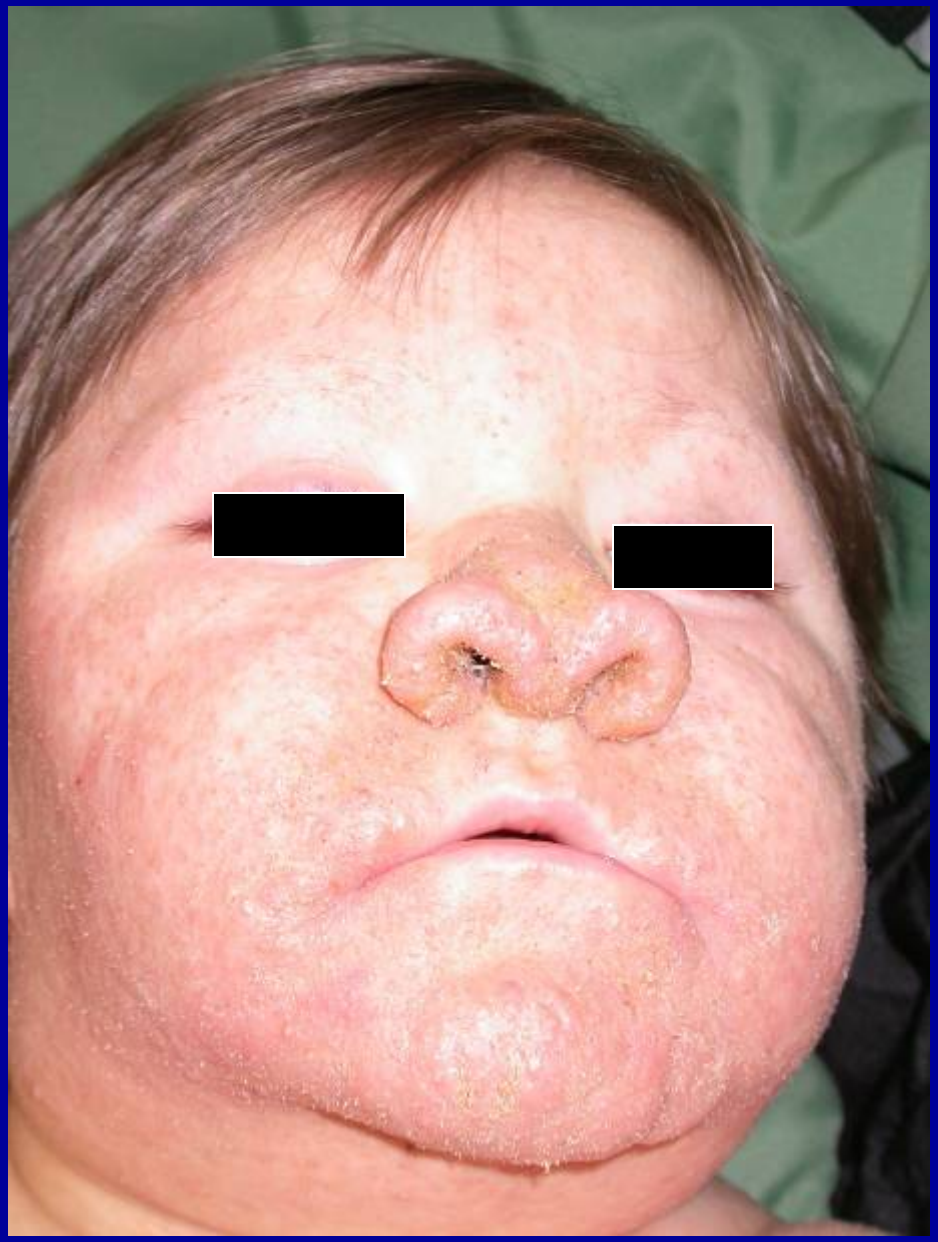

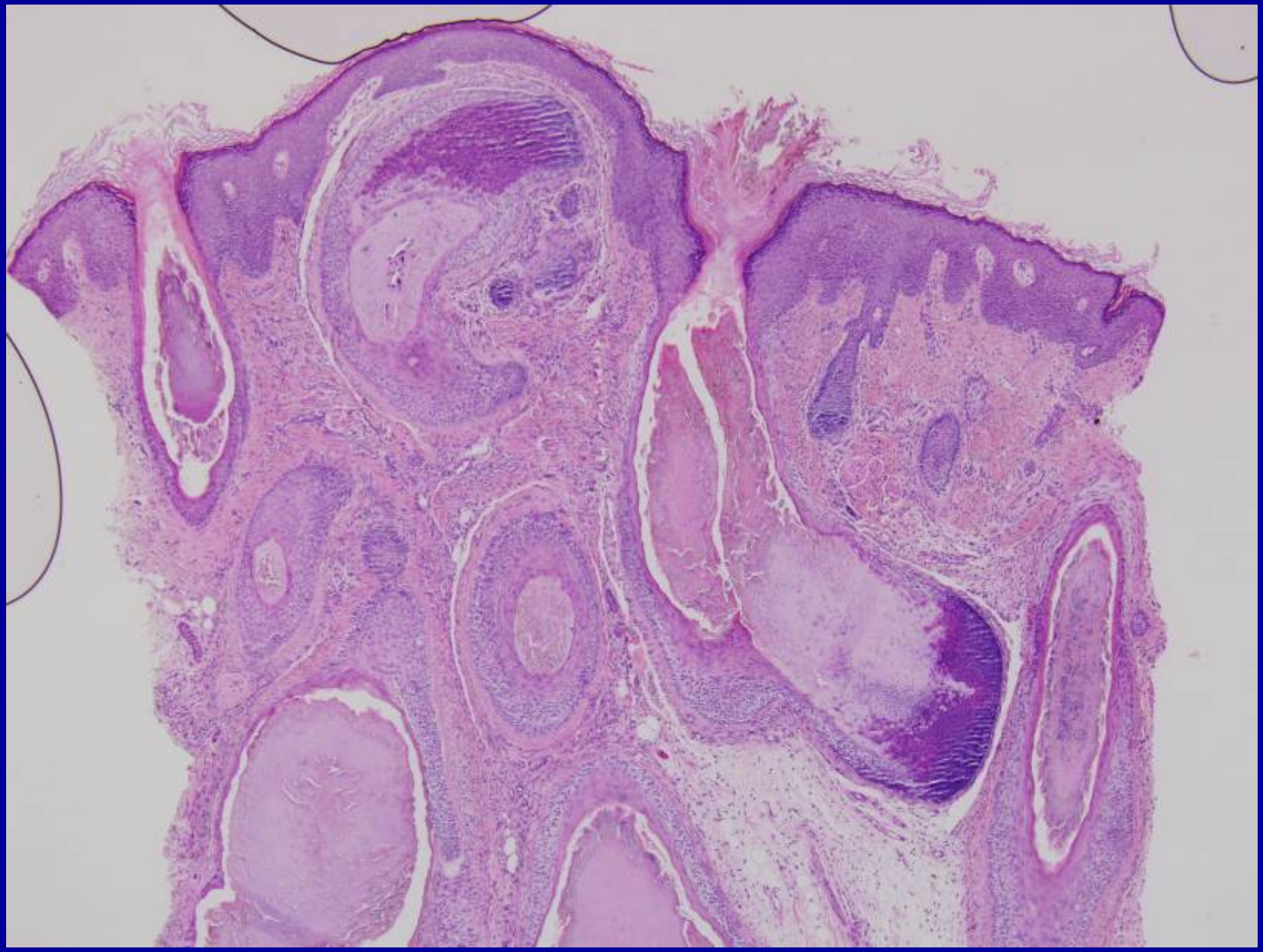

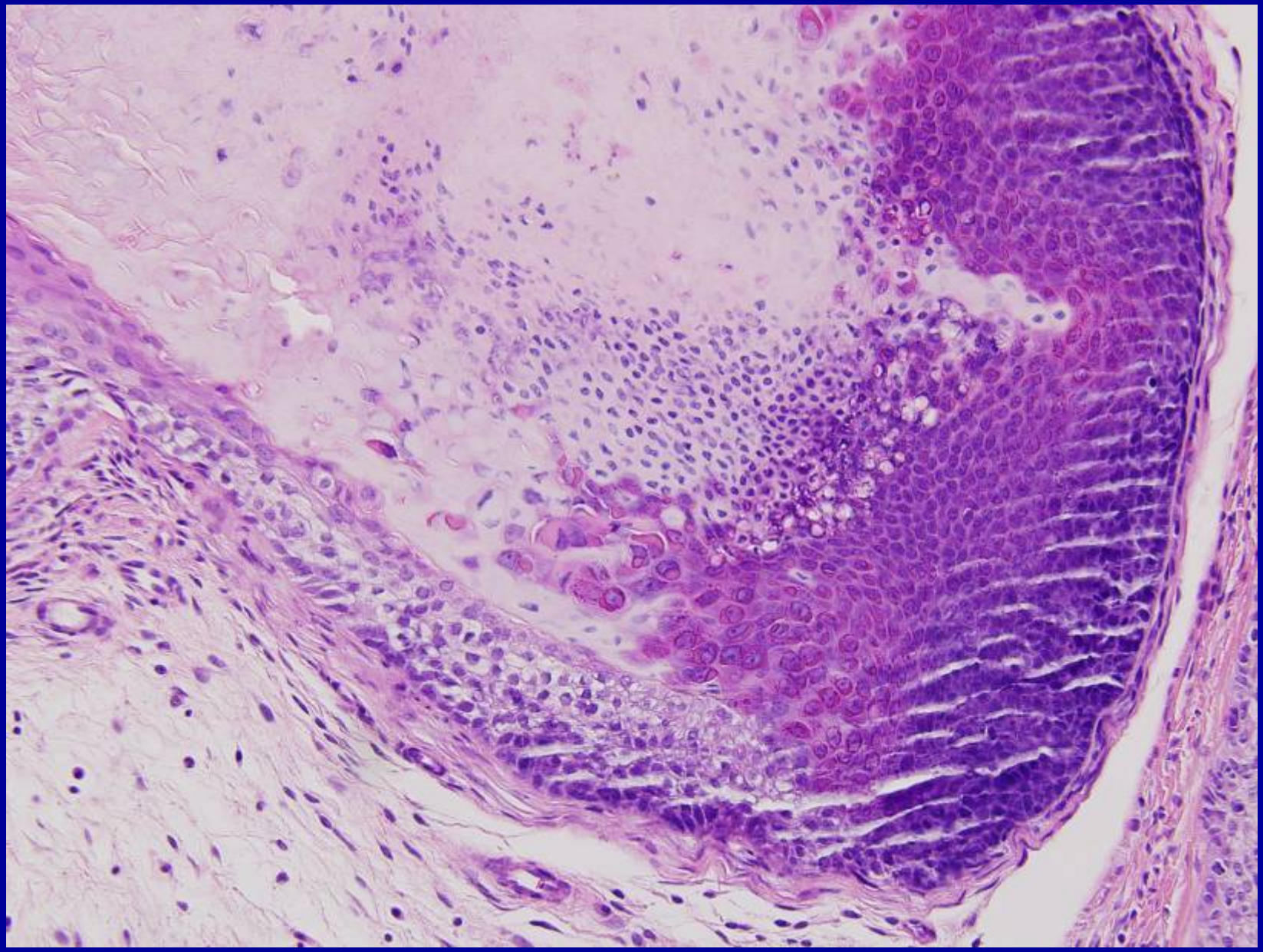

Case #13

MG

Exogenous Ochronosis

CASE # 13
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	MG
Areas of Interest:	Face
History:	65 year-old woman with history of sun exposure applied greater than 4 year application of a skin lightening cream to her face. She reports a gradual darkening of her skin despite discontinuation of the cream.
Past Medical History:	No history of premature osteoarthritis symptoms, renal calculi, scleral color change or valvular heart disease. Otherwise non-contributory.
Family History:	No family history of alkaptonuria.
Physical Examination:	blue-gray patches with areas of erythema and hypopigmentation distributed over forehead and malar cheeks. Spares perioral area and conjunctiva.
Histopathology:	There are yellow-brown, banana-shaped ochronotic fibers deposited in the papillary dermis
Diagnosis:	Exogenous ochronosis
Treatment and Course:	Despite discontinuation of topical hydroquinone there has been little improvement in the pigmentary alterations on the patient's face. She has attempted several cosmetic procedures such as chemical peel and dermabrasion. The areas of hypopigmentation correspond to the mechanical exfoliation.
Comment:	Ochronosis exists in both an endogenous and exogenous form. Endogenous ochronosis or alkaptonuria is an autosomal recessive disorder characterized by a deficiency of homogentisic acid oxidase, an enzyme necessary for the conversion of homogentisic acid to acetoacetic and fumaric acids. Affected individuals have an accumulation of homogentisic acid which is an insoluble pigment that deposits into various tissues including cartilage, skin, and cardiac valves. Exogenous ochronosis is confined to the skin, and is characterized by blue-black to slate-gray color. The discoloration most commonly is a result from the use of hydroquinone, although other agents have been implicated such as phenol, mercury, picric acid, antimalarials, and resorcinol.

The etiology of hydroquinone-induced pigmentation in exogenous ochronosis is still speculative. Some data suggests hydroquinone may cause a local inhibition of homogentisic acid oxidase, leading to local accumulation of homogentisic acid which polymerizes to form pigment. There have been suggestions that sunlight, oxidized by-products of hydroquinone, and melanocytes also play a role in pigment deposition.

Reference:

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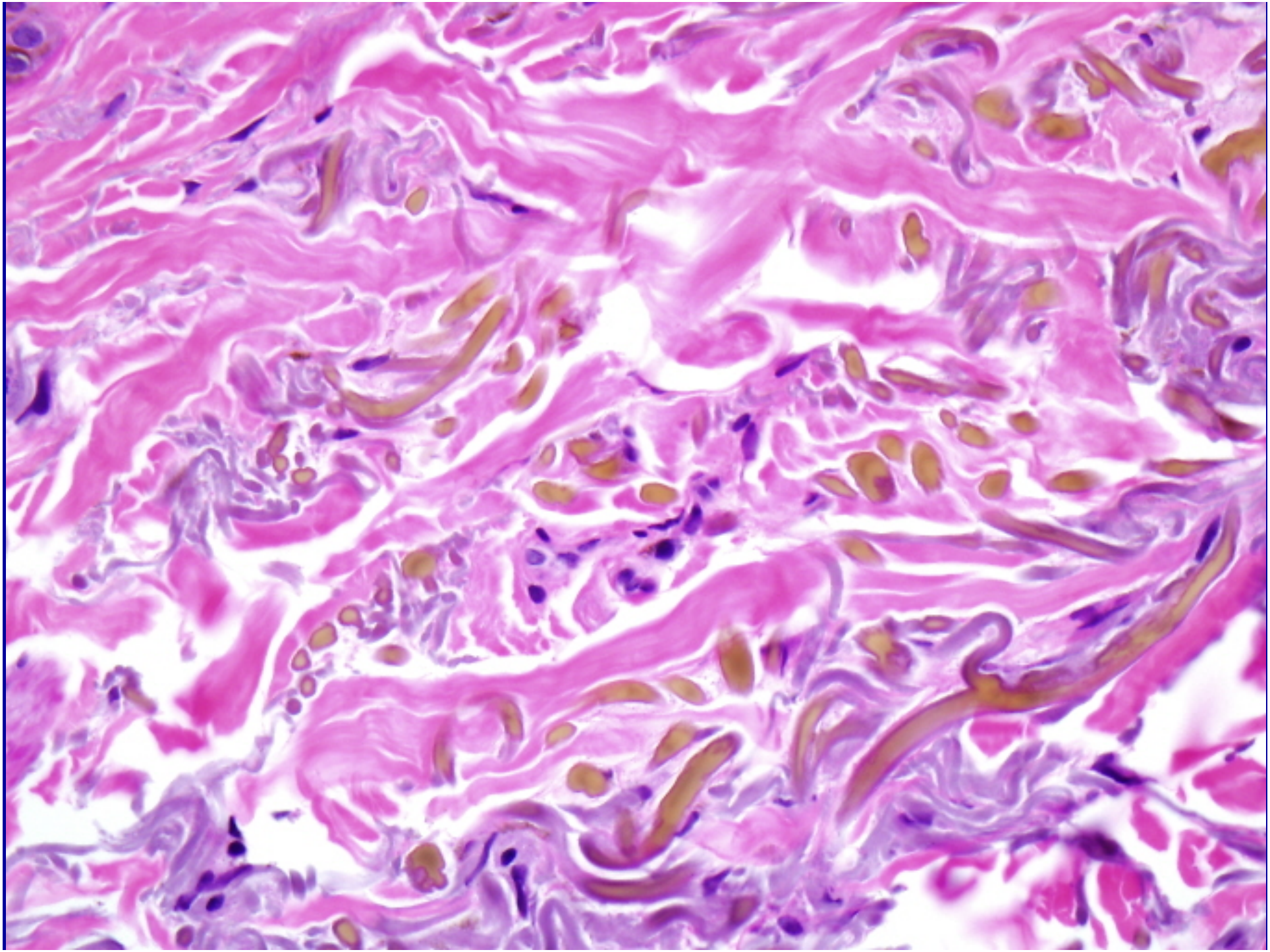

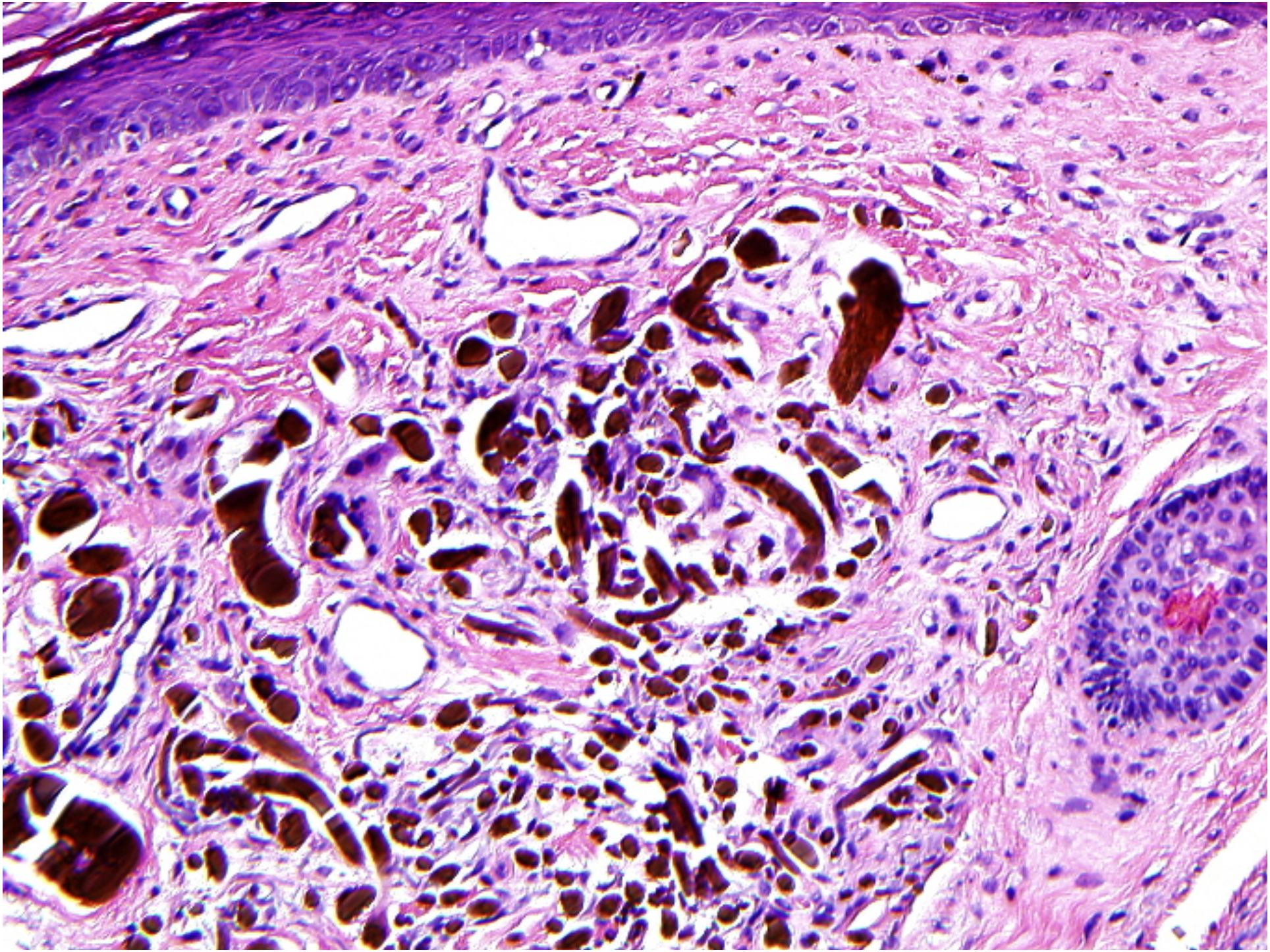

Case #14

WS

Granuloma Annulare and Necrobiosis Lipoidica Overlap

CASE # 14
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	WS
Area of Interest:	Lower legs, abdomen, and arms
History:	This is a 79 year-old woman with a long history of diabetes mellitus type II. She reports her skin lesions began 2007 initially on her bilateral lower extremities. She notes the lesions progressed to other areas of her body, most notably on her abdomen and extremities.
Past Medical History:	diabetes mellitus type II treated with insulin and oral agents; hypertension
Family History:	Both parents with a history of diabetes mellitus, and hypertension. The patient's mother also has a history of heart disease.
Physical Examination:	small firm erythematous to flesh-colored dermal papules distributed over the abdomen and bilateral upper extremities. Yellow indurated plaques with central atrophy on bilateral shins.
Histopathology:	Palisading granulomas with necrobiotic collagen and mucin accumulation within the dermis. Dermal interstitial infiltrate of histiocytes and multinucleated giant cells
Laboratory data:	Noncontributory
Diagnosis:	Granuloma annulare and necrobiosis lipoidica overlap
Treatment and Course:	The patient has tried various topical steroids and topical tacrolimus with little improvement. She had significant improvement with oral ketoconazole, however this effect was temporary. The eruption recurred despite continued therapy. Other systemic therapies include S.S.K.I and dapsone. The patient was recently began combination therapy with Minocycline 100mg BID and nicotinamide 500mg TID.
Comment:	Necrobiosis lipoidica (NLD) and granuloma annulare(GA) are both characterized by granulomatous inflammation and degenerative collagen. These diseases are of unknown etiology, but it has been suggested they are part of the spectrum of the same disease. Clinical and pathologic differentiation can be made difficult as both conditions share several histological similarities. It is rare to see both

of these conditions to be seen together in the same patient simultaneously.

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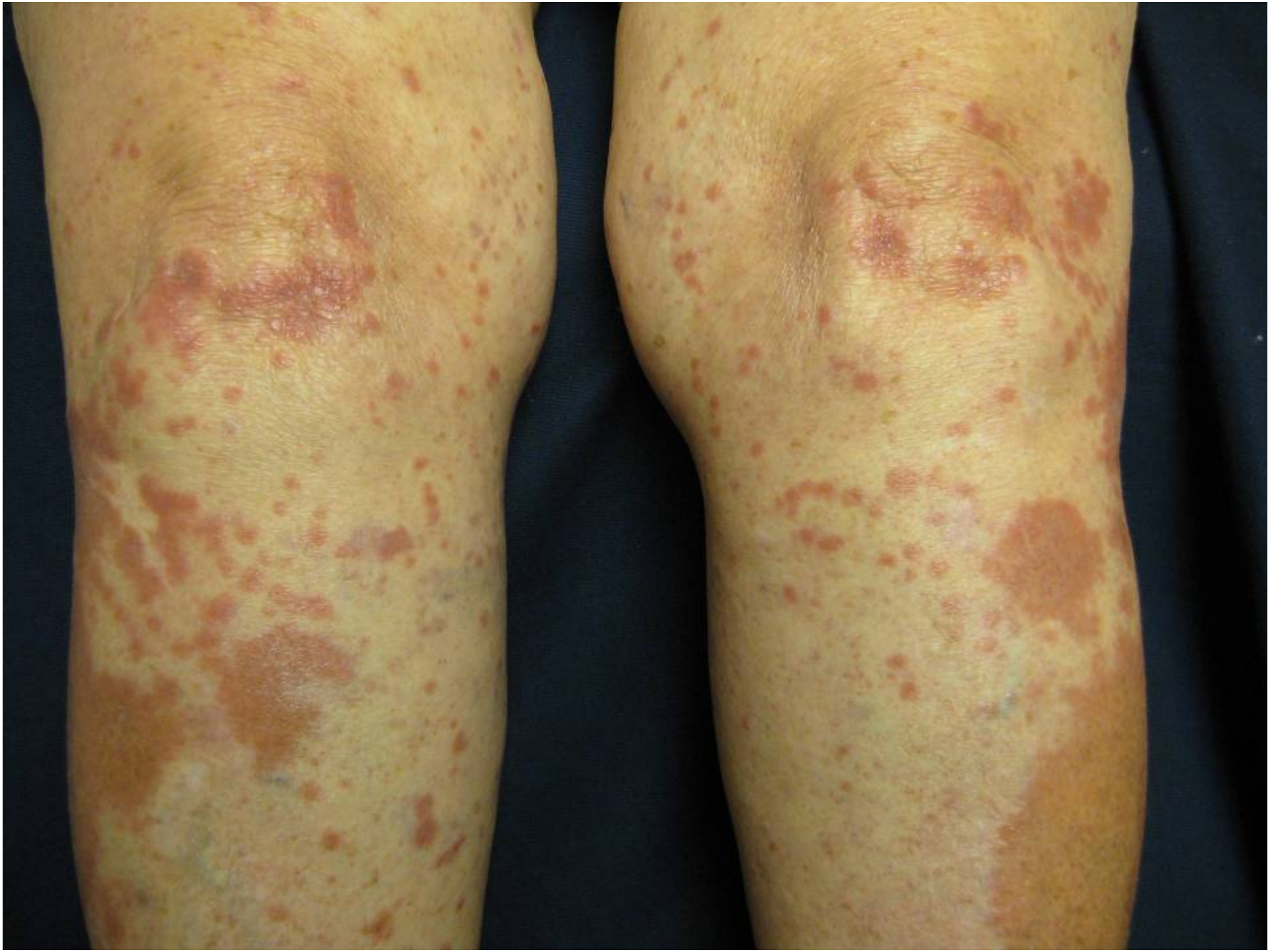
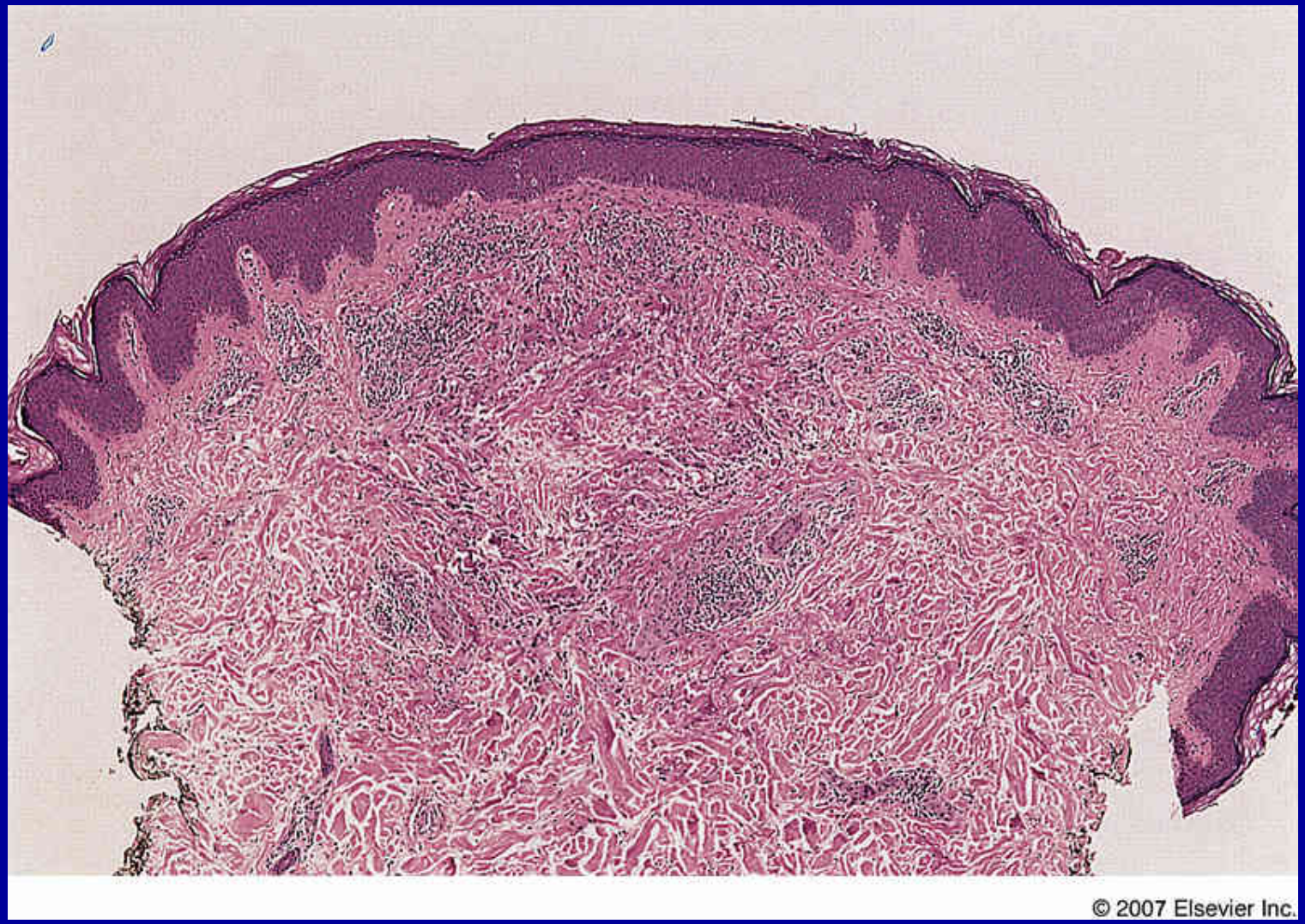
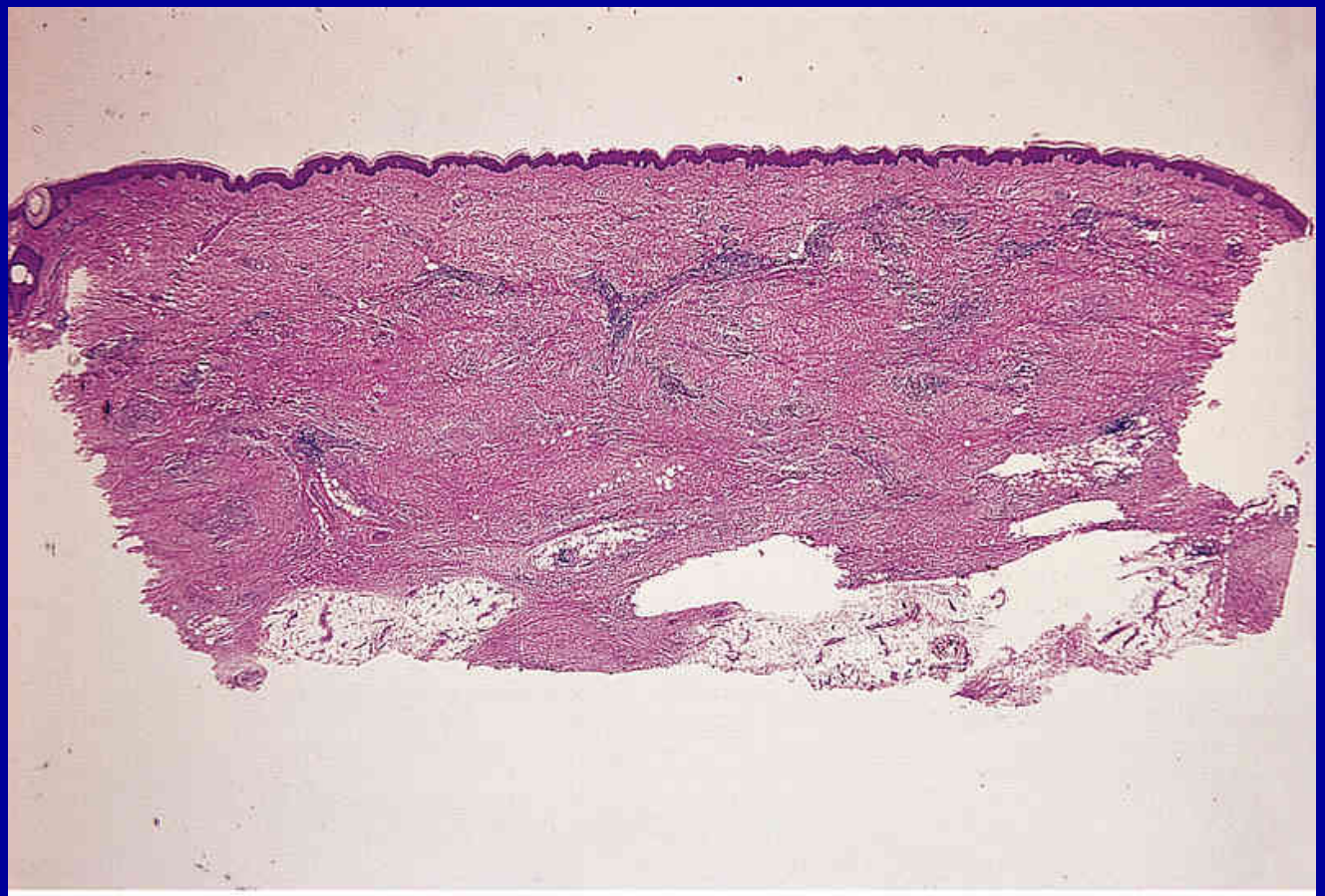


Granuloma Annulare



Necrobiosis Lipoidica



Case #15

LH

Bloom’s Syndrome with
Lupus-like
Histopathologic Features

CASE # 15
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: LH

Area of Interest: Face, dorsal forearms, and dorsal hands

History: This 19 year old Caucasian male with known history of Bloom Syndrome (BS) was referred to the dermatology clinic for management options of worsening facial erythema. The cutaneous manifestations of BS initially included telangiectatic, erythematous patches on the malar cheeks which later spread to involve more of the face, dorsal forearms, and hands. They are exacerbated by sunlight. The diagnosis of BS was confirmed at 4 years old by fibroblast metaphase cytogenetic analyses, and was unique in that it occurred in a patient of non-Ashkenazi Jew and non-consanguineous parentage.

Past Medical History: Poor visual acuity, mild intercurrent gastrointestinal and pulmonary infections throughout childhood, congenital ventral penile hypospadias. Immunologic studies have reproducibly shown low IgG, IgM, and IgA with normal T-cell subsets. No history of solid or hematologic malignancies.

Family History: No family history of BS

Physical Examination: When he was first referred to dermatology at 16 years old, an ulcerative cheilitis with oral ulcers was apparent, along with photodistributed, blancheable, scaling, erythematous, telangiectatic plaques and papules of the face, ears, extensor forearms and dorsal hands. Focal eyelash, eyebrow, and scalp alopecia was observed. Discoid scars were apparent at the right lower cheek, forearms, and ears. Other dermatologic stigmata include café au lait macules present in the bilateral popliteal fossae and on the superior right eyelid. The patient also exhibited red hair, blue eyes, microcephaly, distinct triangular facies, short stature and persistent hyperemia of the right bulbar conjunctiva.

Pathology: Epidermis with effacement of the rete ridge pattern, focal plugging of the follicular orifices, necrotic keratinocytes and vacuoles within the basal and parabasal epidermis were seen. Basement membrane was overtly thickened. A superficial and deep perivascular and prominent periadnexal infiltrate composed of lymphocytes and histiocytes was observed. A colloidal iron stain showed a moderate amount of mucin among collagen bundles in the

dermis. DIF demonstrated a focal linear presence of shaggy fibrin at the dermoepidermal junction. A few scattered IgM cytoid bodies were also seen along the basement membrane zone.

Laboratory Studies: ANA, dsDNA, complement levels (C3 and C4) normal

Diagnosis: Bloom syndrome with lupus-like histopathologic features

Treatment and Course: Patient has been undergoing pulse dye laser treatment to treat his telangectasias with good results. He is followed by genetics and is otherwise stable.

Comment: BS is an autosomal recessive genodermatosis which results from inactivating mutations in the BLM gene. The average life expectancy is 18 years, owing to a high rate of hematologic and solid organ epithelial malignancies. Patients also tend to be deficient in at least one immunoglobulin class, leading to increased infection rates. Telangiectatic erythema is the most common clinical finding. Skin lesions are described as "lupus-like" since they are usually distributed over the malar cheeks, are photosensitive, and relatively transient, lasting from days to one week in duration. The characteristic facial appearance of BS patients involves hypoplasia of the malar area, small mandible, dolichocephaly, and prominent nose.

Dermatopathologic findings in BS remain poorly defined. Nonetheless, a "lupus-like" histopathology has been described in several cases. This case in particular showed striking features of a lupus-like histology, including the DIF pattern showing a linear deposition of fibrinogen. This raises the question whether systemic lupus erythematosus and BS exist concurrently in selected patients or a lupus-like morphology is a clinical expression of BS.

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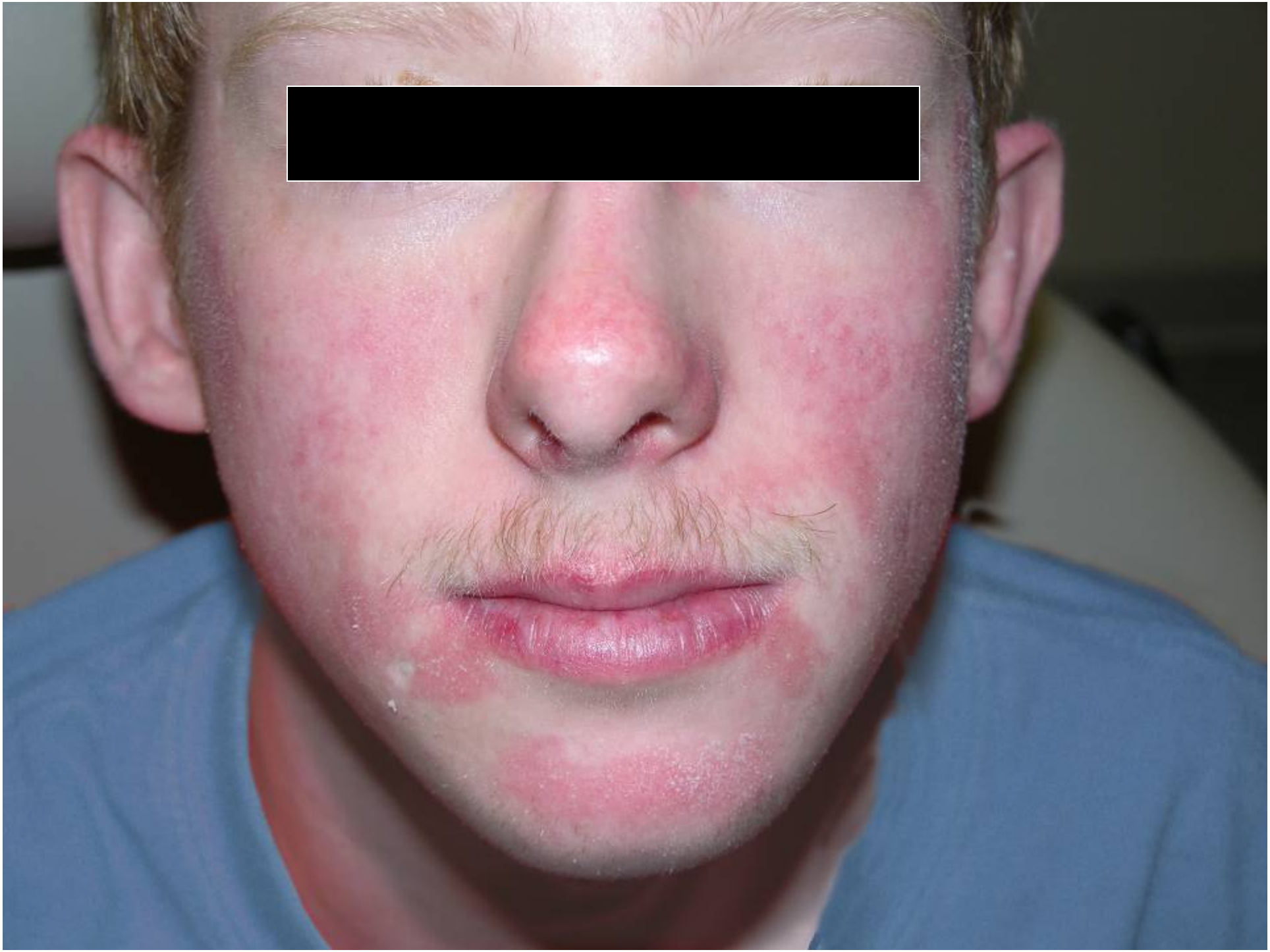

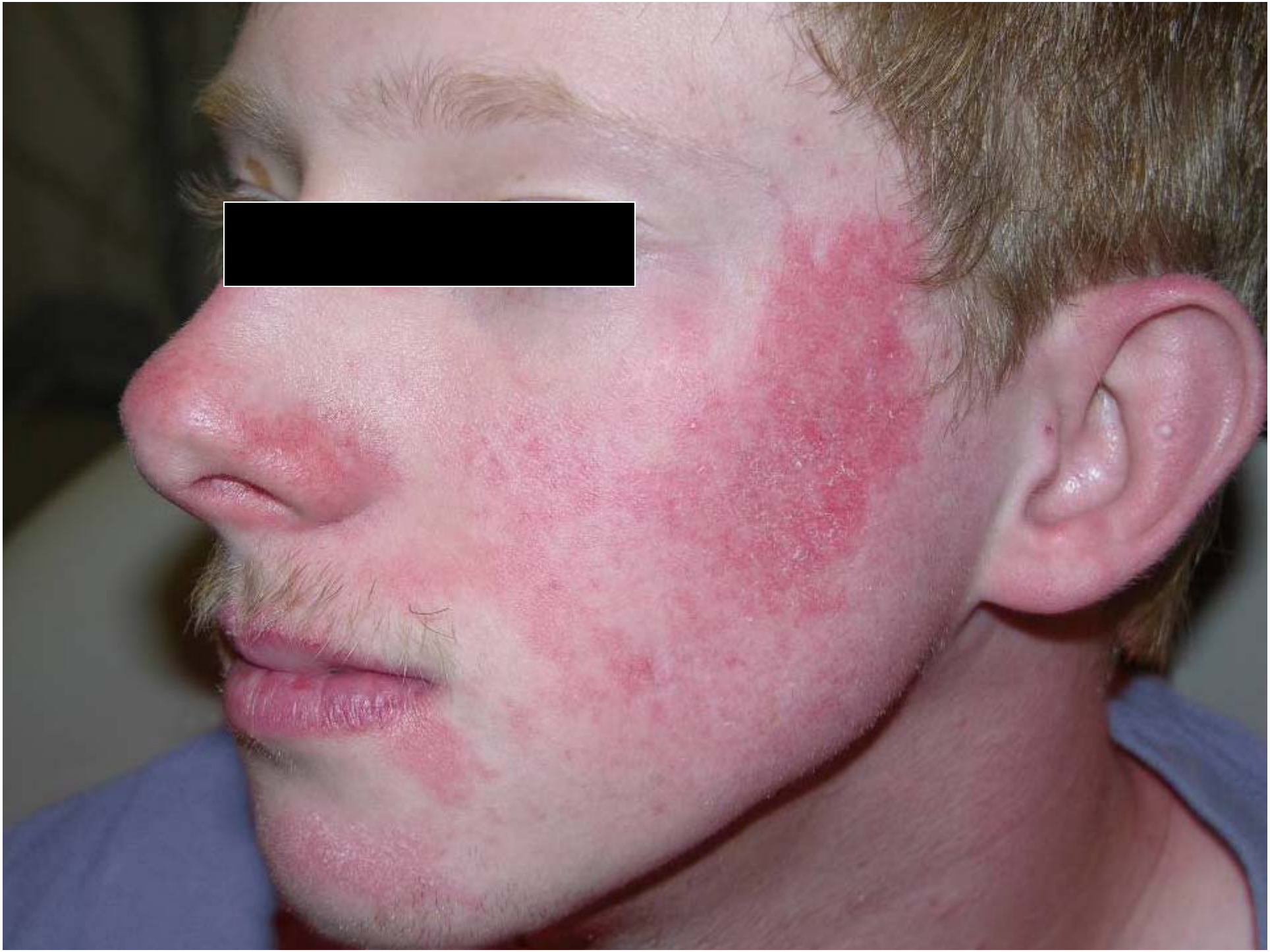

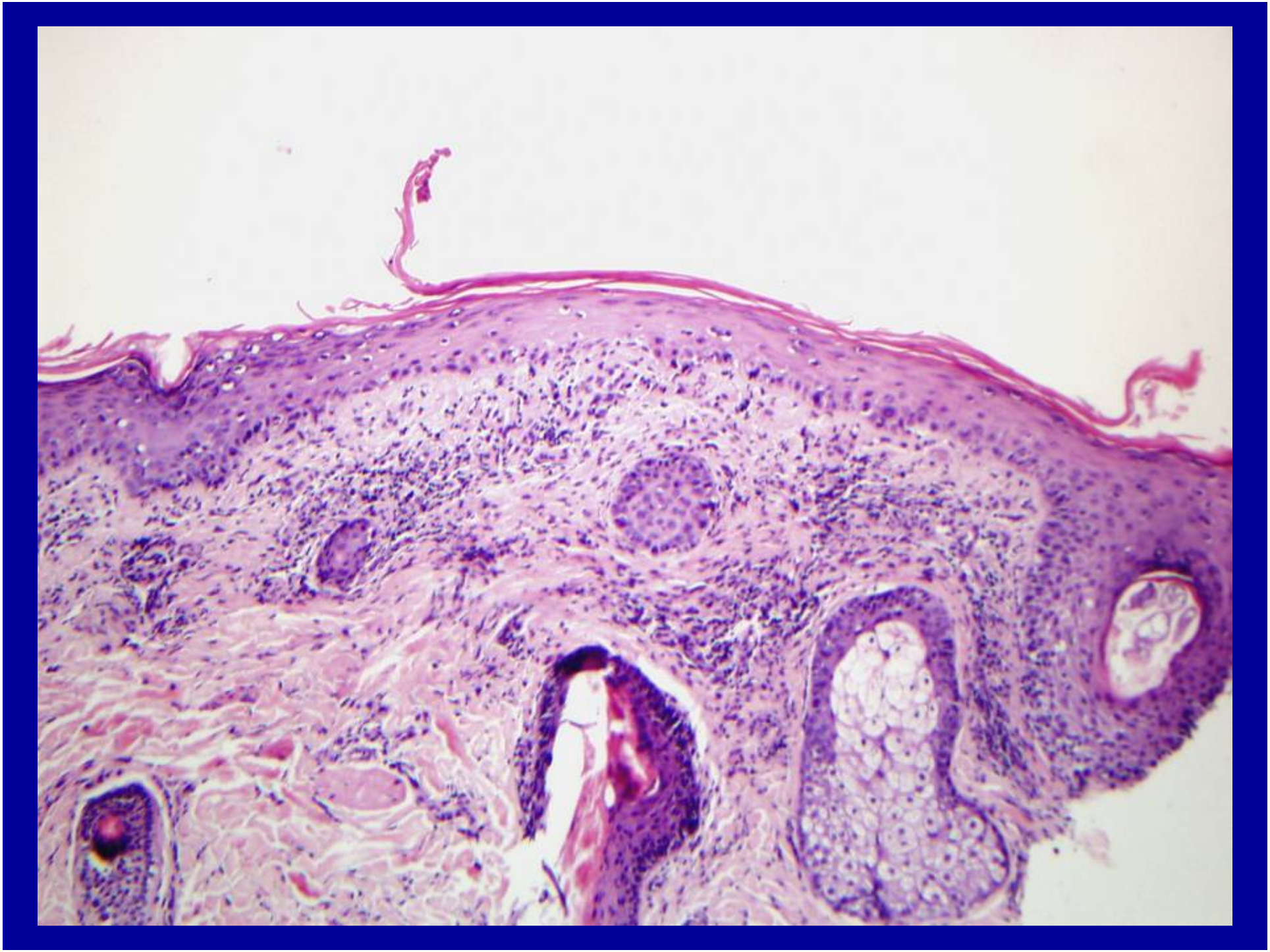

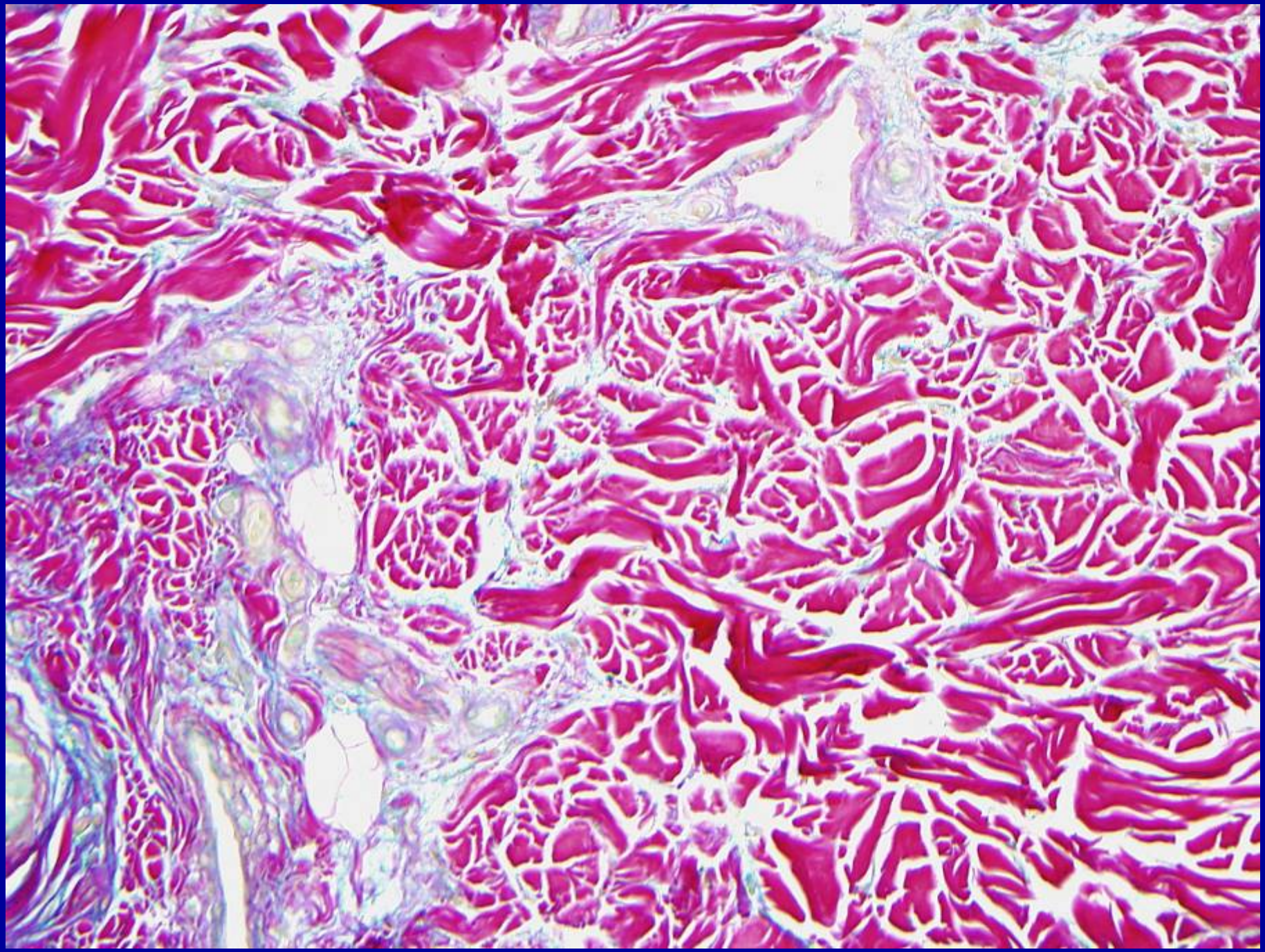

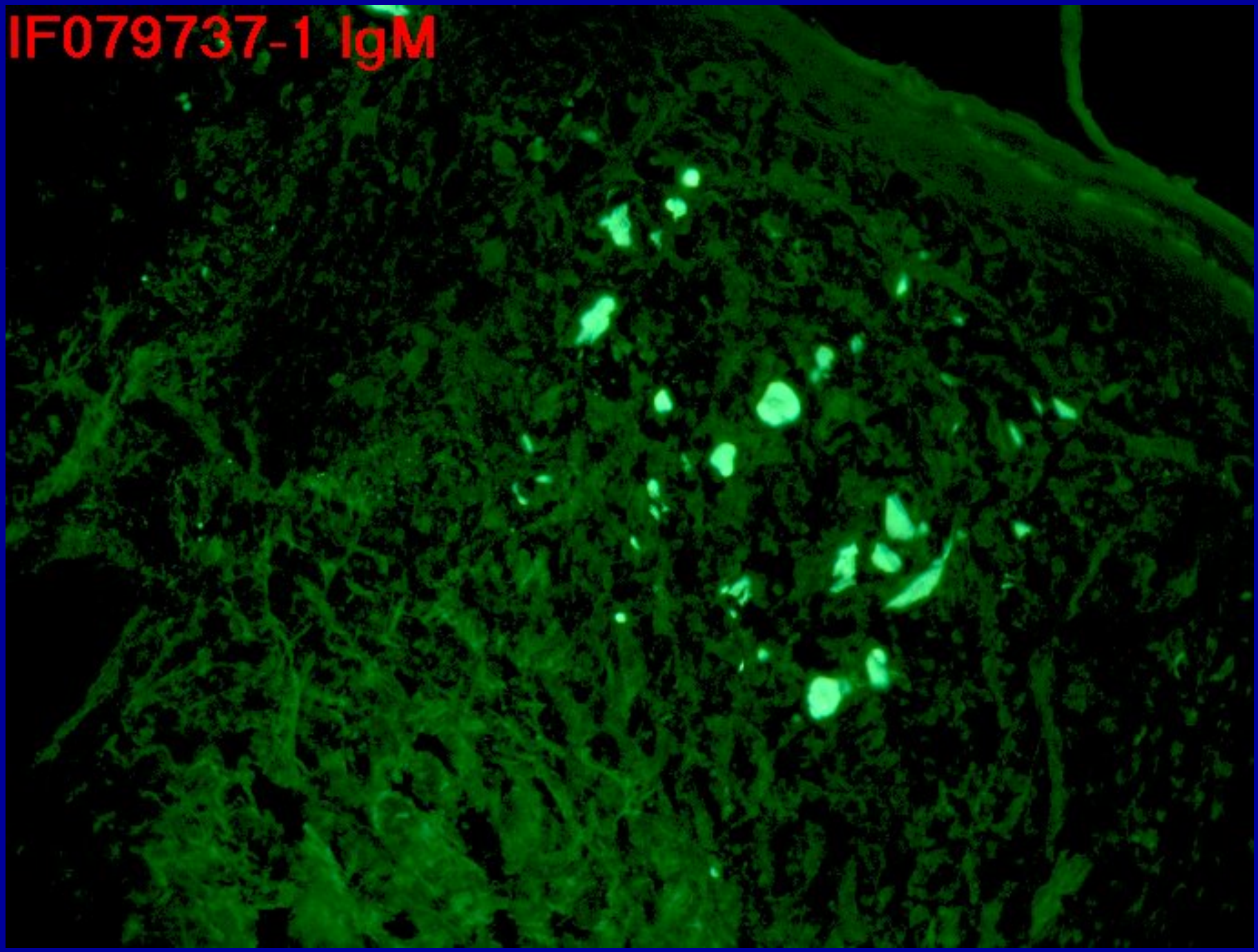
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Case #16

AJ

Aquagenic Syringeal Acrokeratoderma

CASE # 16
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16 – 18, 2009

Patient:	AJ
Area of Interest:	Bilateral palms and palmar fingers
History:	16 year old white female with a two year history of white plaques on the palms that seem to be exacerbated by moisture. When the patient was hospitalized for an unrelated problem and her hands were kept dry, the plaques almost completely resolved. They first appeared when she was being evaluated for a chronic cough that was later diagnosed as a neurogenic cough.
Past Medical History:	GERD, status post Nisson fundoplasty, neurogenic cough
Social History:	High school student. Color guard in marching band and wears gloves often during practice and performances.
Family History:	Father and brothers with sweaty palms. No history of cystic fibrosis, leukemia, keratoderma.
Physical Exam:	Velvety, slightly verrucous white papules coalescing into plaques covering most of the bilateral palms and extending down the palmar surface of all of her fingers bilaterally. Moist palms. No involvement of the dorsal hands or nails.
Pathology:	Thickened orthokeratotic stratum corneum. Normal epidermis with no hypergranulosis, spongiosis, or acanthosis. Dermis had normal collagen pattern and vascular structures with occasional sweat ducts and coils. No inflammatory infiltrate.
Laboratory:	Negative cystic fibrosis genotyping studies
Diagnosis:	Aquagenic Syringeal Acrokeratoderma
Treatment and Course:	Recommended keeping hands dry as much as possible and using aluminum chloride topically daily. The plaques improved with these recommendations, but there is still some recurrence when the patient's hands remain moist for long periods of time. She had a negative work-up for cystic fibrosis, and no other associated systemic findings have been detected.
Comment:	Aquagenic Syringeal Acrokeratoderma (also known as acquired aquageneic wrinkling of the palms, transient reactive papulotranslucent acrokeratoderma, or aquagenic palmoplantar keratoderma) manifests as whitish papules

and plaques on the palms that are accentuated after water immersion and lessens after drying. It can occur only two to ten minutes after water exposure, leading to the "hand-in-bucket-sign" associated with this disease. Often a central prominent pore is noted within each white papule. Pain, pruritus, and burning can be noted in the area. It mostly affects the palms, but the soles an also be involved. It is most commonly seen in adolescents and young adults, mostly females. The cause is unknown, but it may be inherited in an autosomal dominant fashion. Associated findings in case reports include hyperhydrosis, cystic fibrosis, asthma, and allergic rhinitis. Treatment options mainly focus on the associated hyperhydrosis, making aluminum chloride the treatment of choice. Iontophoresis and botulinum toxin have also been effective.

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Case #17

BG

Giant Fibrous Hamartoma
of Infancy

CASE # 17
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: BG

Area of Interest: Abdomen

History: A three month old Caucasian male presented with an abdominal tumor since birth which is growing and changing color. A hemangioma was suspected, but an ultrasound showed a large anterior abdominal not consistent with hemangioma. The patient's development and growth was normal, and he had a normal male karyotype.

Physical Examination: Skin-colored firm tumor on left upper abdominal quadrant that extended to left flank with a deeper component palpable. There were purpuric areas, suggestive of hemorrhage. A supraumbilical, synchronous tumor was also present. No apparent musculoskeletal abnormalities were discovered.

MRI showed a large left flank abdominal wall mass (6.2 X 2.1 X 6.6 cm), with the tumor border appearing indistinct and infiltrating into the abdominal wall musculature, fascia, overlying subcuticular fat and skin. Three separate foci were revealed within the abdominal wall which looked to be anatomically distinct lesions. No anomalies were identified within the abdominal wall, liver, biliary system, spleen, pancreas, gastrointestinal tract, urogenital tract or abdominal vessels.

Pathology: Histopathologic findings included a flattened rete ridge pattern with primitive follicular germ and sebaceous epithelia. The underlying dermis was altered by the presence of thinner collagen bundles and an increased vasculature compared to normal. A prominent dilated duct was present with intraluminal decapitation secretion suggestive of apocrine differentiation. There were monomorphous spindle cells in a fascicular arrangement, extending down from the reticular dermis into the subcutaneous fat. Some of these cells exhibited a "plump" appearance, suggestive of myofibroblastic differentiation. An organoid pattern of fibrous septae dividing primitive spindle cells and mature adipocytes was observed.

Diagnosis: Giant Fibrous Hamartoma of Infancy (FHI)

Treatment and Course: Patient was referred to pediatric surgery for an extensive resection involving much of the left upper internal and

external oblique and rectus abdominus muscles. A large abdominal wall defect was primarily closed using an acellular dermal matrix, peritoneum, and subcutaneous tissue. The patient recovered uneventfully without complication. No recurrence has been noted to date and the patient is developing normally.

Comment:

This is an unusual case of FHI because of its large size and presence of a synchronous lesion. FHI is a benign mesenchymal tumor of myofibroblastic derivation with prototypical histopathologic features. Most FHI tumors are solitary, range in size from 0.5 – 4.0 cm, and arise within the first 12 months of life. The most common sites of involvement include the axillae, upper arms, inguinal area and external genitalia. Though characteristically slow-growing and asymptomatic, FHI may exhibit rapid growth or symptoms of pain, warmth, or tenderness. Most tumors are freely moveable; however, some reports of infiltrative, sclerotic borders have been described, which can limit mobility. This correlates well with the poor circumscription and infiltrative margins seen histologically and propensity for clinical recurrence. Recurrence occurs in approximately 10% of cases, usually within 2 to 11 months. Risk factors for recurrence included male gender and axillary or upper extremity involvement. FHI does not spontaneously regress, and surgical treatment is usually curative. FHI exhibits no metastatic potential.

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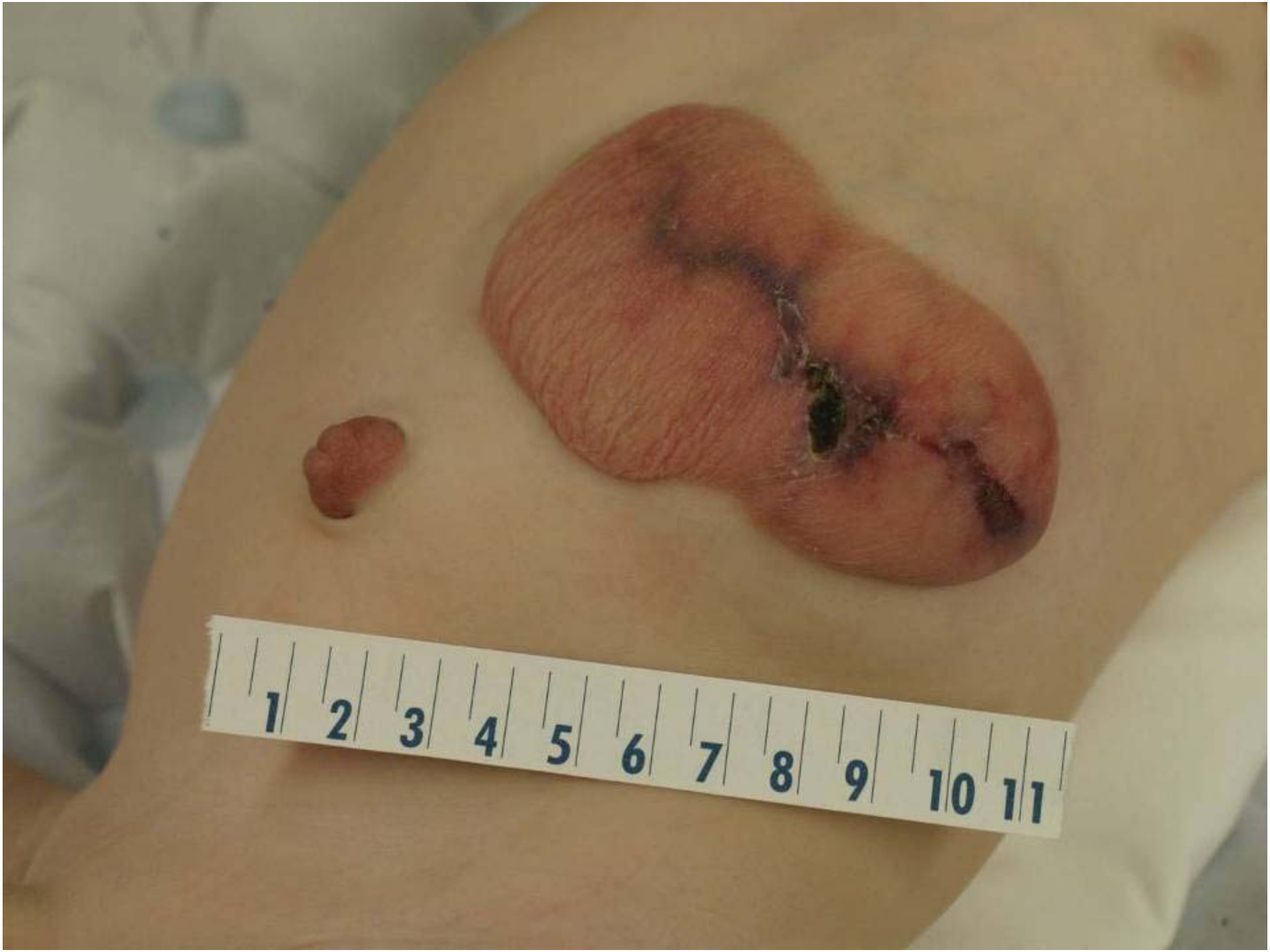
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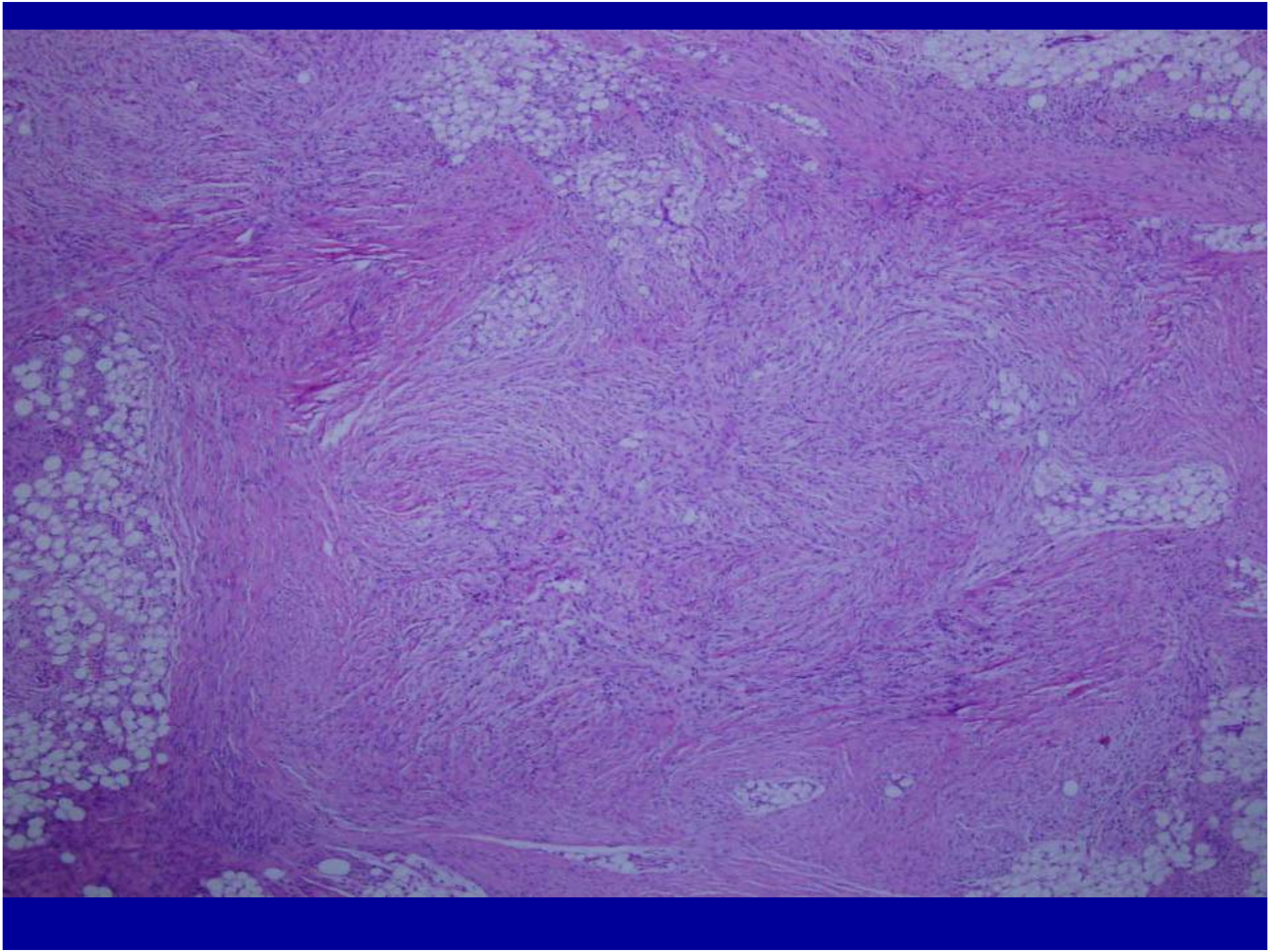

Case #18

DE

Follicular Mucinosis
(Alopecia Mucinosa)

CASE # 18
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: DE

Area of Interest: Lower face

History: A 57 year old white male presented with a 3 year history of rash on the lower face and concomitant rosacea which was treated with metronidazole cream and minocycline. The rash on his lower face would not clear completely with the rosacea treatment. This area became more pruritic and secondarily infected with *Staphylococcus aureus*. Herpes viral cultures were negative. Several courses of antibiotics were given which would provide temporary relief, but the lesions would recur and he started getting similar lesions on the other side of his face.

Past Medical History: hypertension, rosacea

Physical Examination: Bilateral jawline, chin, and cheeks multiple boggy, erythematous papules and plaques with crust and scale. No evidence of discrete alopecia in the plaques. No cervical lymphadenopathy.

Pathology: In most of the follicles there are large collections of lymphocytes and a few eosinophils involving follicular epithelium and sebaceous glands. A colloidal iron stain revealed follicular mucin. These changes were consistent with follicular mucinosis.

Laboratory: CBC and blood chemistries within normal limits.

Diagnosis: Follicular mucinosis (alopecia mucinosa)

Treatment and Course: A punch biopsy was performed with was consistent with follicular mucinosis. T cell gene rearrangement studies were negative, he had no systemic symptoms, and no other evidence of an associated cutaneous T cell lymphoma (CTCL). The patient was started on isotretinoin and the areas of follicular mucinosis cleared.

Comment: Alopecia mucinosa is hallmarked by the pathologic finding of follicular mucinosis, although many people use these terms interchangeably. It is characterized by hypopigmented or erythematous scaly eczematous plaques and flesh-colored follicular papules. They are usually distributed on the face, and pruritus is a common symptom. Alopecia can occur within the lesions. The disease may be skin-limited (primary) or may be

associated with follicular mycosis fungoides. Spontaneous resolution can occur with primary follicular mucinosis, but treatment was needed in this patient because of the chronic and intermittent nature of the lesions. This case is interesting because of the patient's complete recovery with isotretinoin. Other treatment options include topical or intralesional corticosteroids, dapsone, PUVA, radiation therapy, minocycline, and indomethacin. Those cases associated with CTCL are more refractory to treatment and have a worse prognosis than the classic CTCL.

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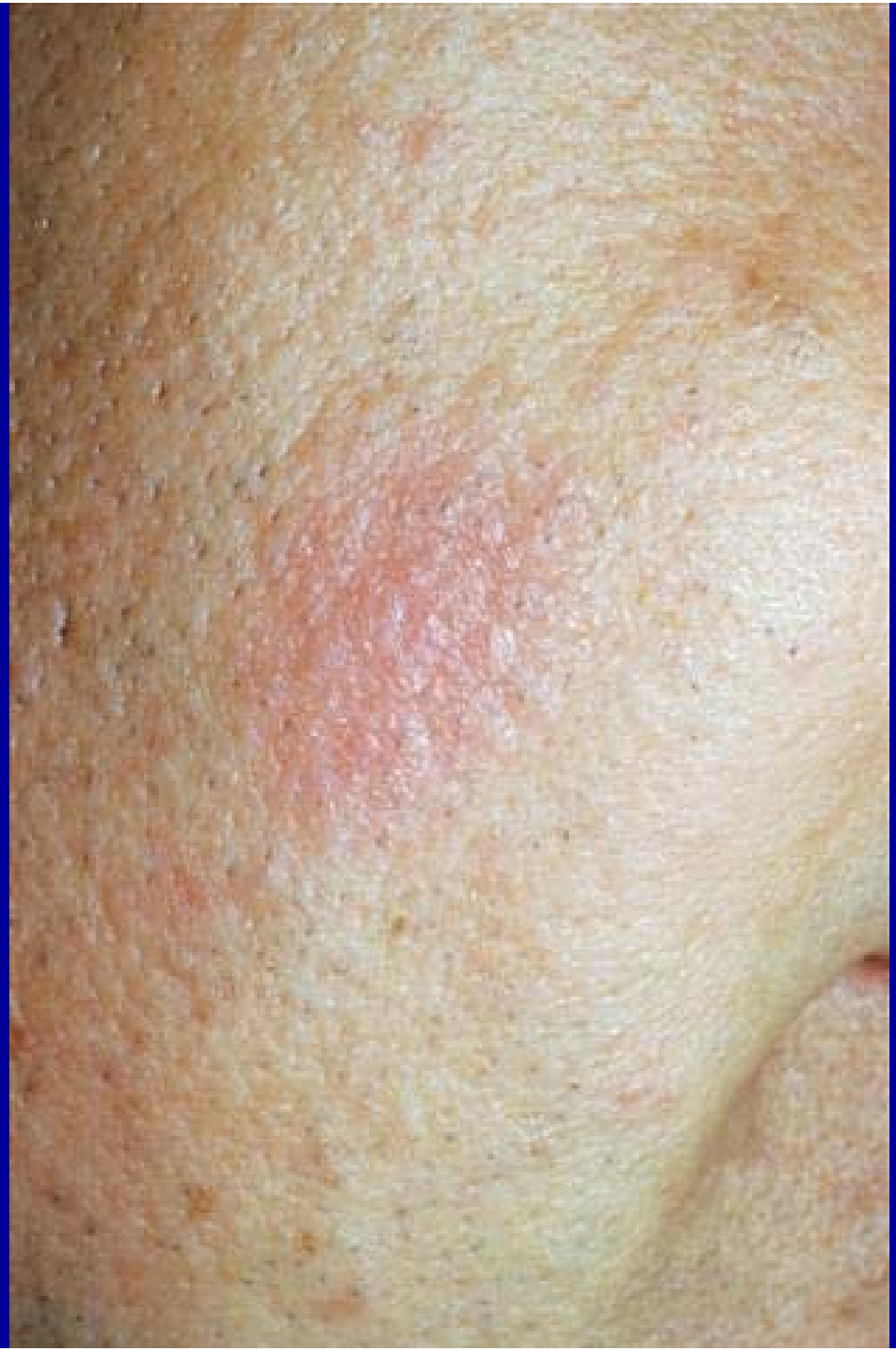
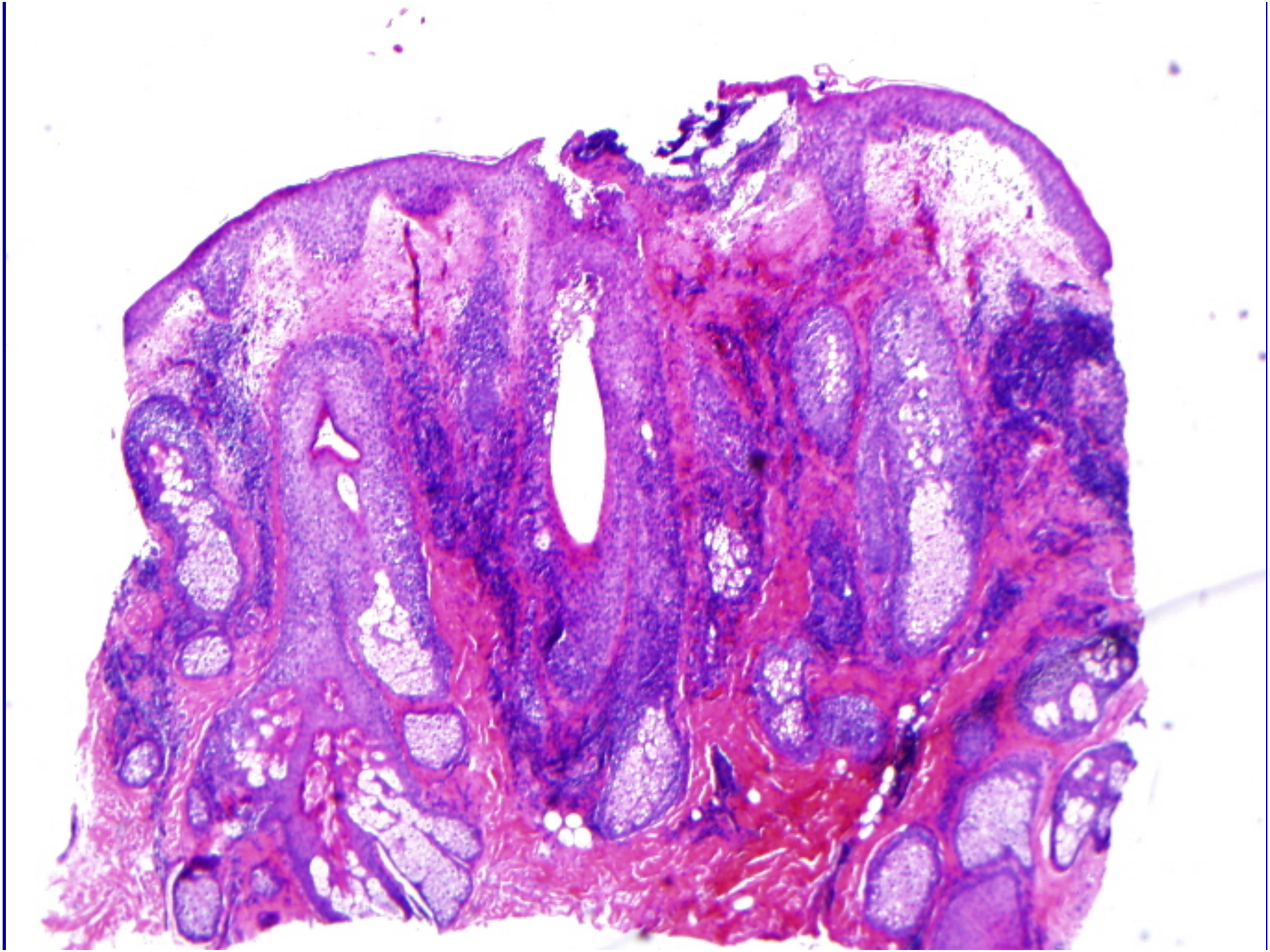
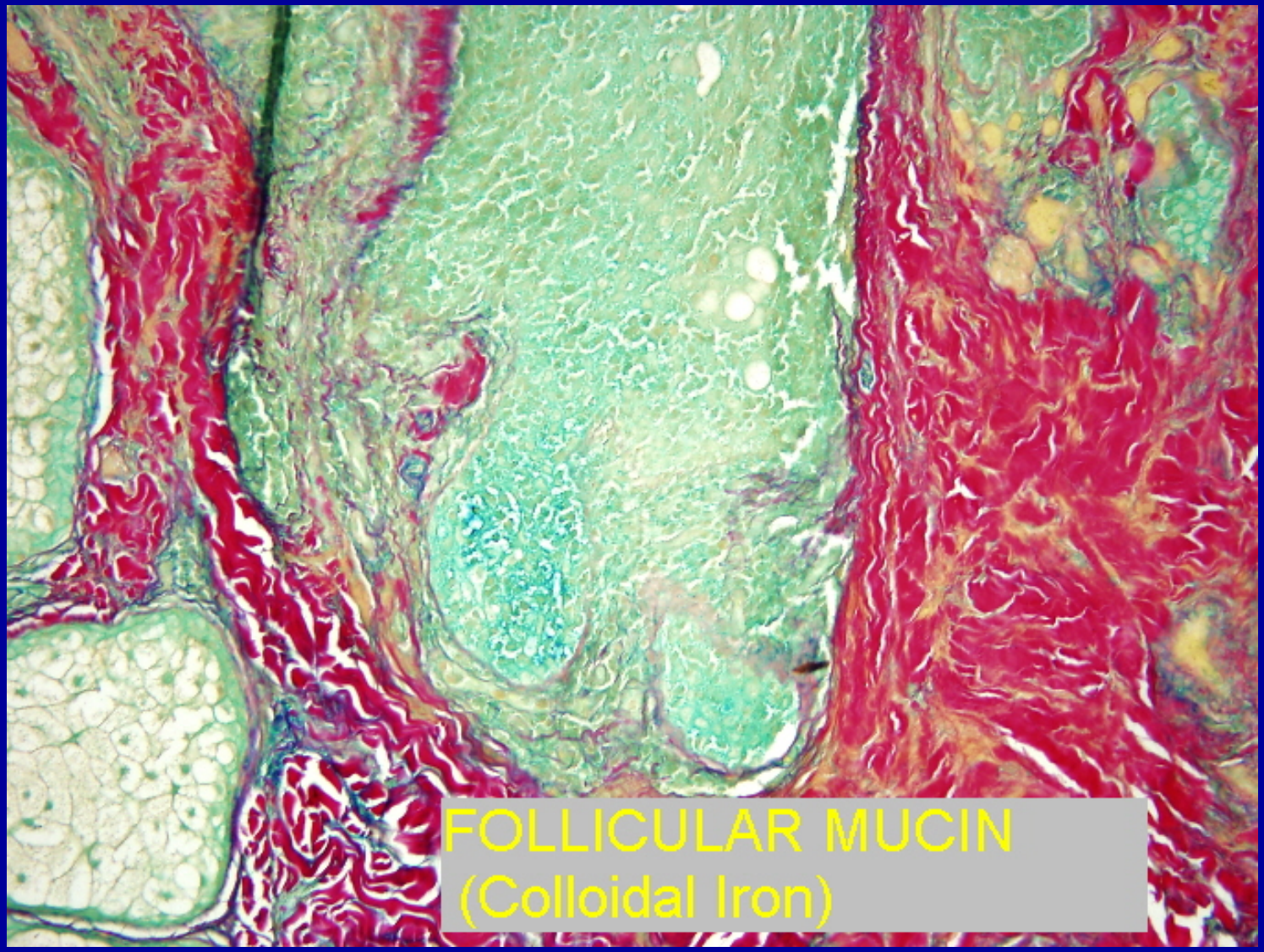
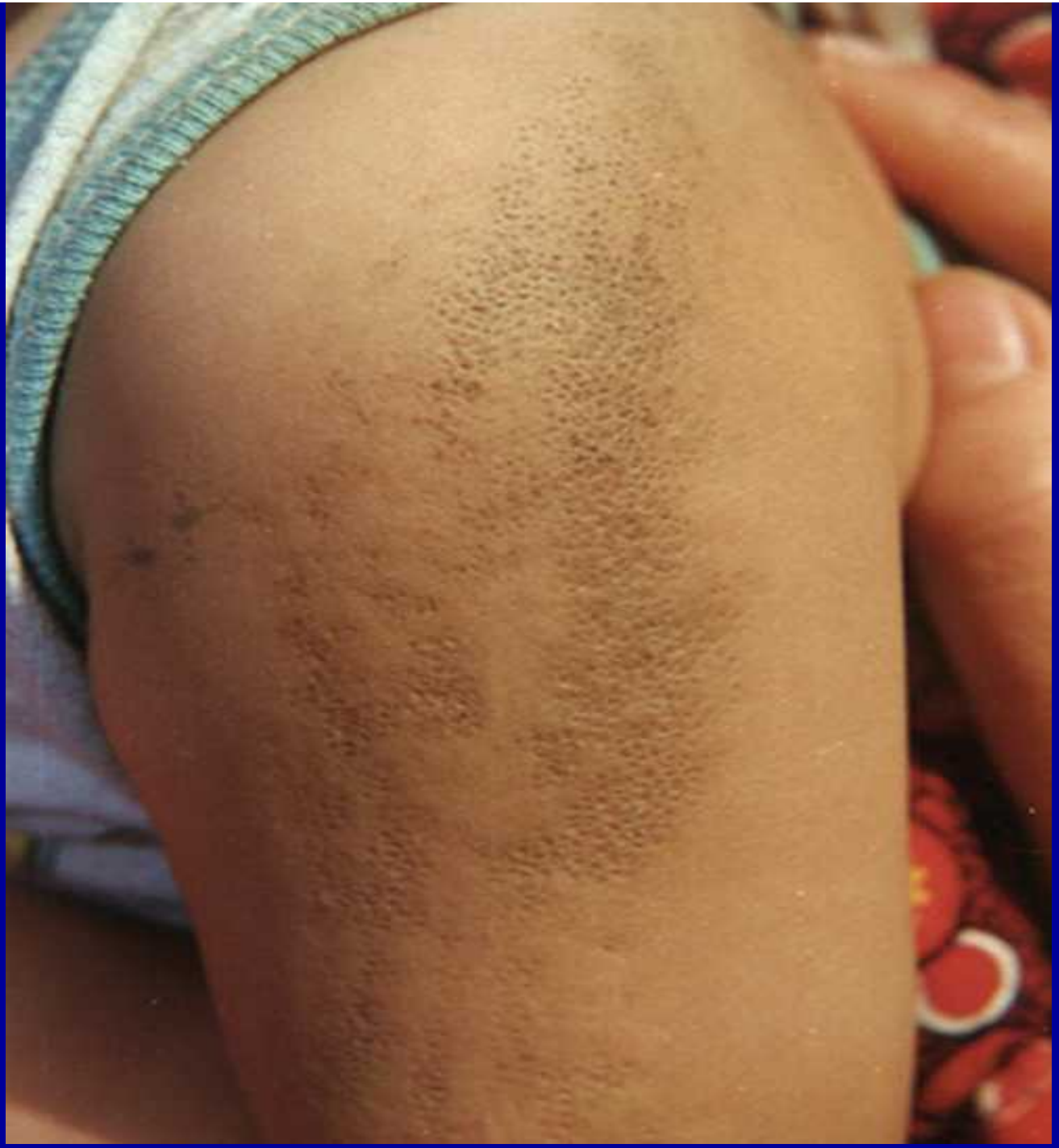
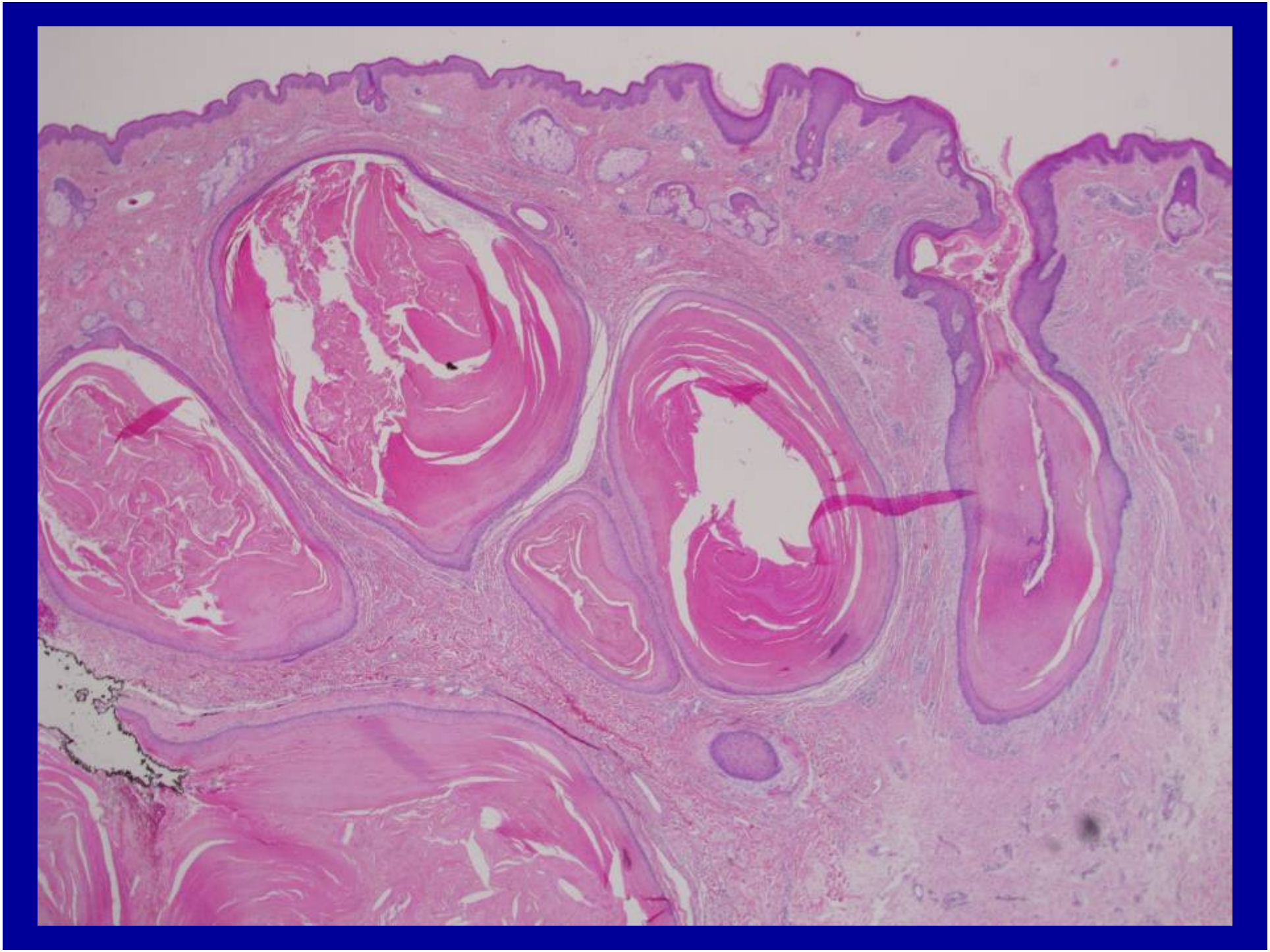
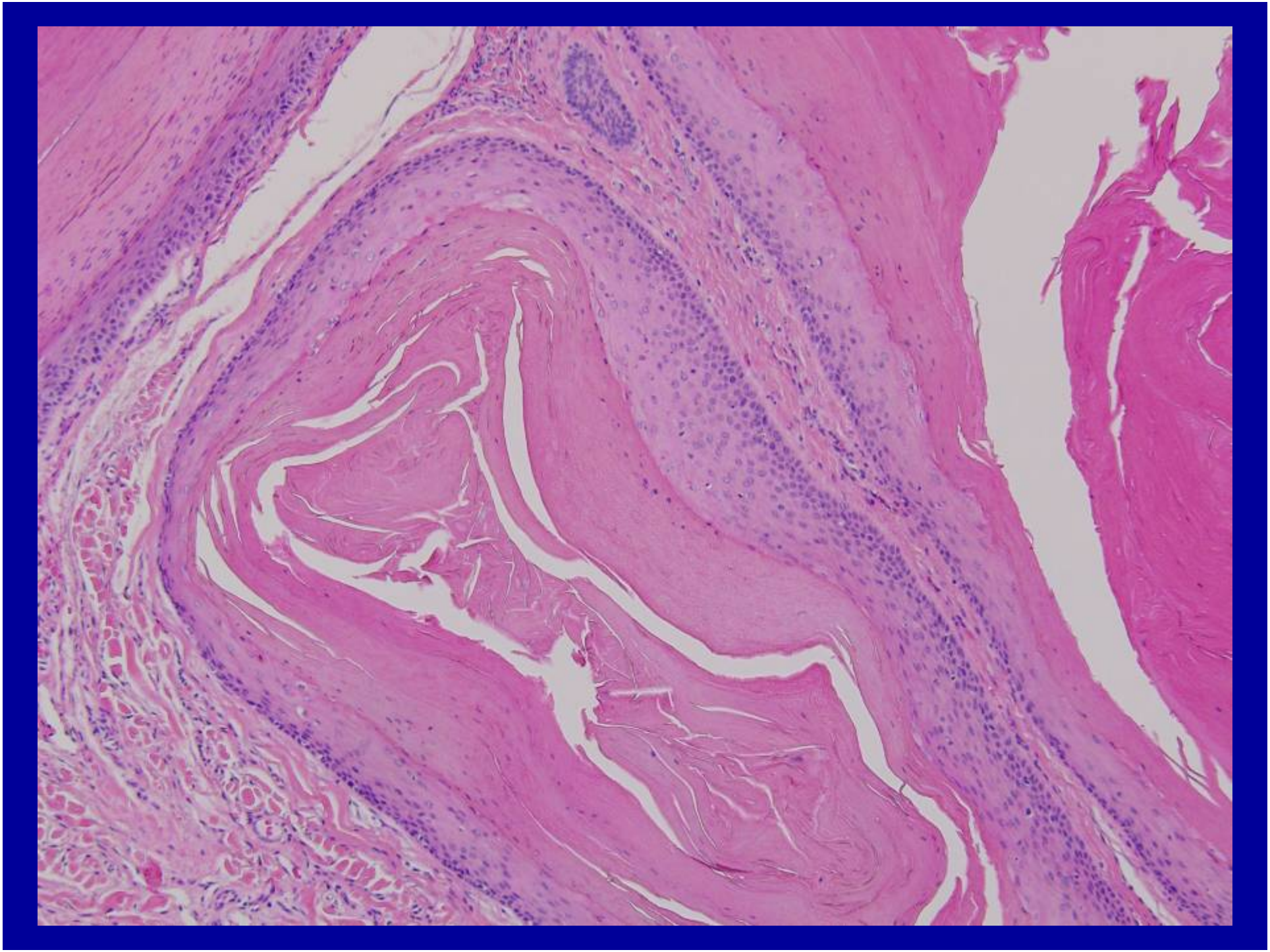





FOLLICULAR MUCIN
(Colloidal Iron)







Case #21

TA

Klippel-Trenaunay-Weber Syndrome

CASE # 21
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: TA

Area of Interest: Right Lower Leg

History: Pt is a 2yo male referred from his PCP for evaluation of right lower leg hypertrophy.

Past Medical History: none

Family History: N/A

Physical Exam: His general habitus is notable for significant lower limb asymmetry of the bone and soft tissues. The right lower limb shows significant size enlargement over the left. This is particularly noted on the dorsal right foot. There is a widespread nevus flammeus occupying the foot, leg, thigh, and buttock. There are small phlebectasias of the medial thigh.

Pathology: N/A

Diagnosis: Klippel-Trenaunay-Weber Syndrome

Treatment and Course: Pt has been followed by both Dermatology Surgery and Pediatric Orthopedics at MUSC. Long bone xray shows bone asymmetry with right leg being 1.2 cm longer than the left leg however pt is ambulating well. MRA/MRV shows no deep vascular anomaly. No need for intervention at this time.

Comment: Klippel-Trenaunay syndrome is characterized by a triad of port-wine stain, varicose veins, and bony and soft tissue hypertrophy involving an extremity.

The exact cause of Klippel-Trenaunay-Weber syndrome (KTWS) remains to be elucidated. Most cases are sporadic, although a few cases in the literature report an autosomal dominant pattern of inheritance.

KTWS generally affects a single extremity, although cases of multiple affected limbs have been reported. The leg is the most common site followed by the arms, the trunk, and rarely the head and the neck.

Arteriovenous fistulas, the feature that distinguishes Klippel-Trenaunay syndrome from Parkes-Weber syndrome, are rarely found in the affected extremity. If present, they can occasionally be palpated as a pulsatile mass, thrill, or bruit on physical examination.

Treatment of KTWS is conservative and symptomatic.

Compression garments are indicated for chronic venous insufficiency, lymphedema, recurrent cellulitis, and recurrent bleeding from capillary or venous malformations of the extremity.

Regarding limb hypertrophy, heel inserts are generally sufficient for limb discrepancies of 1.5 cm or less. For greater discrepancies, orthopedic surgery may be considered.

Laser treatment of the hemangioma can be effective in lightening the color of the port-wine stain. Currently, the flashlamp-pumped pulsed dye laser is the treatment of choice in vascular lesions. Laser treatment is also indicated in the case of ulceration. Ulceration of hemangiomas can be painful and can impair functional abilities.

Stable disease can be followed clinically. KTWS is not always a static disease process. If progression of the disease arises, imaging studies should be performed. Medical or surgical intervention should be pursued if indicated.

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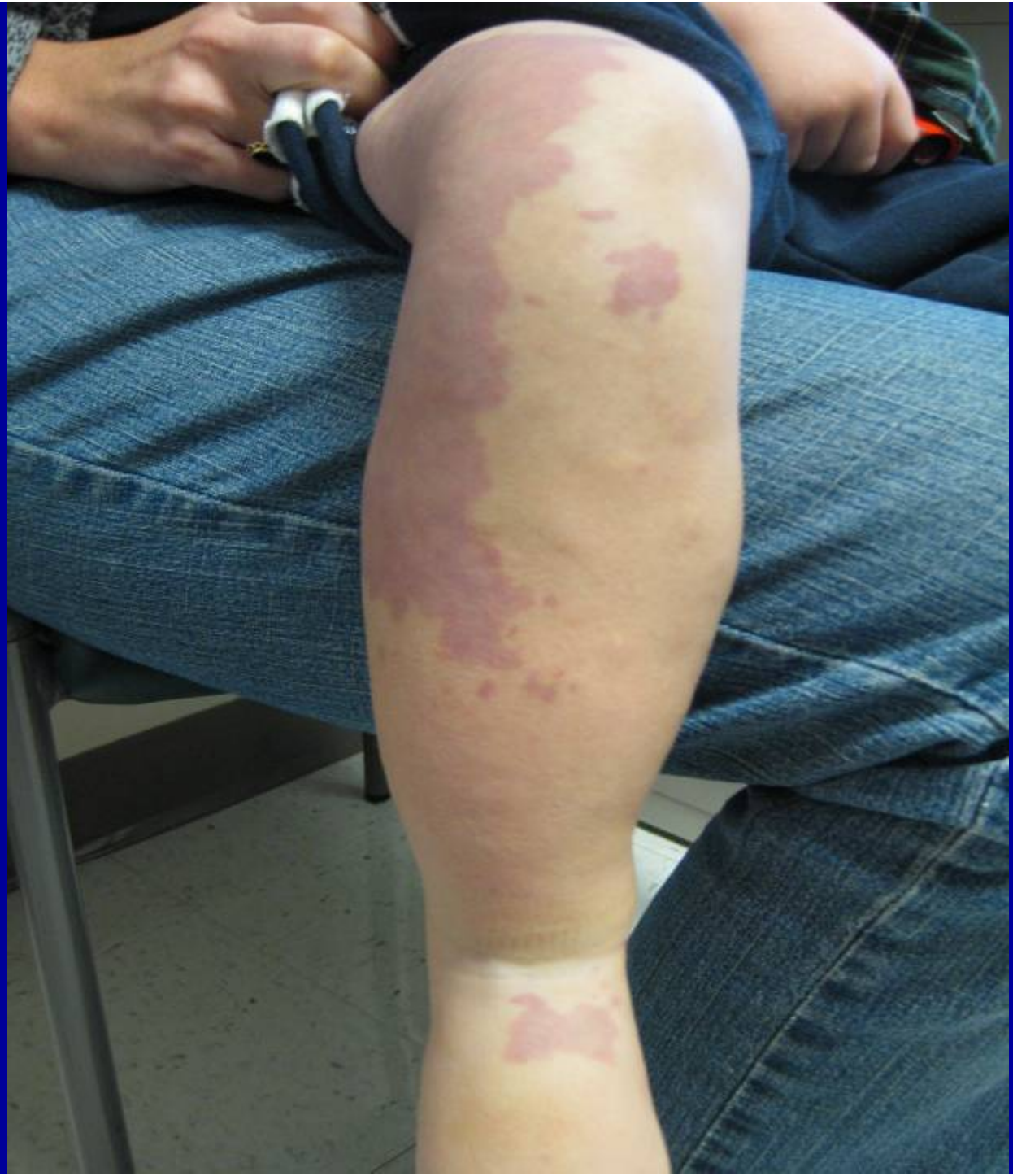

Case #23

JS

Keratosis Follicularis
(Darier's Disease)

CASE # 23
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	JS
Area of interest:	Scalp, Neck , Trunk, Legs, Nails
History:	The patient is a 43 year old male with a long history of skin lesions and intermittent flares of malodorous lesions.
PMHx:	Bipolar disorder
Family History:	Multiple family members with similar skin findings
PE:	Yellow-brown, greasy, hyperkeratotic papules and plaques on the face, neck, trunk, and extremities. Nails show longitudinal alternating red and white bands and distal v-shaped nicking.
Pathology:	Marked hyperkeratosis, papillomatosis, acanthosis, acantholytic dyskeratotic keratinocytes forming "corps rounds" and "grains"
Laboratory:	N/A
Diagnosis:	Darier's Disease (Keratosis Follicularis)
Treatment and Course:	Treated over the years topically with steroids, retinoids, and silvadene. He has also been admitted to the hospital multiple times due to secondary infection with MRSA for which he has been treated with multiple antibiotics. The patient has taken Lithium for a long time which is known to cause flares but he has difficulty coming off of it. The patient has done very well with acitretin 25mg but this has been difficult to acquire at times secondary to finances. At other times, he has been treated with 100,000 units of vitamin A with some improvement.
Comments:	Darier's disease is an autosomal dominant genodermatosis with characteristic hyperkeratotic greasy papules that tend to be in a seborrheic distribution. The disease usually becomes apparent during puberty and leads to formation of papules which become confluent into malodorous plaques. There are also common nail changes including red and white longitudinal bands, nail ridges, and

v-shaped nicking. It has been associated with psychiatric problems and secondary infections.

The disease occurs secondary to mutations to the ATP2A2 gene which codes for the SERCA2 protein. This leads to faulty intracellular calcium signaling which affects cellular adhesion. More than 113 familial and sporadic mutations have been identified.

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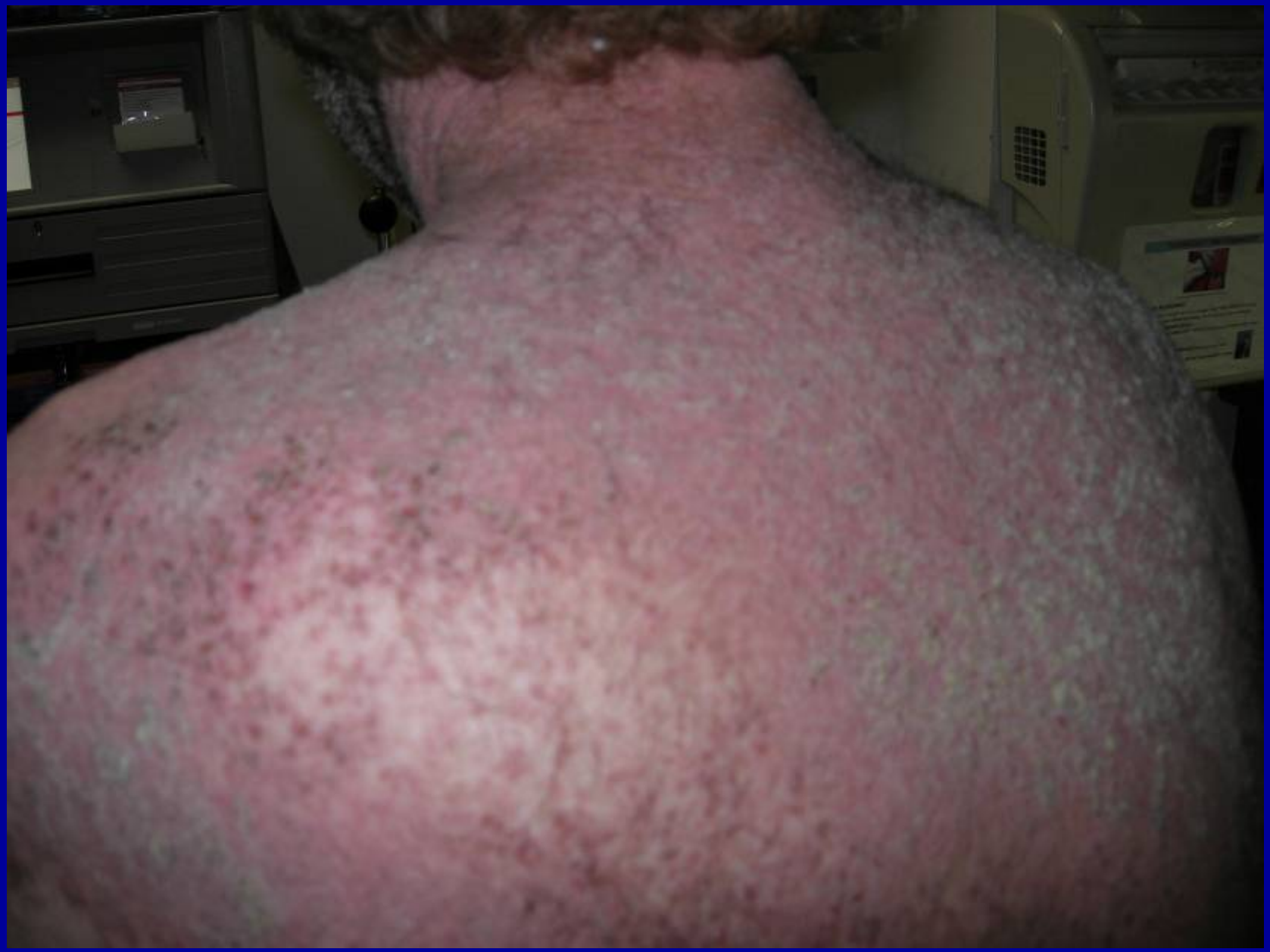

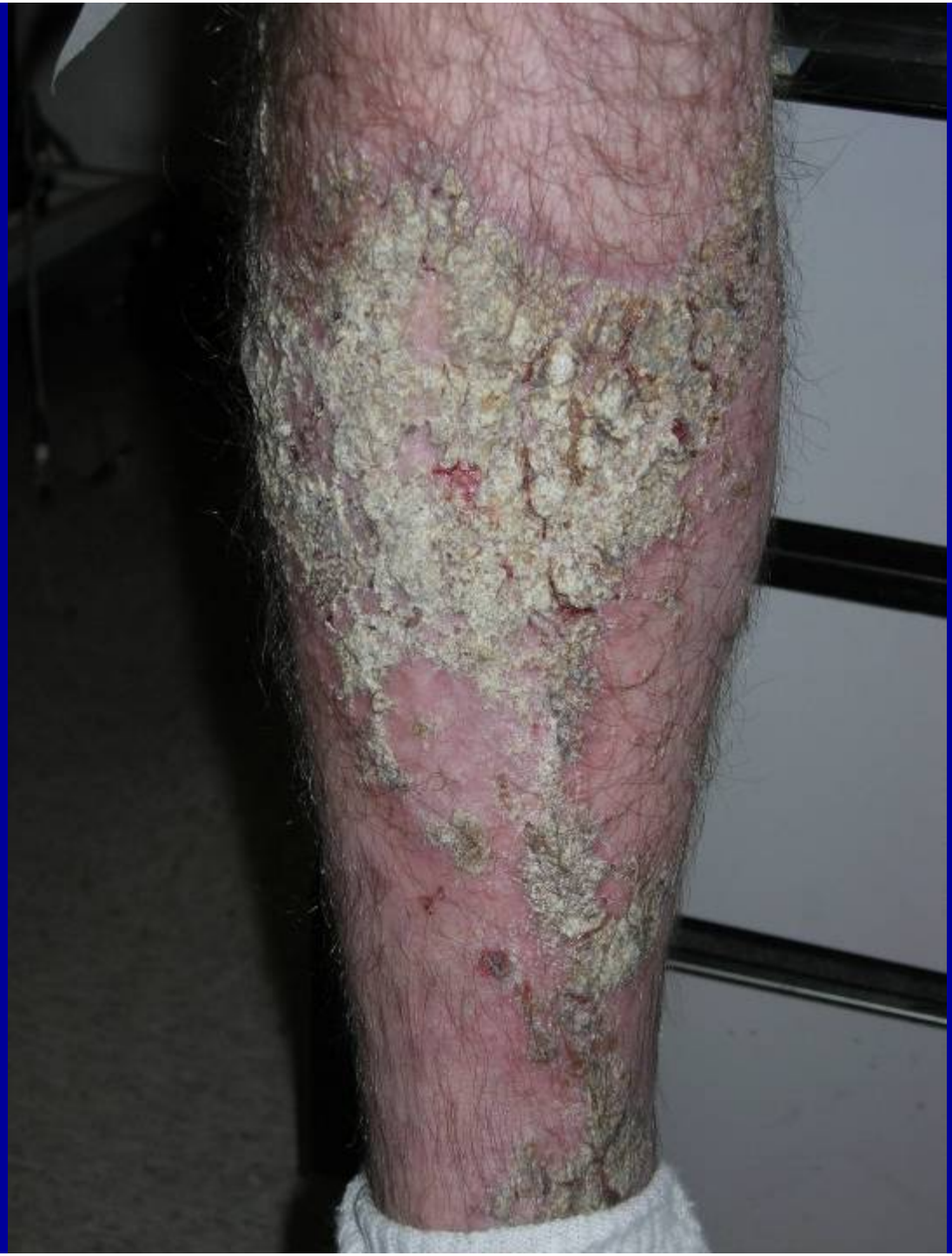

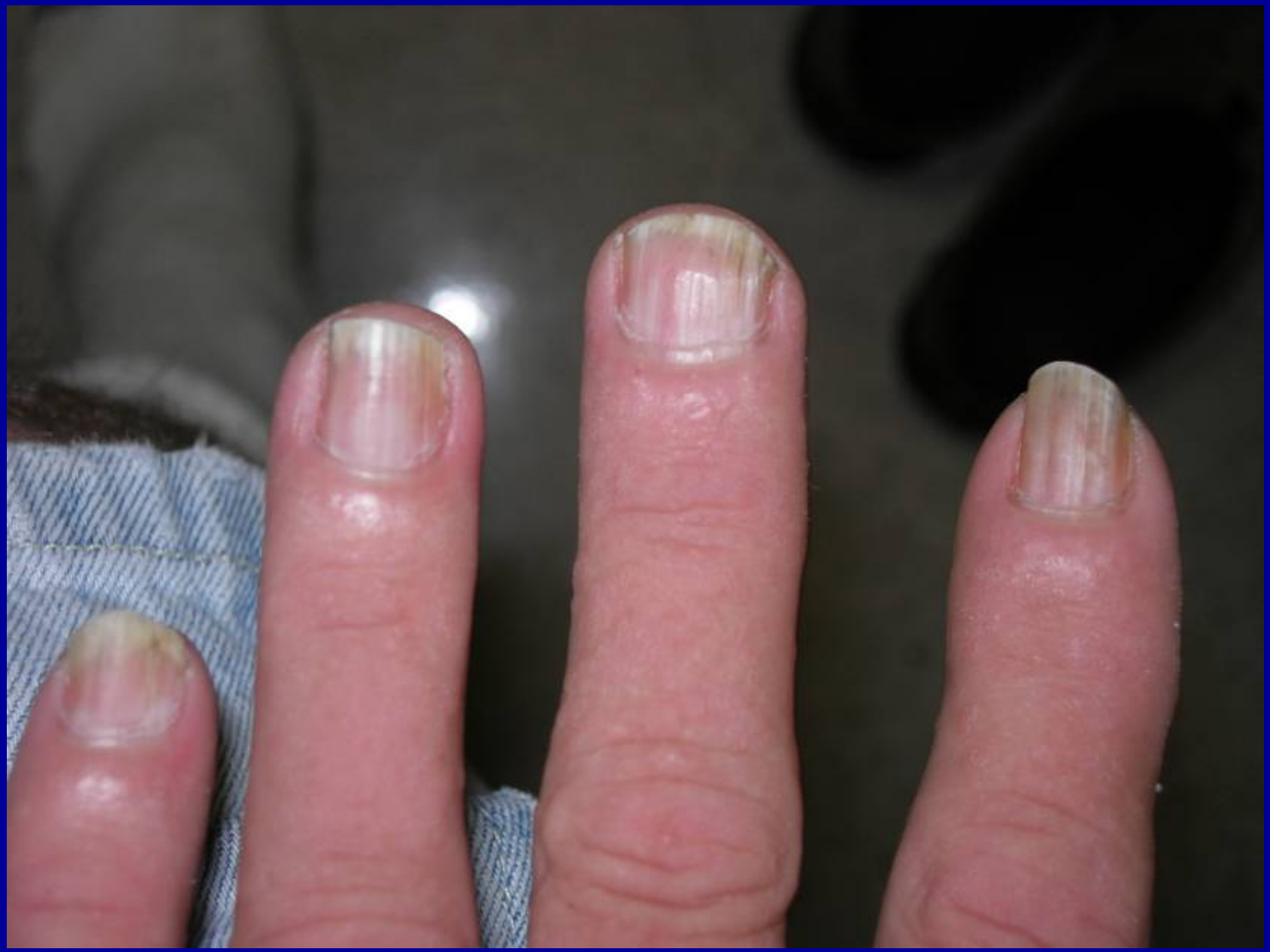

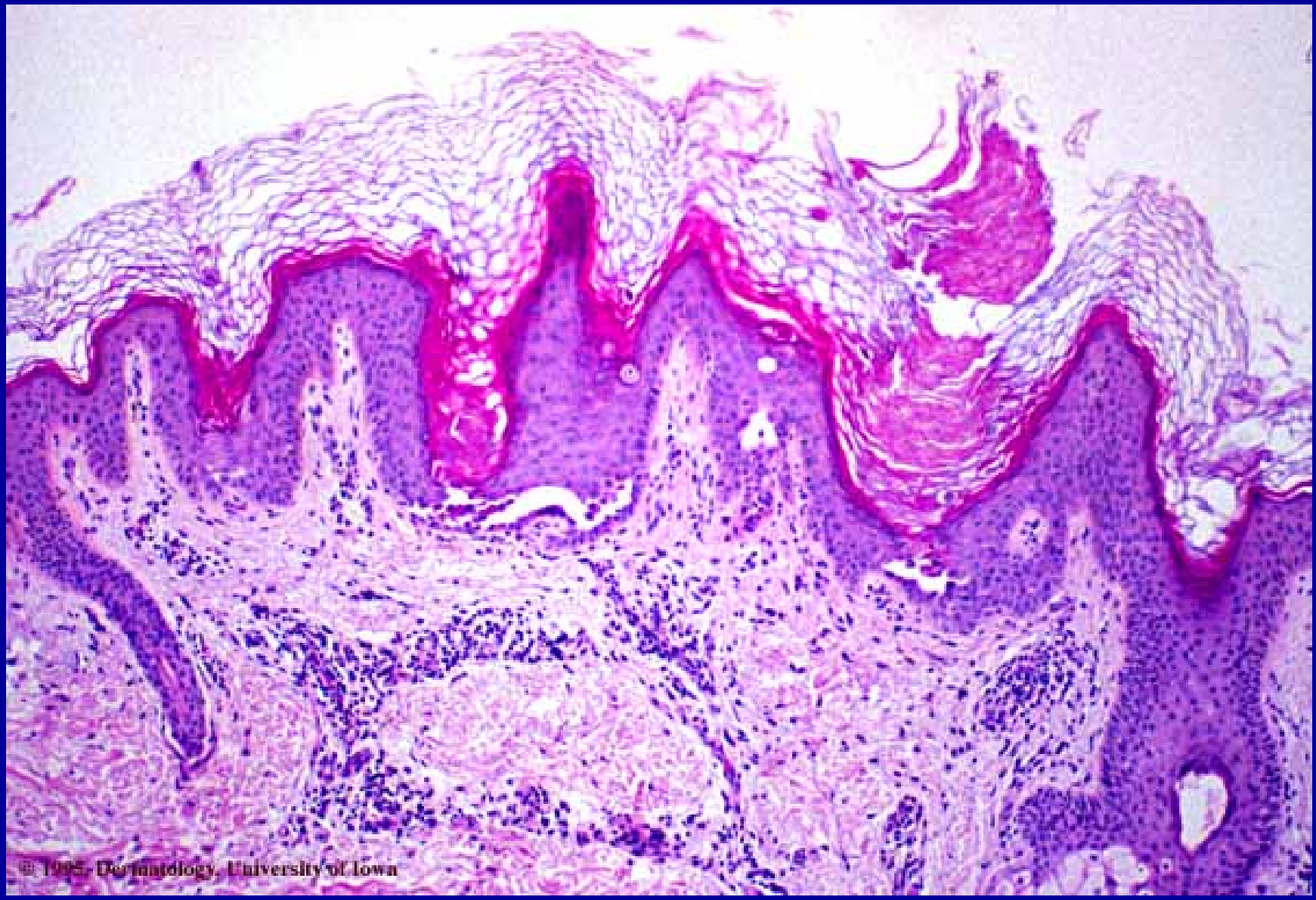
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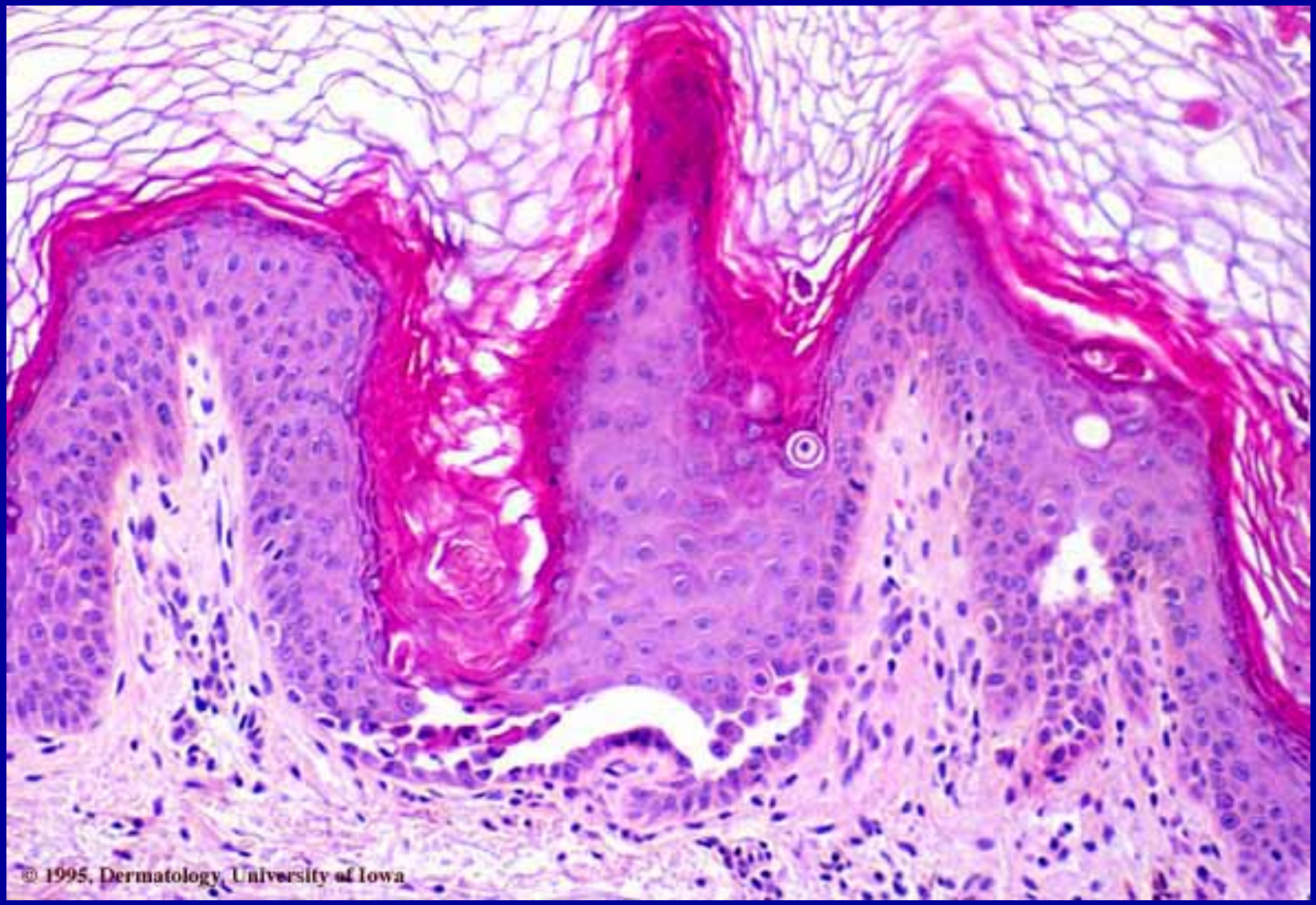
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Case #24

JS

Hurler’s Syndrome

CASE # 24
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	JS
Area of interest:	Scalp, face, neck
History:	5 year old WM with a history of bone marrow transplant was seen by dermatology to evaluate a rash to rule out GVHD.
PMHx:	Mitral valve thickening and prolapse, aortic valve and ascending aortic root dilatation, pericardial effusion, developmental delay, gastroesophageal reflux, bone marrow transplant, and pulmonary hemorrhage. Past surgical history include bilateral inguinal hernia repair, umbilical hernia repair, pyloric myotomy for pyloric stenosis, cholecystectomy, and PEG placement as well as port placement and then removal. Family history is noncontributory. Birth weight of 10lb 2 oz.
PE:	Well nourished WM in NAD, Exam is notable for hypertrichosis of the entire body including trunk, arms and legs. He also has trichomegaly. He has scoliosis and is wearing a back brace. He has mildly coarse facial features. At the time of presentation he had erythema and greasy scale in the scalp, face, ears, as well as on the back and on his lower extremities.
Pathology:	The cornified layers show focal parakeratosis and serum crust. The vital edpidermis displayed foci of spongiosis and exocytosis of lymphocytes. There were no signs of GVHD.
Laboratory:	N/A
Diagnosis:	Hurler's syndrome (Mucopolysaccharidoses 1)
Treatment and Course:	A punch biopsy was performed and was consistent with seborrheic dermatitis. He was treated successfully with dermasmooth scalp oil. There were no signs of GVHD. His large extent of seborrhea was thought to be due to his hypertrichosis.
Comments:	Mucopolysaccharidosis type I is a rare, autosomal recessive, lysosomal storage disorder. Hurler syndrome is caused by mutation in the gene (*IDUA*) that encodes alpha-L-iduronidase on chromosome 4 which results in an

inability of the lysosome to break down glycosaminoglycans, namely dermatan sulfate (DS) and heparan sulfate (HS). These accumulate in the lysosomes, ultimately causing cell, tissue, and organ dysfunction by largely unknown pathophysiological mechanisms.

The disease has spectrum of severity. Children with Hurler syndrome appear normal at birth and develop the characteristic appearance over the first years of life. Symptoms include facial dysmorphism, corneal clouding, hepatomegaly, valvular heart disease, obstructive airway disease, developmental delay, skeletal deformities, and joint stiffness. There is a spectrum of disease with some having early onset and much shortened lifespan. Average life span is 10 years.

There have been studies that have shown improvement with bone marrow ~~biopsy~~ transplant at an early age that helps decrease many of the symptoms including retardation. Some patients are given the medication Laronidase which is a polymorphic variant of the human enzyme a-L-iduronidase produced by recombinant DNA technology.

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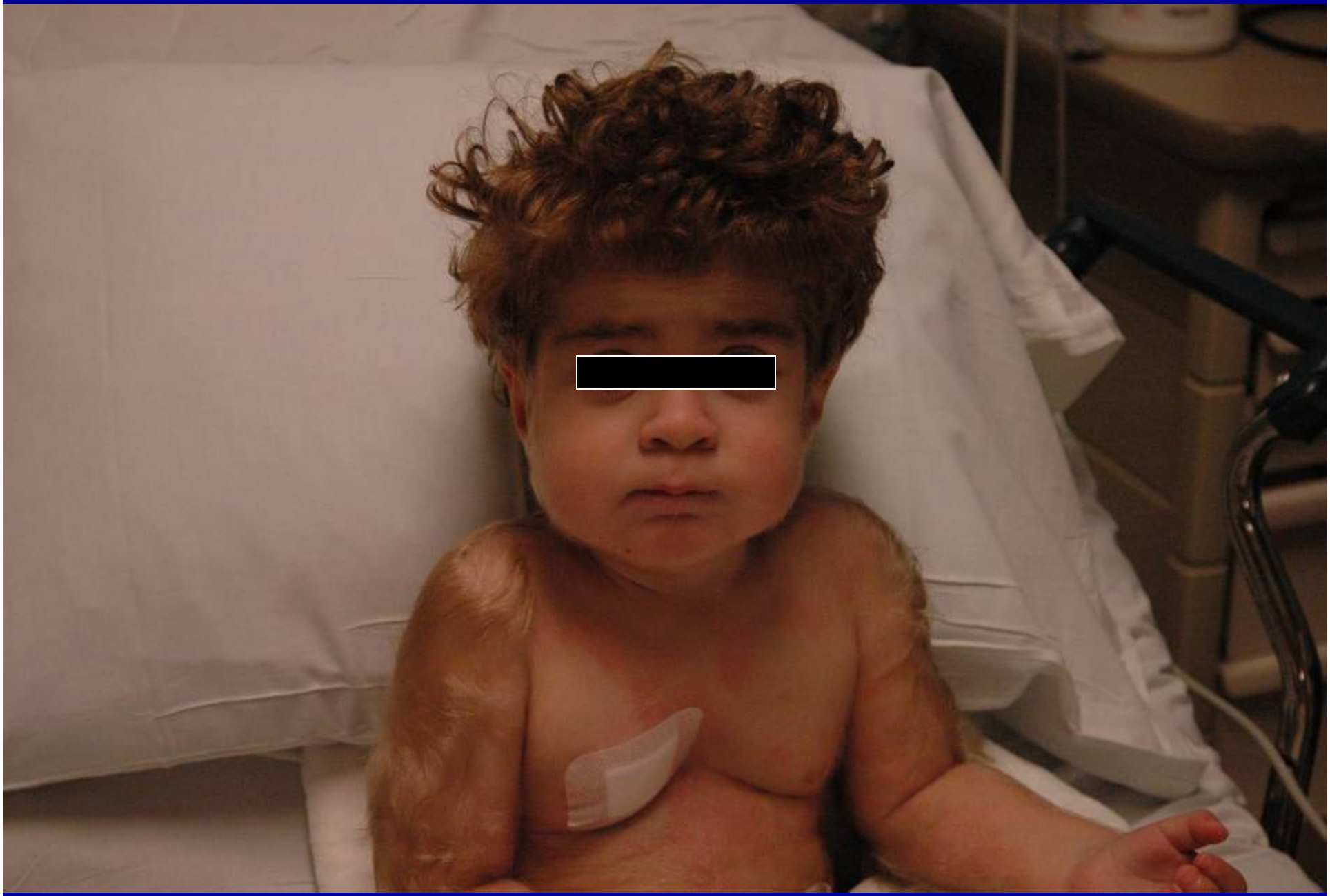

Case #25

LS

Nodular Localized Cutaneous Amyloidosis

CASE # 25
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	LS
Area of interest:	Scalp
History:	Pt is a 63 yo WM with no past medical history of skin cancers or other skin lesions who presents with a 9 month history of a tan-colored plaque on his forehead.
PE:	The patient is well-nourished, well-developed, pleasant, alert and oriented x 3. On exam his hair, scalp, chest, back, abdomen, arms, hands and nails were normal. Remarkable on examination was a yellow-tan plaque about 2 cm in size on his left scalp.
Pathology:	Histologic sections show a a fragmented skin punch biopsy. The cornified layer is normal basket weave orthokeratosis. The vital epidermsis is within normal limits. Within the dermis, there is diffuse deposition of pink amorphous material and associated interstitial hemorrhage. There is a perivascular lymphoplasmacytic infiltrate. Congo red histochemical stain was performed and demonstrates brick red positive staining with apple green birefringence on polarization.
Laboratory:	Normal CBC, Normal CMP, Normal Protein Electrophoresis (Total protein- 5.9, Albumin-4.1 Alpha 1 Globulin-0.2, Alpha-2-Globulin-0.7, Beta Globulin-0.9, Gamma Globulin-0.9).
Diagnosis:	Nodular Localized Cutaneous Amyloidosis
Treatment and Course:	The patient was recently biopsied and diagnosed and will likely undergo Moh's surgery to the site as we have had positive results with this in the past.
Comments:	Nodular Localized Cutaneous Amyloidosis (NLCA) is a rare primary skin condition found in adults due to the deposition of amyloid or "amyloid-like"proteins in the dermis. NLCA is the rarest form of LCA and was first reported by Gottron in 1950. The amyloid is thought to be due to a population of plasma cells. This is in contrast to keratinocyte derived amyloid found in lichinoid or macular amyloidosis. Some have found monoclonality of the plasma cells, suggesting a neolplastic disorder, but others

have found polyclonality. Progression to systemic disease is rare but has been described. Several of the cases in the literature have found an association with Sjögren's Syndrome. Firm nodules can present anywhere on the skin, including the face, scalp, extremities, trunk, and genitalia and are usually pink to brown to red. The nodules range in size from a few millimeters to a few centimeters.

The typical workup when NLCA is considered is to rule out any extracutaneous manifestations of the disease. Normal serum protein electrophoresis and urine protein electrophoresis studies exclude multiple myeloma. Positive antinuclear, anti-Ro, and anti-La antibodies indicate Sjögren syndrome. Laboratory studies, such as CBC, serum chemistry profile, and liver function tests are typically normal.

Multiple treament modalities including topical and intralesional corticosteroids, cryotherapy, dermabrasion, shaving, curettage and electrodesiccation, laser, and surgery have been attempted. Steroids and cryotherapy have not been found to be helpful but there have been good results with surgical removal and electrodesiccation and curettage.

References:

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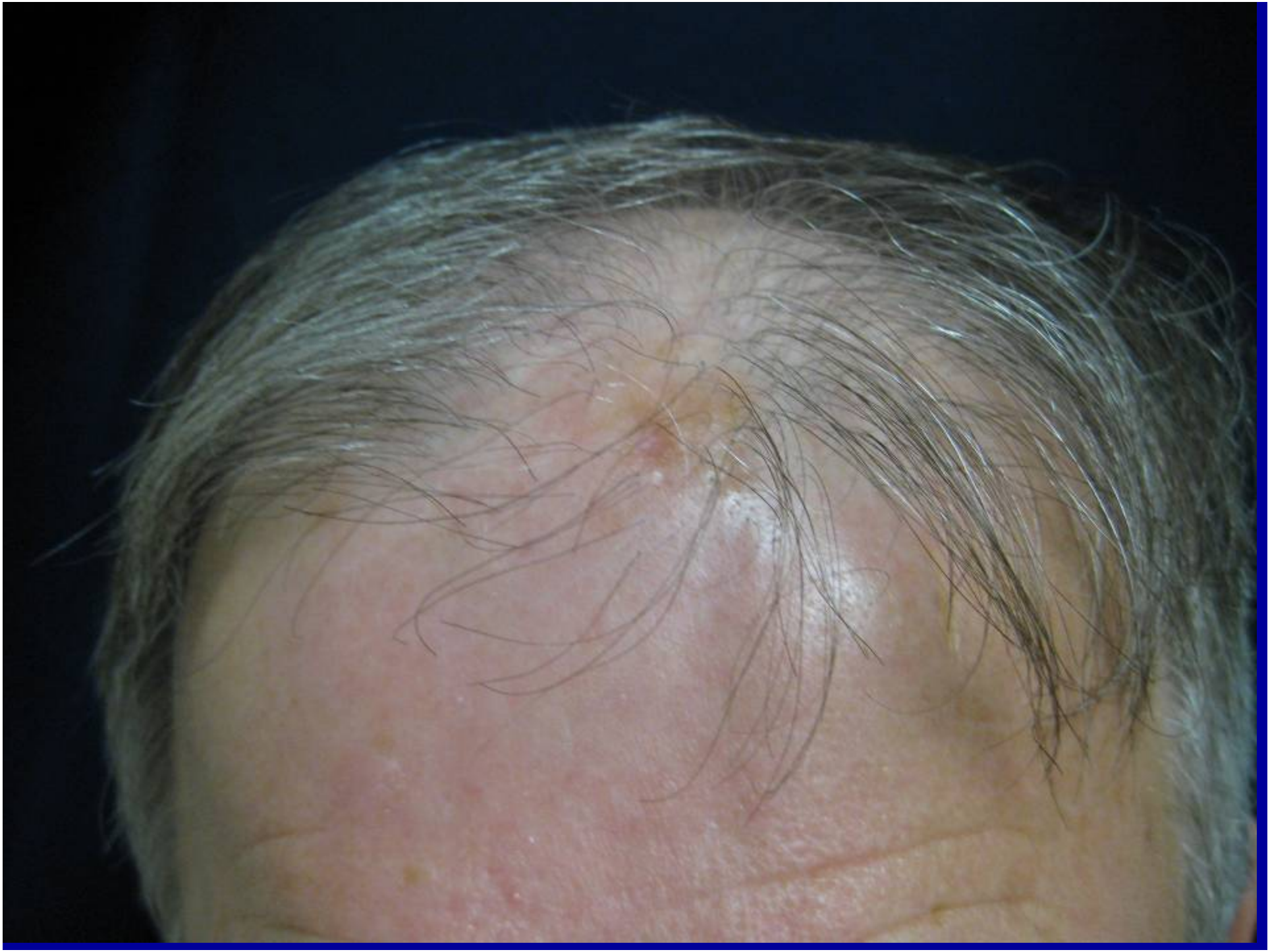
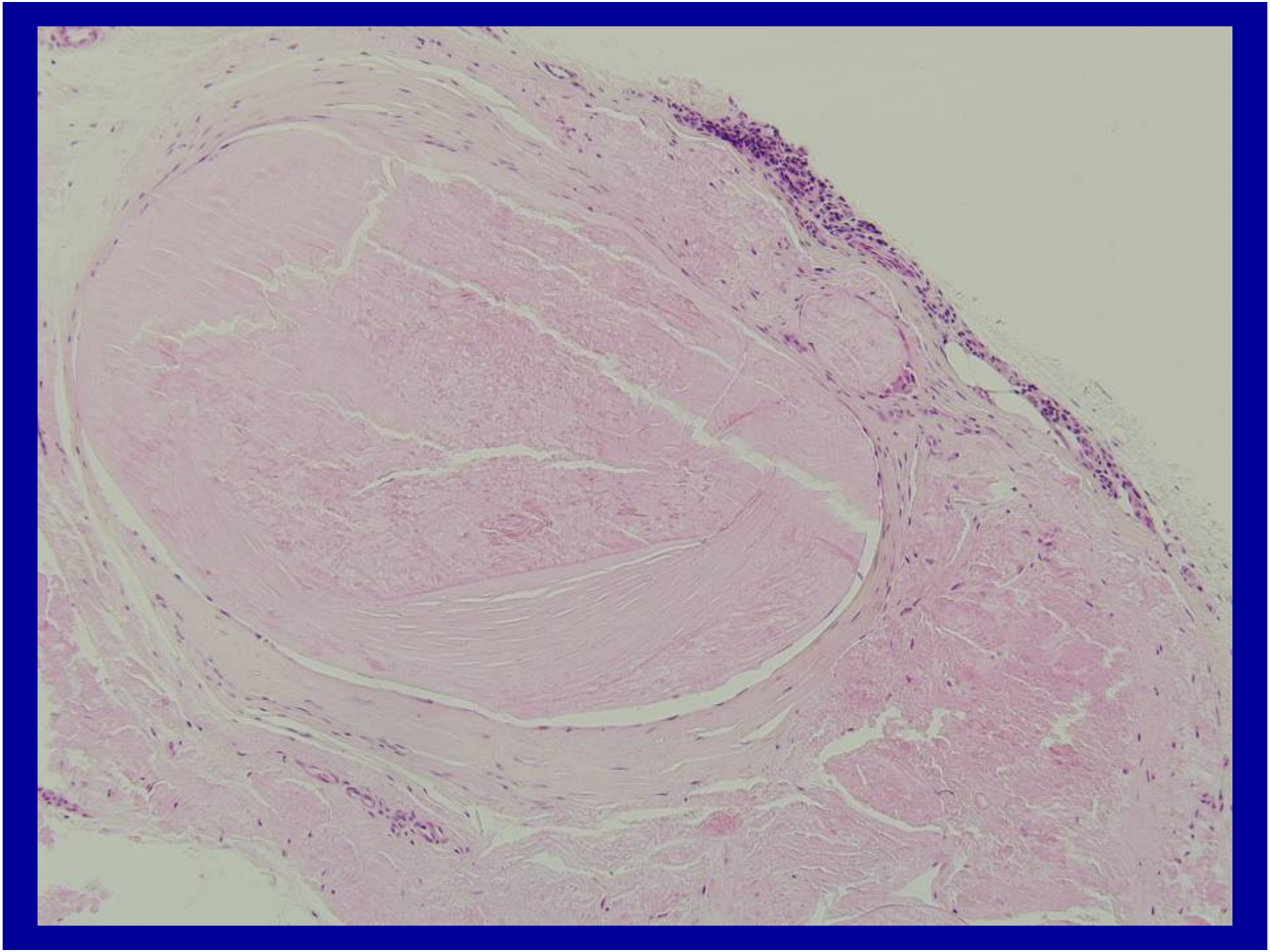
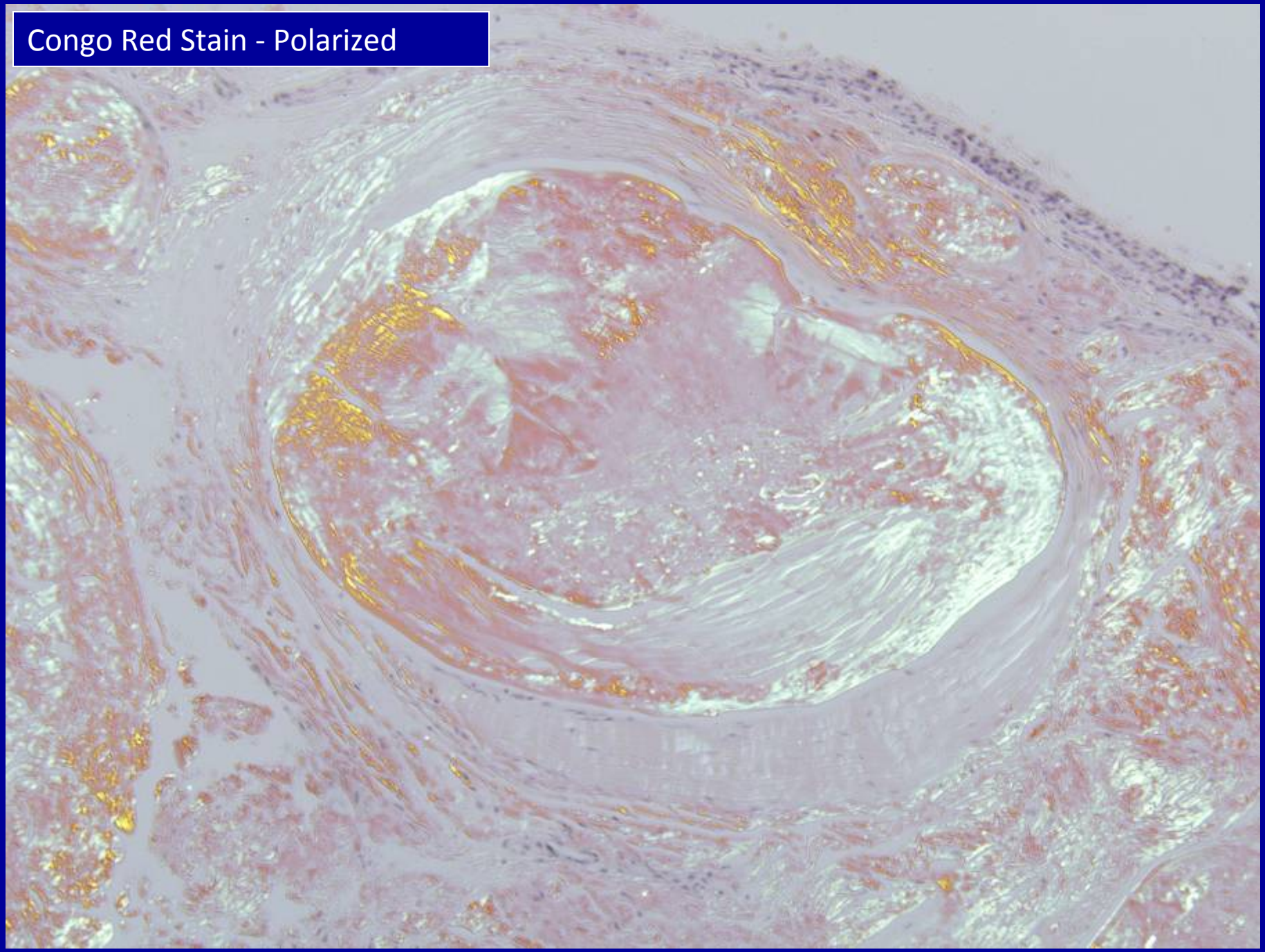
4. Yoneyama K, Tochigi N, Oikawa A, et al. Primary localized cutaneous nodular amyloidosis in a patient with Sjogren's syndrome: a review of the literature. J Dermatol. Feb 2005;32(2):120-3.





Congo Red Stain - Polarized



Case #26

AF

Xeroderma Pigmentosa

Case # 26
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	AF
Area of interest:	Face, chest and hands
PMHx:	Multiple skin cancers including SCC, BCC and Melanoma, Positive for migraine headaches.
Family History:	Multiple family members with skin cancers.
PE:	14 yo with extensive erythema and scaling of the back of sun exposed areas including the hands, face and upper chest. There is full facial actinic damage with atrophic damage of the lips which affects the aperture of his mouth. There are multiple scars from previous excisions.
Pathology:	Site 1: Right Preauricular- Invasive well differentiated squamous cell carcinoma extending greater than 2.0mm in depth. Site 2: Left Posterior Neck- Infiltrating basal cell carcinoma with perineural involvement. Site 3: Left cheek- Superficial spreading type melanoma. Clark's level IV, Breslow thickness of 1.7mm and 7 mitoses per HPF. He has had multiple other skin cancers in the past.
Laboratory:	The available genetic tests for Xeroderma Pigmentosum (XPA and XPC gene) were performed and were negative.
Diagnosis:	Xeroderma Pigmentosum (XP)
Treatment and Course:	This is a 14 year old male that has been diagnosed with non melanoma skin cancers starting at the age of five. Within the past year he presented with a pigmented lesion on his left cheek which was diagnosed as melanoma. He is currently being treated with Soriatane 10mg but was previously treated topically with 5-fluorouricil and diclofenac sodium (Solaraze) as well. His hobbies include hunting and fishing but he maintains that he performs his hobbies at night or when wearing appropriate cover-up including a large hat, long sleeves, long pants and extensive sunscreen. Staging workup related to his recent melanoma included a PET CT scan showing increasing activity in the bilateral cervical nodal basin, hypermetabolic enlargement of the right mylohyoid muscle, and findings suggestive of

mandibular involvement. He is set to undergo lymph node biopsy and is scheduled for excision of the melanoma via Moh's surgery.

Comments:

Xeroderma Pigmentosum is a heterogeneous autosomal recessive genodermatosis caused by a defective thymidine dimer excision repair mechanism that normally functions to repair DNA damage caused by ultraviolet light. Cumulative skin damage at a young age and skin cancers are the primary manifestations. The patients commonly get multiple premalignant and malignant neoplasms (1000x increased risk) including squamous cell carcinoma, basal cell carcinoma and melanoma. Patients also commonly develop ocular pathology due to ectropion, corneal opacities or ocular neoplasms. Many times patients get neoplasms on the tip of the tongue as well. Neurological abnormalities are common as well including acquired microcephaly, diminished or absent deep tendon stretch reflexes, progressive sensorineural hearing loss, and progressive cognitive impairment. XP has been found to be associated with mutations in XPA, XPB, XPC, XPD, XPE, XPG, XPF, and XP-V genes. Mutations in the XPA and XPC genes have been found to cause greater than 50% of cases and are the only tests clinically available to detect this disorder.

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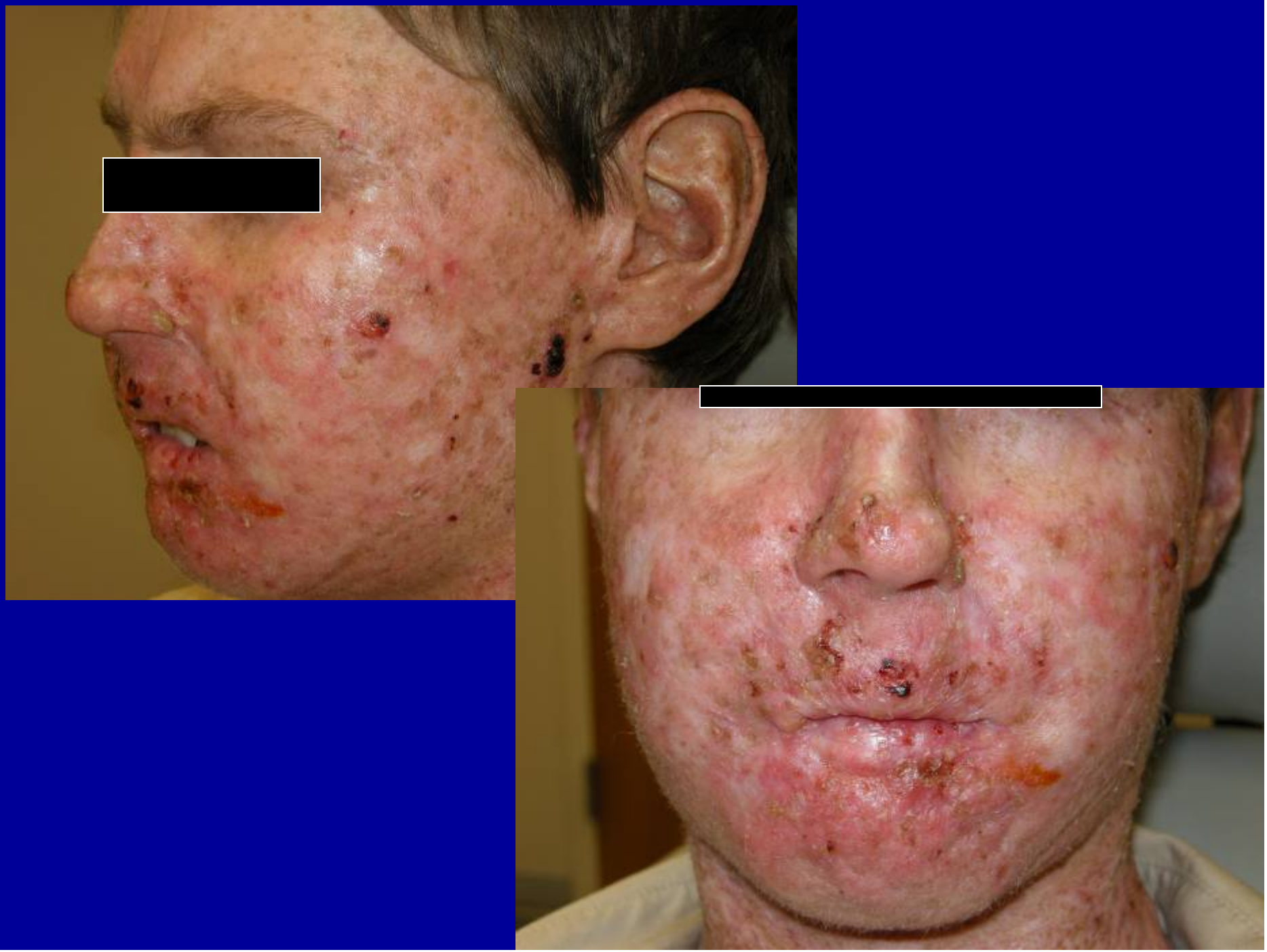

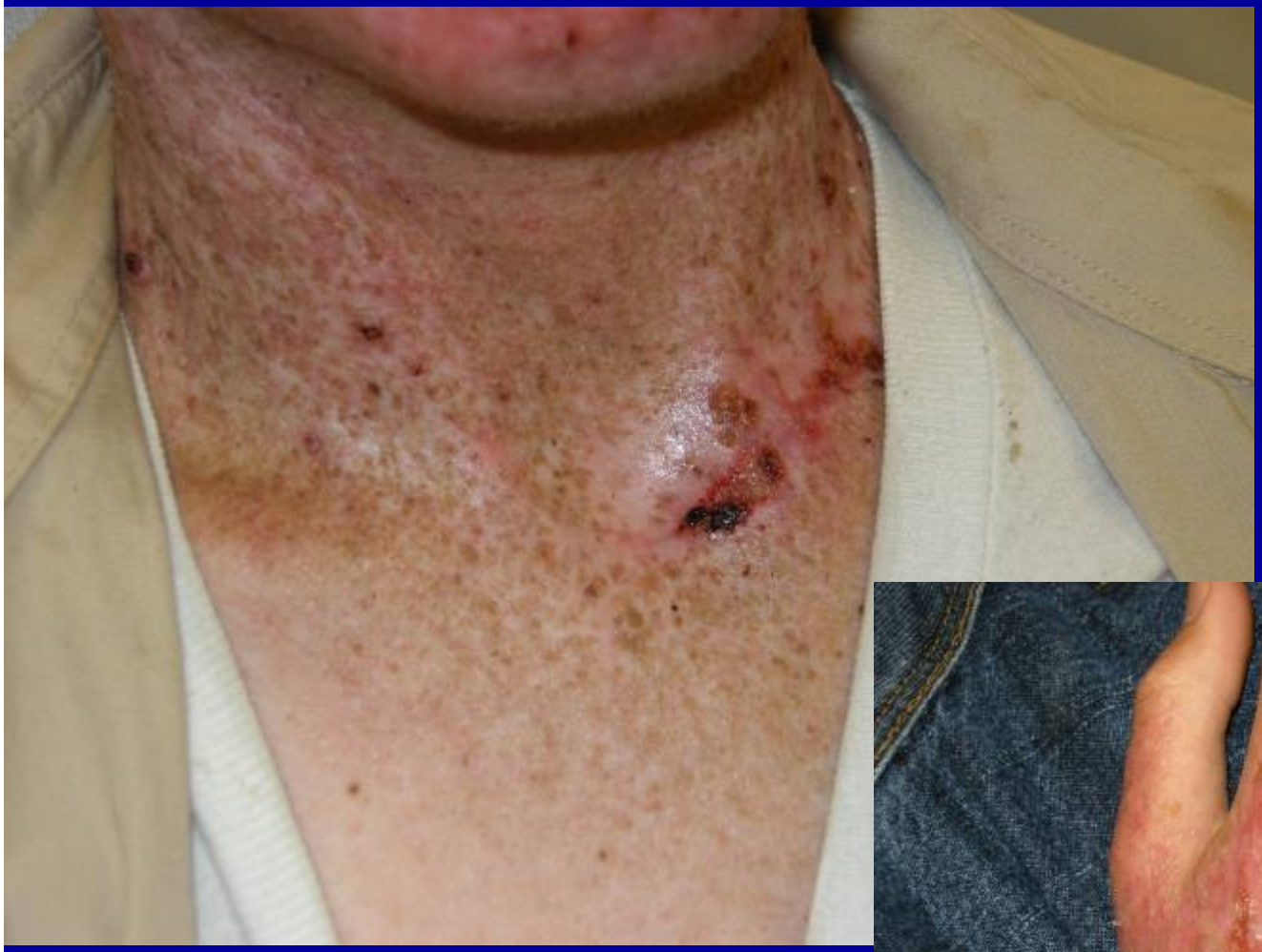

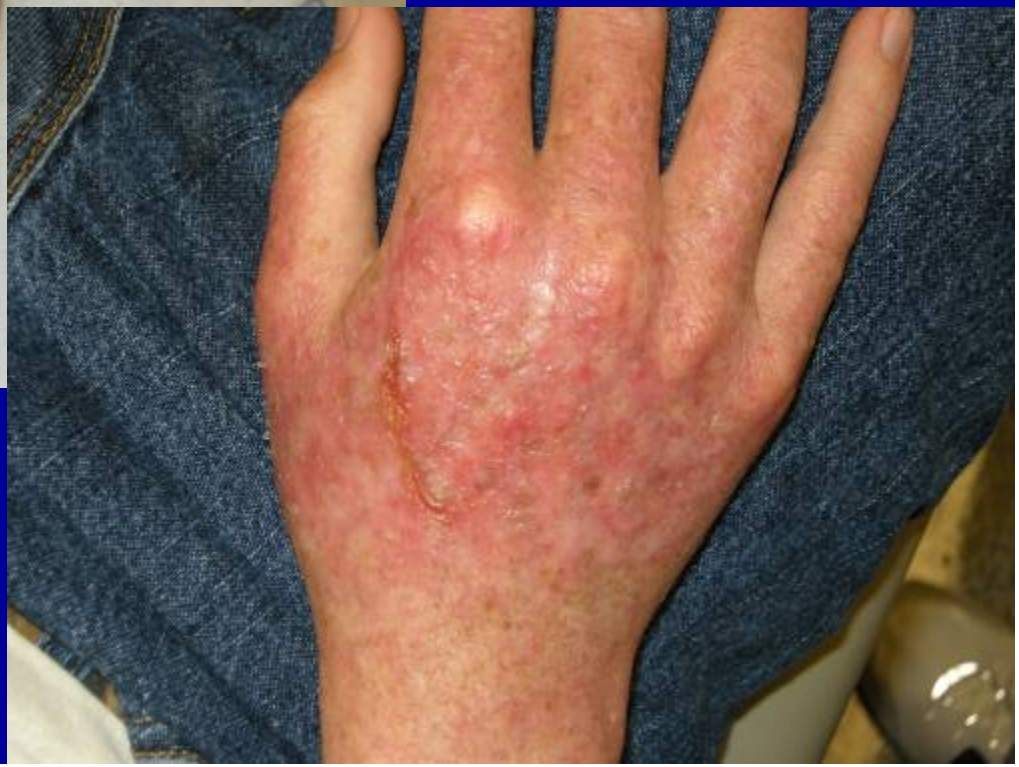

Case #28

NT

Adult Histiocytosis X

CASE # 28
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	NT
Area of Interest:	face, anterior chest
History:	69 year old Caucasian man initially presented in 2004 with a papular eruption located primarily on the frontal hairline. The patient also had similar lesions that occur on other areas of his face, anterior chest, shoulders, and pubic region. The lesions will resolve and then reappear, and they are associated with intense pruritis.
Past Medical History:	Hypertension, dyslipidemia, glaucoma.
Family History:	Prostate cancer, hypertension
Physical Exam:	There are scattered, dome-shaped, flesh-colored nodules located along the lateral forehead, preauricular areas, and the angle of the jaw. In addition, there are some smaller 3-4mm flesh-colored papules located on the anterior chest.
Pathology:	A dense dermal infiltrate which includes large cells with abundant cytoplasm. There are many smaller lymphocytes admixed among the larger cells. The infiltrate is centered on or adjacent to hair follicles in areas. Immunoperoxidase staining reveals scattered CD3-positive T-cells throughout the infiltrate with rare scattered CD20-positive B-cells. The majority of the infiltrate labels avidly and diffusely with CD1a, consistent with Langerhans cells.
Laboratory Studies:	Basic metabolic panel and liver function tests are within normal limits. Complete blood count is significant for leukopenia with a WBC of 2.82 and a mild anemia with a hematocrit of 38. CRP is also mildly elevated.
Diagnosis:	Adult Histiocytosis X, limited to cutaneous involvement
Treatment and Course:	Pt was initially treated by his internist with a short course of prednisone in addition to antihistamines, with no improvement in the skin lesions. Pt was eventually referred to a dermatologist, and a skin biopsy was performed which was consistent with Langerhans Cell Histiocytosis. For the skin lesions, the patient was started on topical steroids to apply as needed for pruritis, which did improve his symptoms.

The patient was also referred to a hematologist/oncologist who performed a bone scan, in addition to CT scans of chest, abdomen, and pelvis to rule out systemic involvement. In addition, the hematologist/oncologist recommended further skin biopsies to be obtained to confirm the diagnosis given the patient's older age at diagnosis. The additional skin biopsy was consistent with Langerhans cell histiocytosis.

The patient's skin lesions have waxed and waned, and pt has developed a new lesion on the right outer eye which is being followed closely by Ophthalmology. The hematologist/oncologist plans a trial of systemic methotrexate if the patient has a symptomatic flare.

Comment:

Langerhans Cell Histiocytosis (Histiocytosis X) is a disease characterized by an abnormal proliferation of Langerhans cells that has a wide range of clinical presentations. These include lytic bone lesions, lymphoproliferative malignancies,various degrees of organ dysfunction, cutaneous manifestations, and diabetes insipidus. The etiology and pathogenesis of this disorder are unknown.

This case is of particular interest because the majority of patients affected by this disease are children from the ages of 1 to 4 years. In patients that present as adults, the average age of presentation is 33 years. In addition, this patient has localized disease of the skin, which is less common in adult patients. In the majority of adult patients, multisystem disease is most common.

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2. Hussein MR. Skin-limited Langerhans' cell histiocytosis in children. Cancer Invest. 2009 Jun;27(5):504-11.

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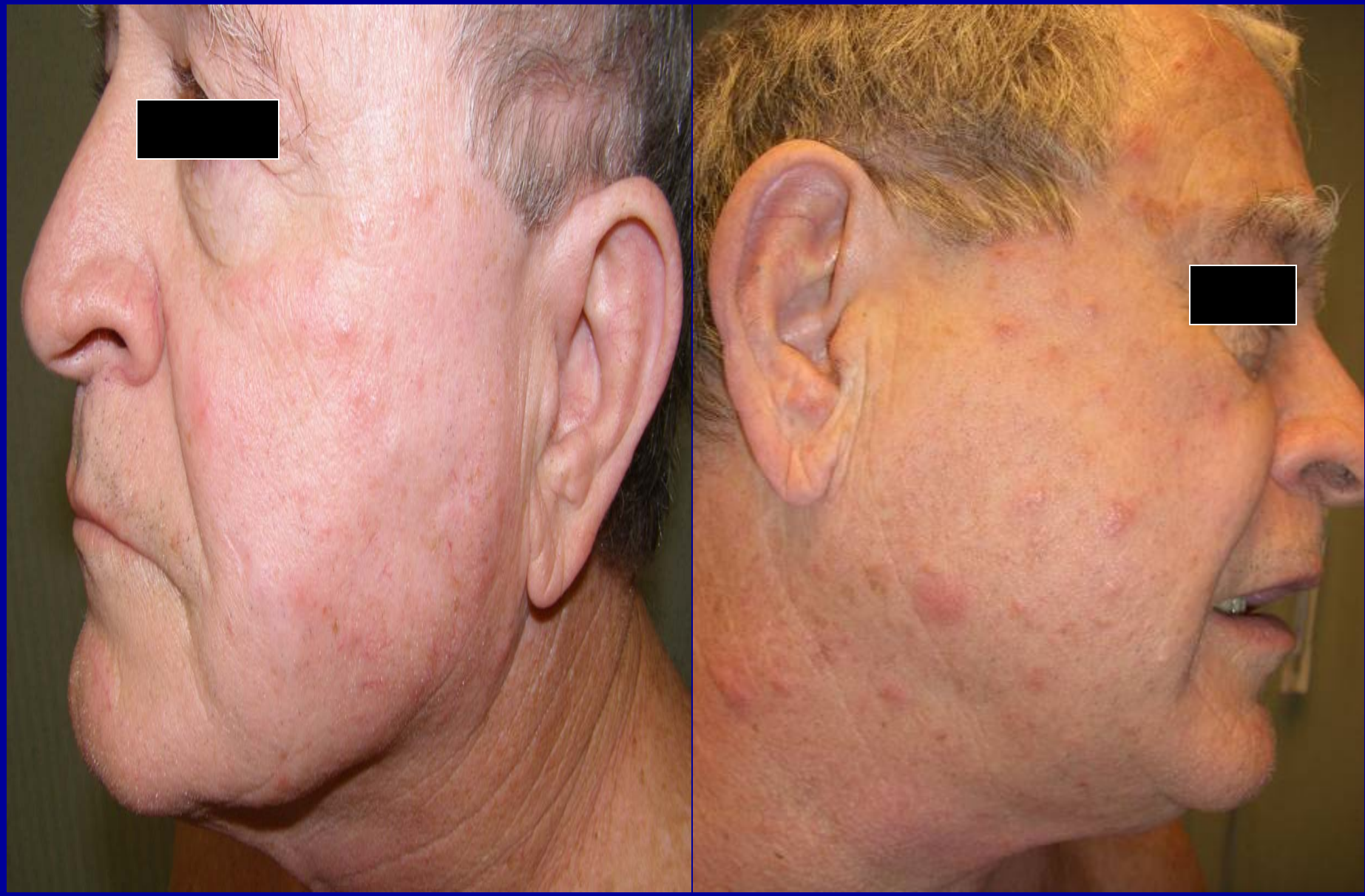

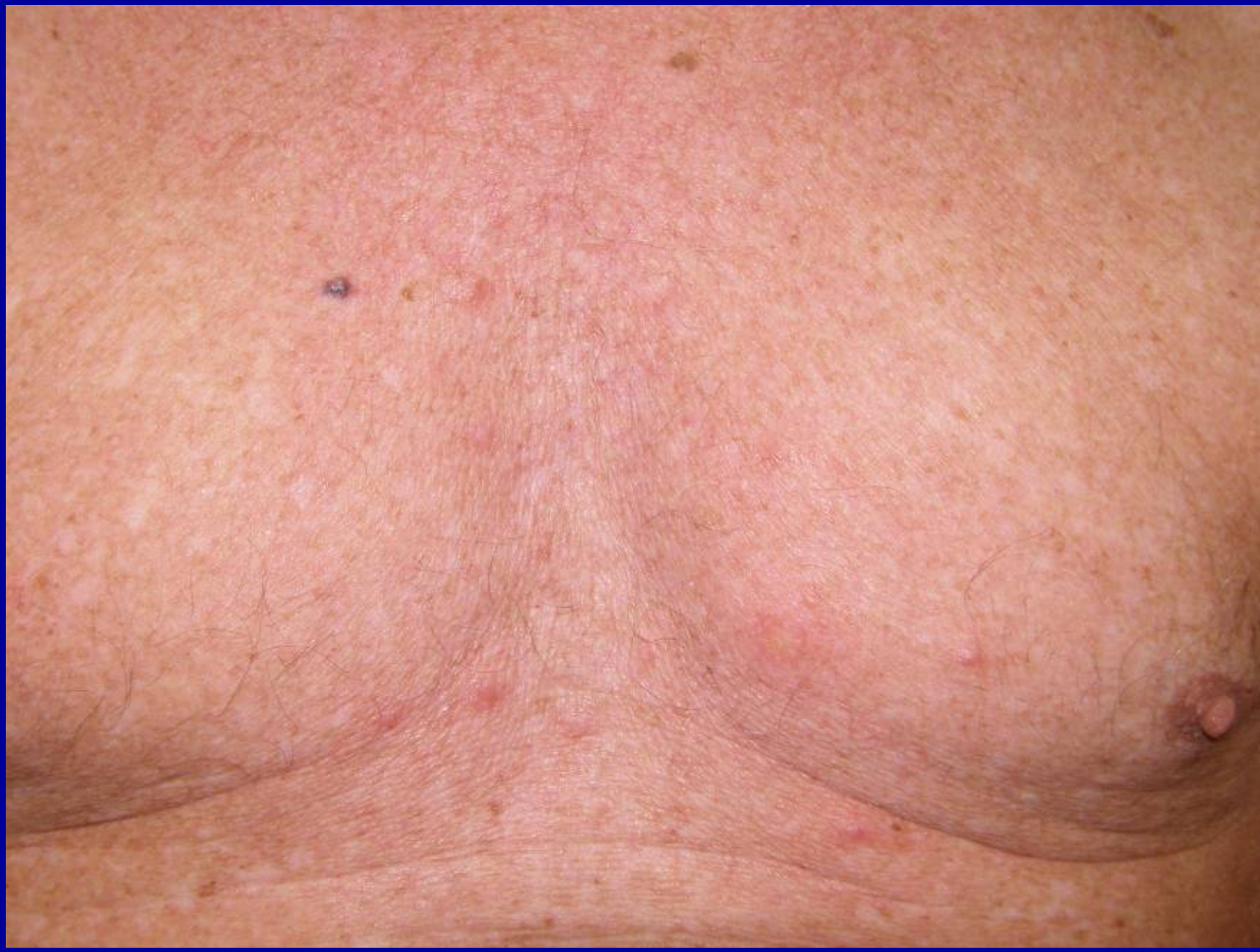

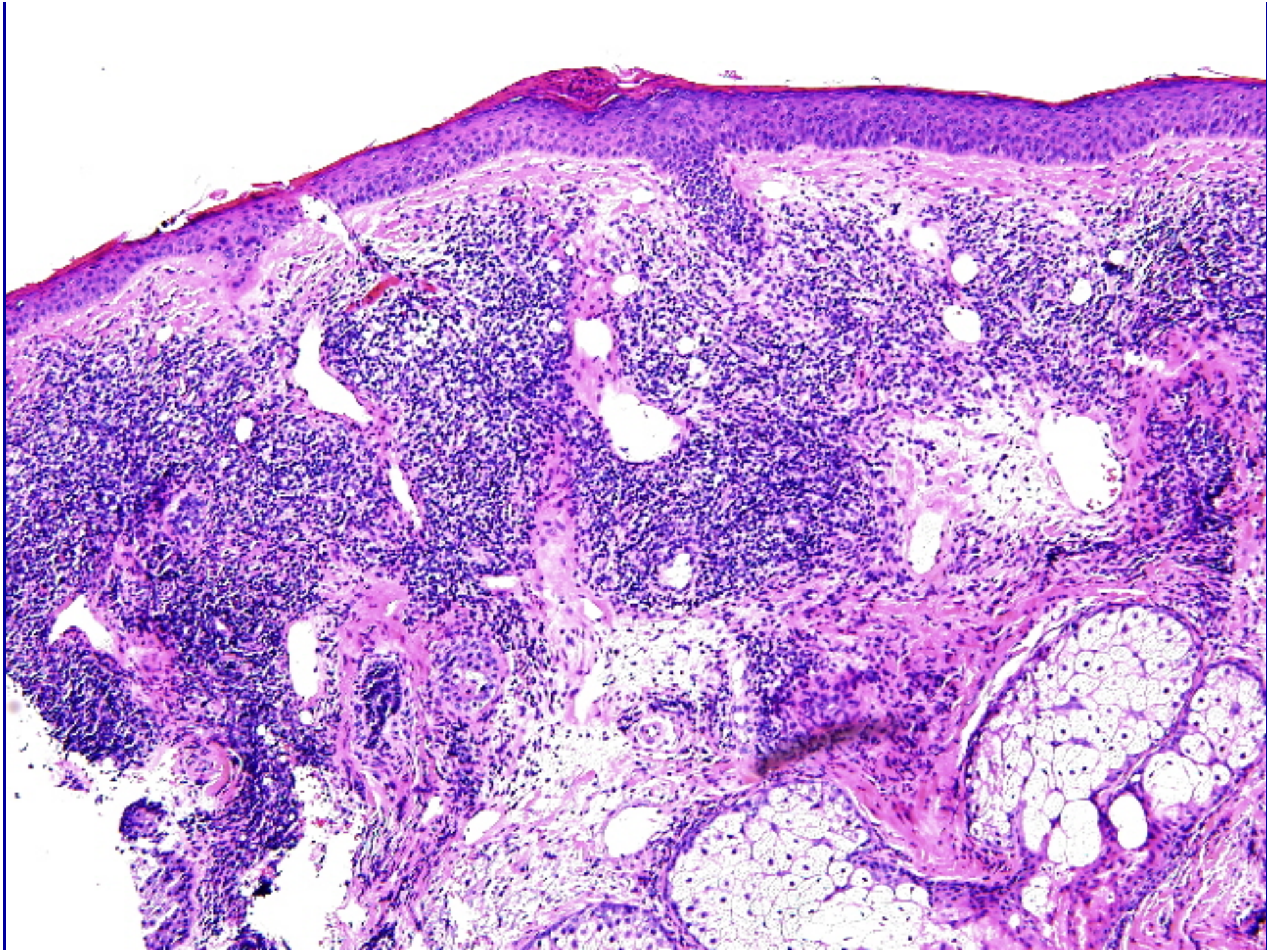

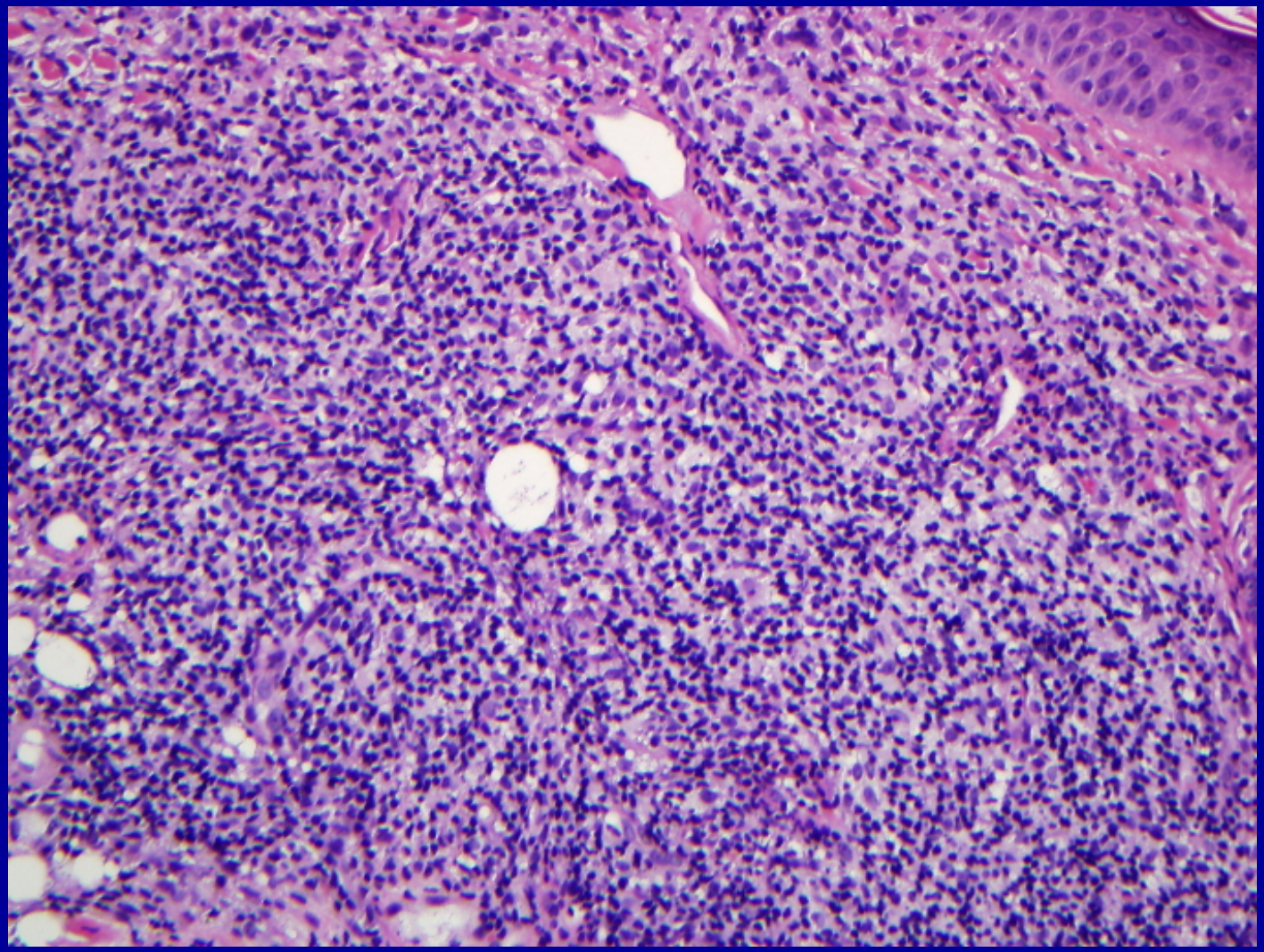

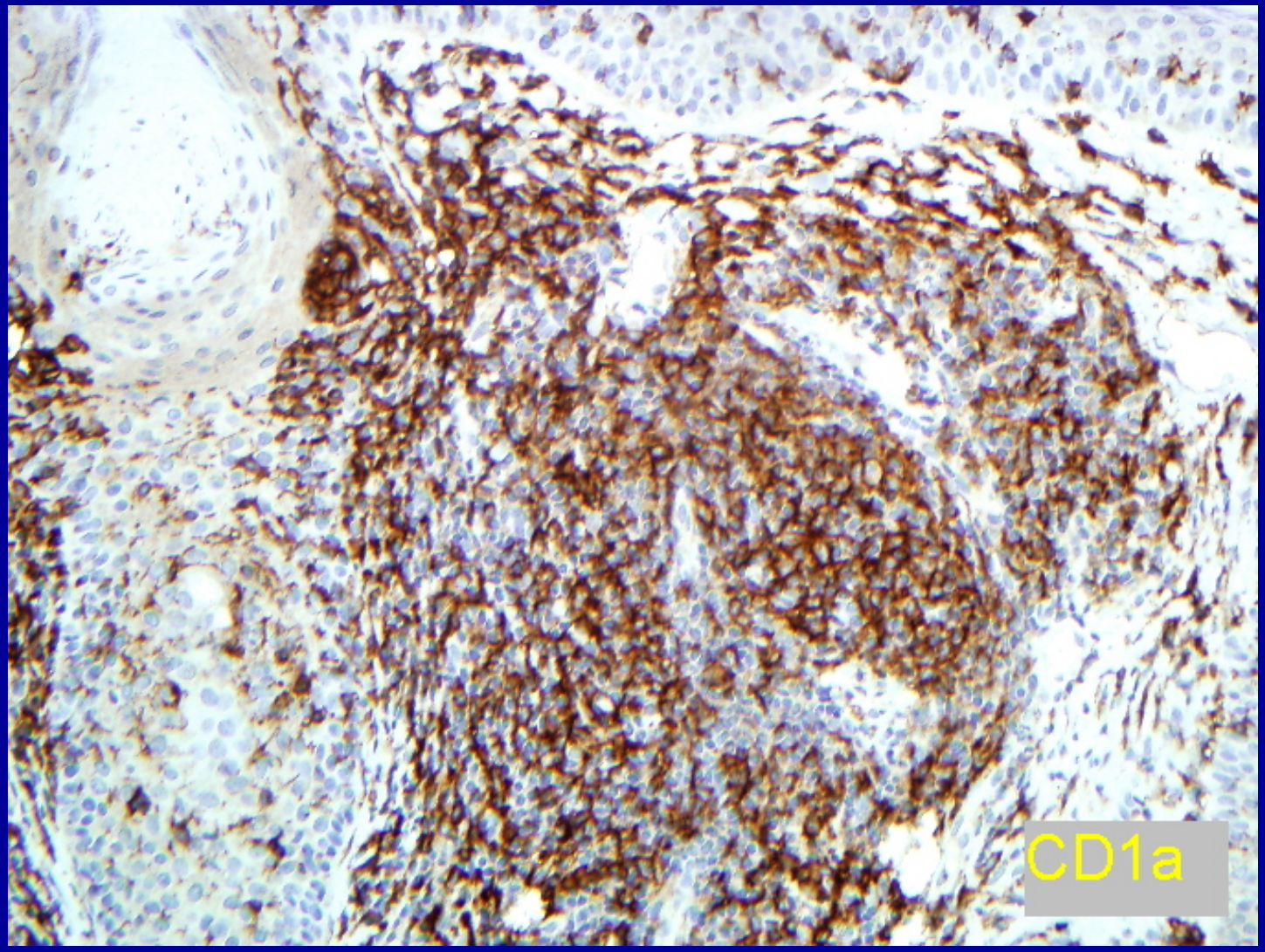
CD1a

Case #29

SB

Eruptive Keratoacanthoma Syndrome

CASE # 29
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	SB
Area of Interest:	upper and lower extremities, soles, palms
History:	32 year old Caucasian woman presents with a 16 year history of several nodules scattered primarily on her bilateral upper and lower extremities, including the palms and soles. The lesions will sometimes ulcerate, and occasionally become infected. When the nodules resolve, they will leave a scar or area of darker pigmentation.
Past Medical History:	Depression
Family History:	None
Physical Exam:	On the bilateral upper and lower extremities, there are shallow, cribiform scars present. In addition, there are multiple hyperkeratotic nodules on an erythematous base, with overlying crust and a central depression present on bilateral upper and lower extremities. These lesions are tender to palpation. In addition, there are flat-topped, verrucous plaques present on bilateral palms and soles, resulting in a cerebriform appearance of the feet.
Pathology:	There is a proliferation of pale-staining atypical keratinocytes with ample cytoplasm arrayed as bulbous lobules extending from the epidermis. Keratinocytes at the periphery of the lobules display more prominent atypia. At the periphery of the lobules, there is an inflammatory infiltrate of lymphocytes and eosinophils. In some foci, the infiltrate obscures a portion of the neoplasm. To the sides of the neoplasm, there is marked dermal elastosis.
Laboratory Studies:	Basic metabolic panel and complete blood count are within normal limits. Liver function tests are normal, except for a mildly elevated ALT. Lipid panel shows elevated total cholesterol of 222, and LDL of 162. HDL is low at 33. Triglycerides had been previously elevated, but are within normal limits now.
Diagnosis:	Eruptive Keratoacanthoma Syndrome (Grzybowski Variant)
Treatment and Course:	Pt has been taking Soriatane for several years, which seems to help control the number of keratoacanthomas. Pt initially started at 25 mg Soriatane daily, then alternated between 25 mg and 50 mg daily, and is now taking 50 mg

Soriatane daily. If the patient discontinues Soriatane, she notices an increase in the number of lesions. Pt is also using Efudex 5% to the lesions of concern every 2-3 days. We will continue to monitor liver function tests and lipid panels.

Comment:

Eruptive Keratoacanthoma Syndrome is very rare and most patients have no history of affected family members. The etiology of this disease is unknown, although chronic sun exposure plays a role in the development solitary keratoacanthomas. The potential for malignant behavior of these lesions is controversial; however, the current practice is to treat keratoacanthomas as malignant neoplasms. Despite the fact that patients have multiple lesions in the Grzybowski variant, no increased risk for internal malignancy has been shown, and there are no known cases of metastasis.

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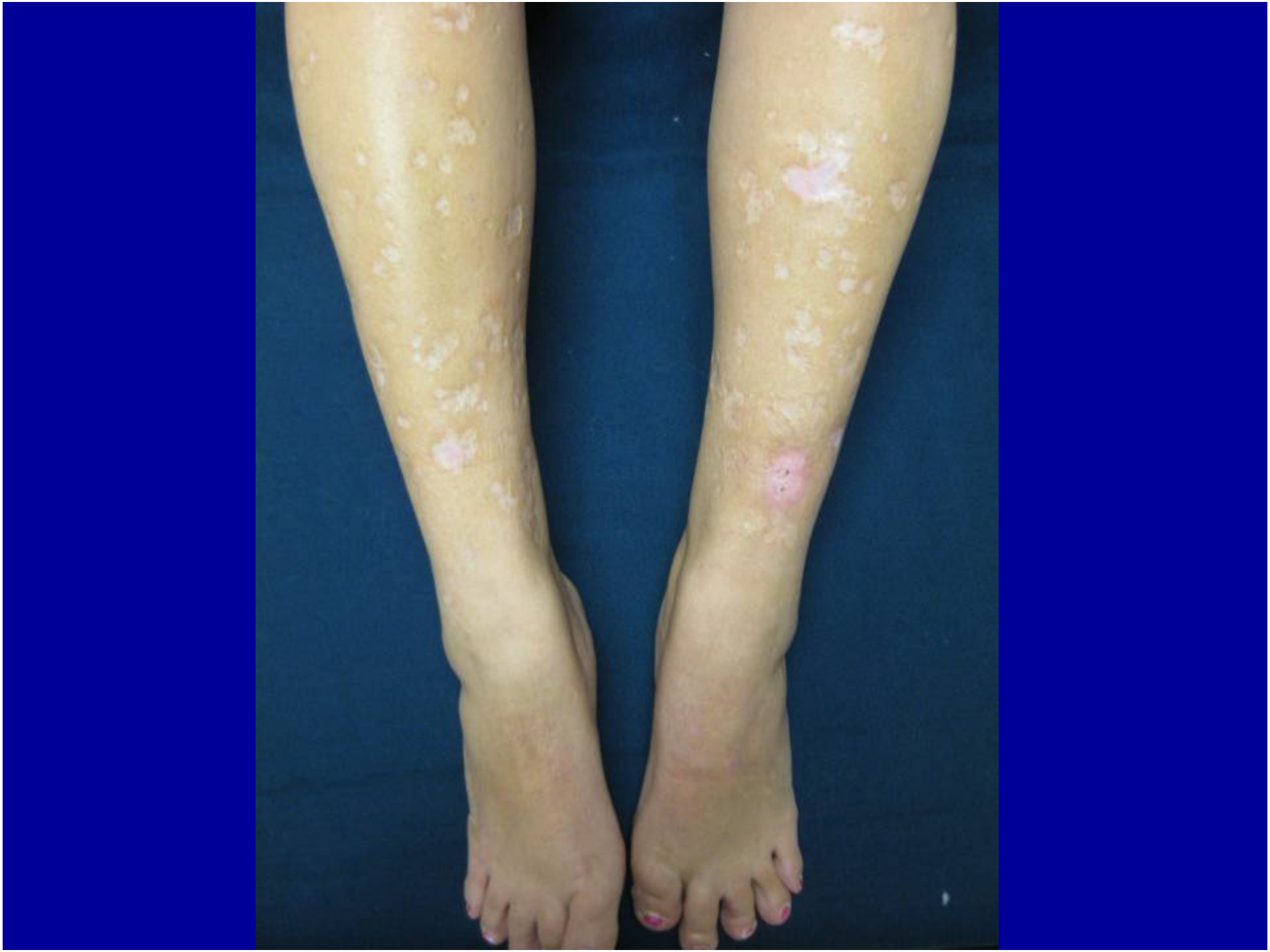

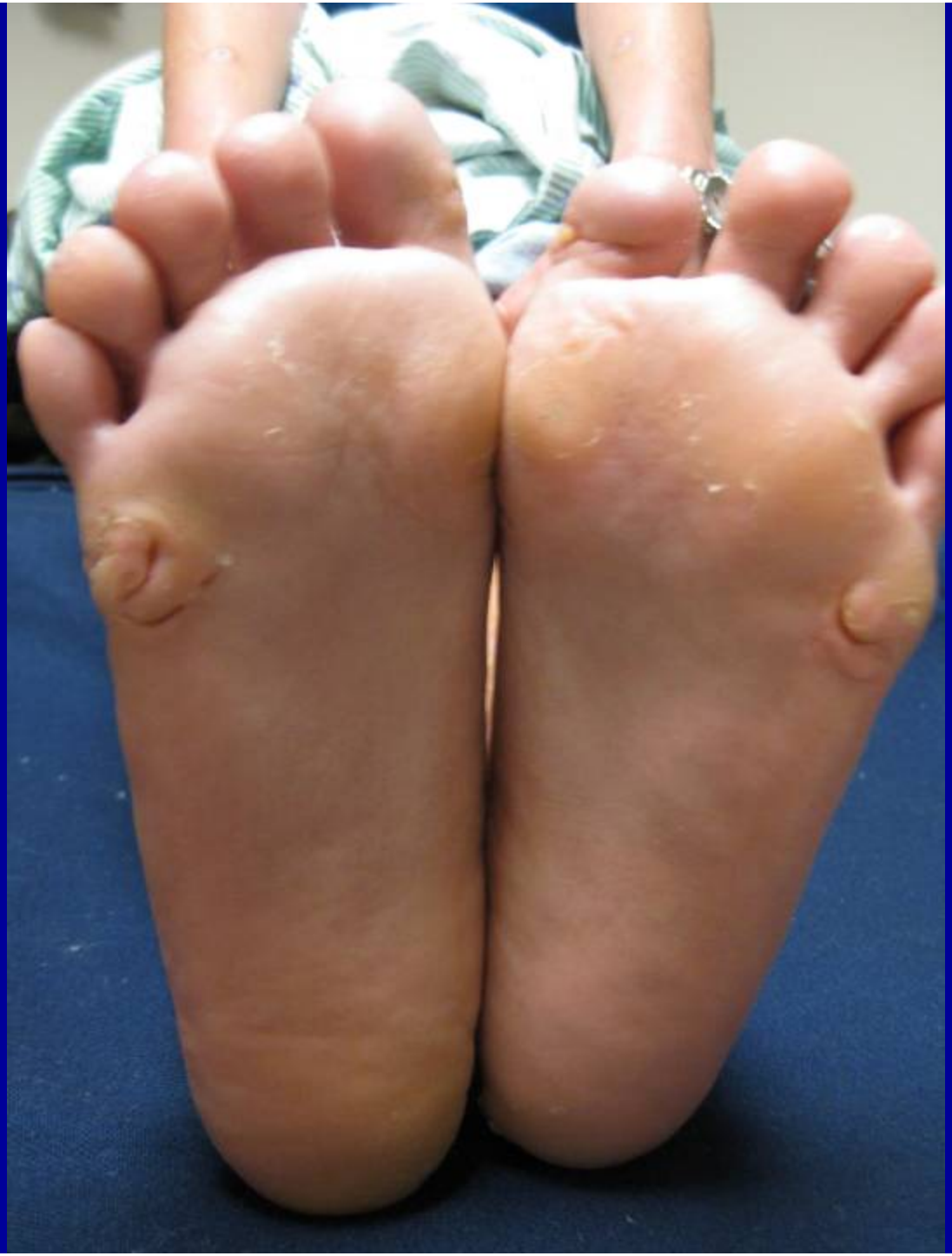

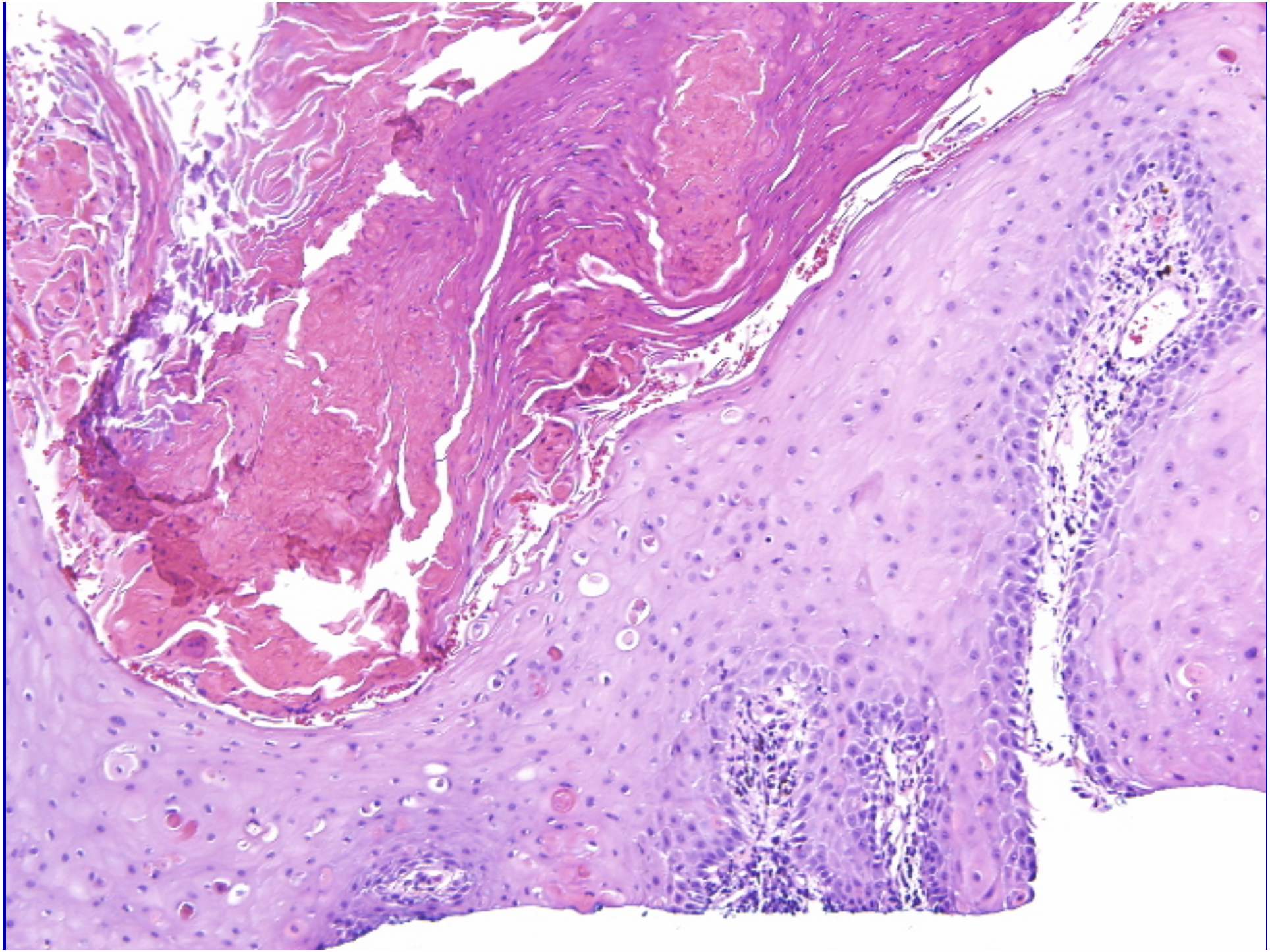

Case #30

TB

Anhidrotic Ectodermal Dysplasia

CASE # 30
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: TB

Area of Interest: Full body, focusing on bilateral lower extremities, oral cavity.

History: 48 year old African American Male with longstanding history of anhidrosis and loss of teeth. Patient presented to dermatology clinic when his symptoms progressed to a burning in the skin on hot days.

Past Medical Hx: DM II

Physical Exam: Diffuse xerosis with slight ichthyotic change on distal lower extremities. Loss of teeth.

Diagnosis: Anhidrotic Ectodermal Dysplasia

Treatment and Course: The patient was treated with a combination of topical medications and education about his condition. For his xerosis, icthyosis, and burning sensation a triamcinolone and Eucerin mixture was recommended as well as Aveeno and Atarax for symptom control for the body. Fluocinonide was recommended on the scalp. He was also told to avoid petroleum products due to the increase in occlusion and therefore body temperature. He was already aware of his inability to cool himself off by sweating and was practicing heat avoidance appropriately.

Discussion: Anhidrotic Ectodermal Dysplasia is a disorder composed of hypohidrosis or anhidrosis, hypotrichosis, and anodontia. The disease is most commonly X-linked recessive and therefore seen more commonly in males. It is possible for female carries to have partial expression of some of the characteristic findings from the disorder, such as patches of hypohidrosis. Other characteristic findings are conical shaped teeth, thin skin, prominent frontal ridge, nail findings, or facial features similar to congenital syphilis. Special care should be taken to prevent hyperthermia in these patients. Fever of unknown origin in a child can be a clue to diagnosis. Associated findings are asthma, atopic dermatitis, and increased susceptibility to infection.

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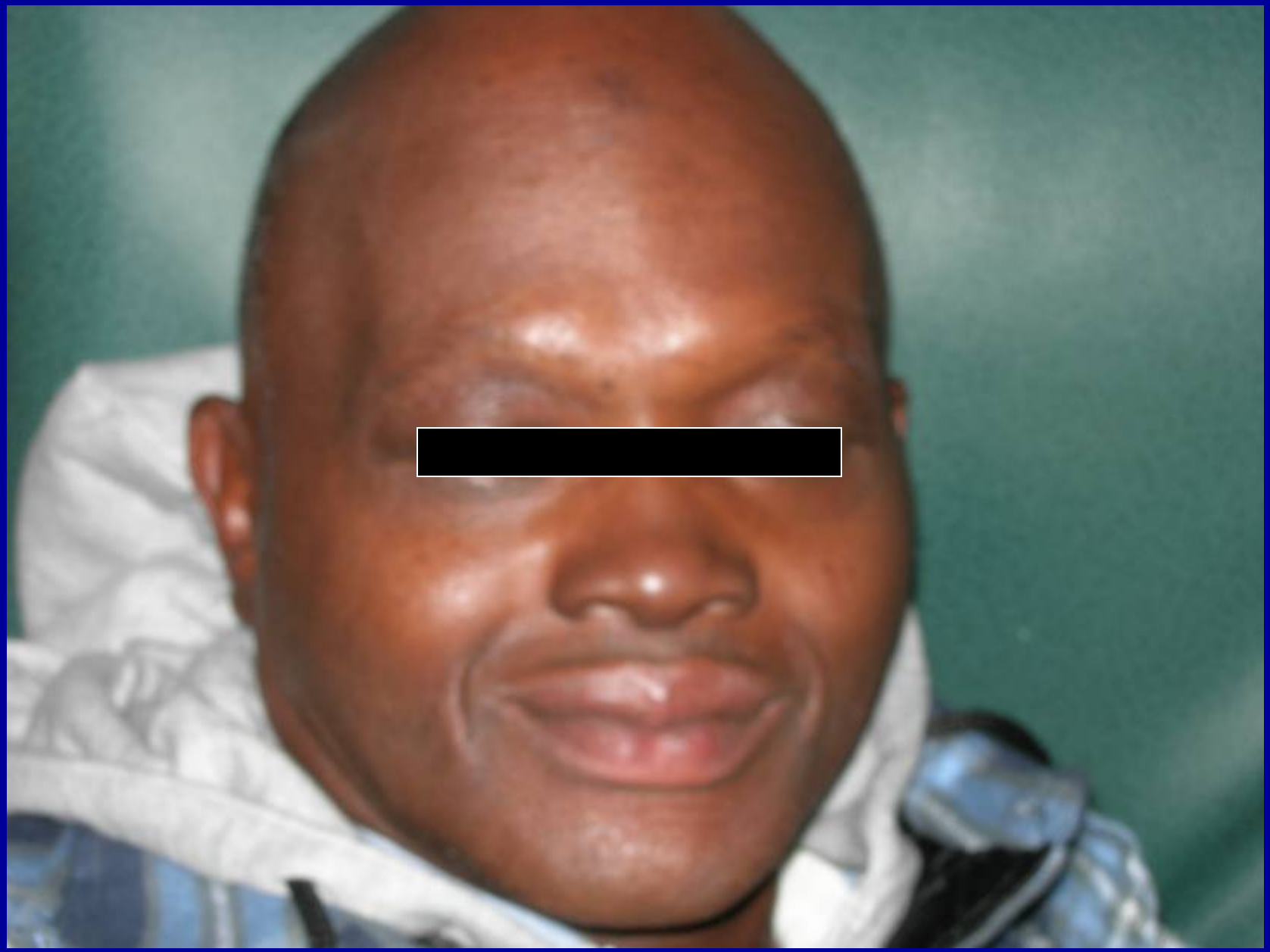

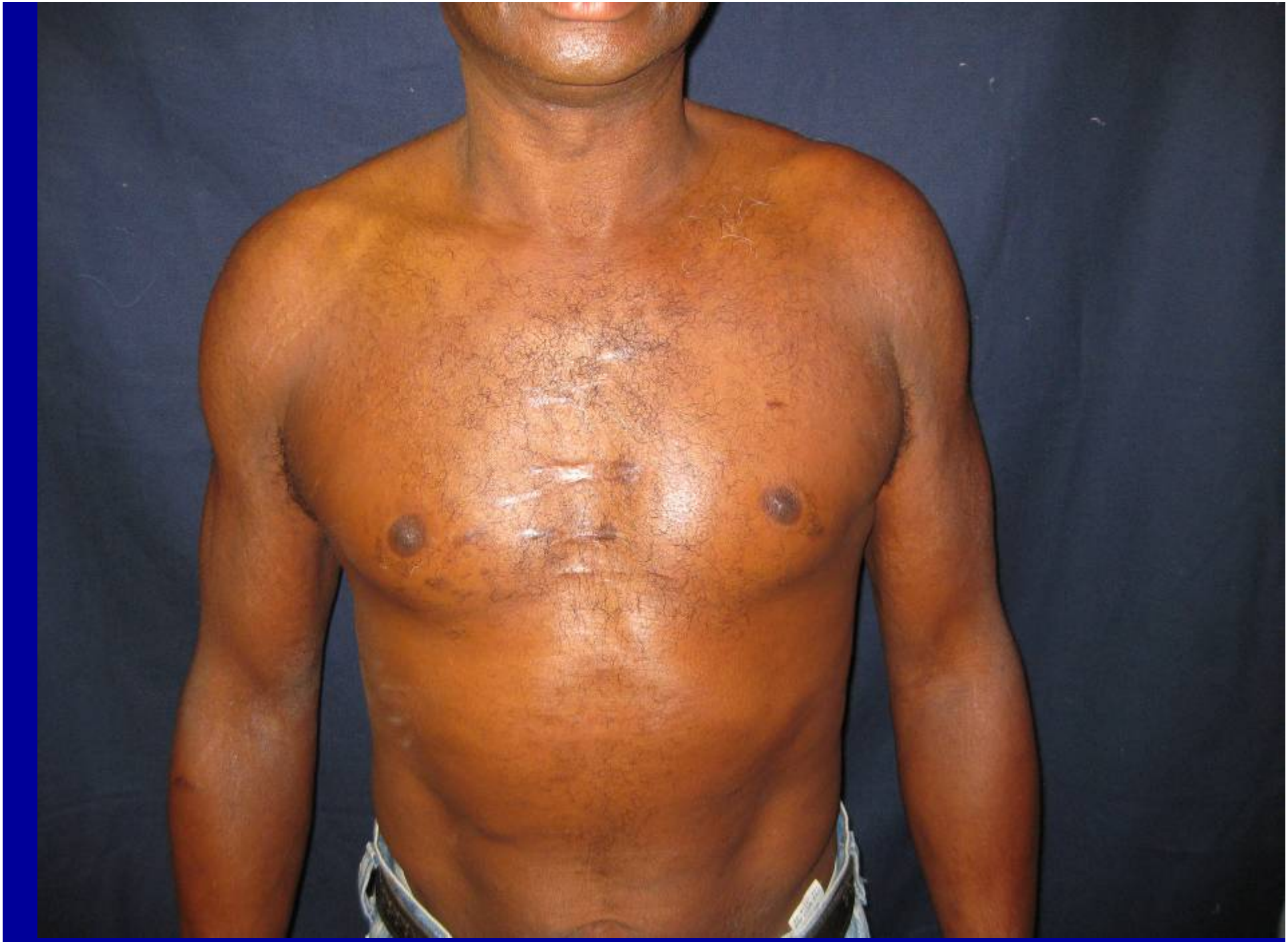

Case #31

JE

Basal Cell Nevus Syndrome

CASE # 31
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	JE
Area of Interest:	Face
History:	46yo Caucasian man with multiple BCCs removed during his lifetime, the majority being on the face. Patient also has mandibular cysts documented by x-ray in 2002 and macrocephaly.
PMH:	Multiple BCCs, Multiple Sclerosis, Meningioma at age 3, chondrosarcoma of left hip.
FH:	No family history of multiple skin cancers.
Physical Exam:	Macrocephaly with head circumference in the 98th percentile. Multiple biopsy scars on the face. No palmar pits.
Pathology:	Multiple different biopsies with similar characteristics of predominant basal cell types with peripheral palisading nuclei and clefting aftifact between the epithelium and the stroma.
Diagnosis:	Basal Cell Nevus Syndrome
Treatment and Course:	The patient is followed closely by the Dermatology Department for frequent skin checks and monitoring for any new papules that could represent a new basal cell carcinoma. The patient's lesions have been treated by surgical excision.
Discussion:	Basal Cell Nevus Syndrome is an autosomal-dominantly inherited syndrome characterized by multiple to numerous BCCs developing as early as childhood and persisting through life. Odontogenic keratocysts, palmar and plantar pits, as well as facial abnormalities are also seen in the syndrome. It is caused by a mutation in the ptch gene, a tumor suppressor gene, but can also be caused by sporadic mutations. Treatment modalities range from surgical excision, laser, PDT, cryotherapy, and imiquimod. Prevention of further gene alteration by UV light is important and sunscreen use should be stressed in this syndrome.

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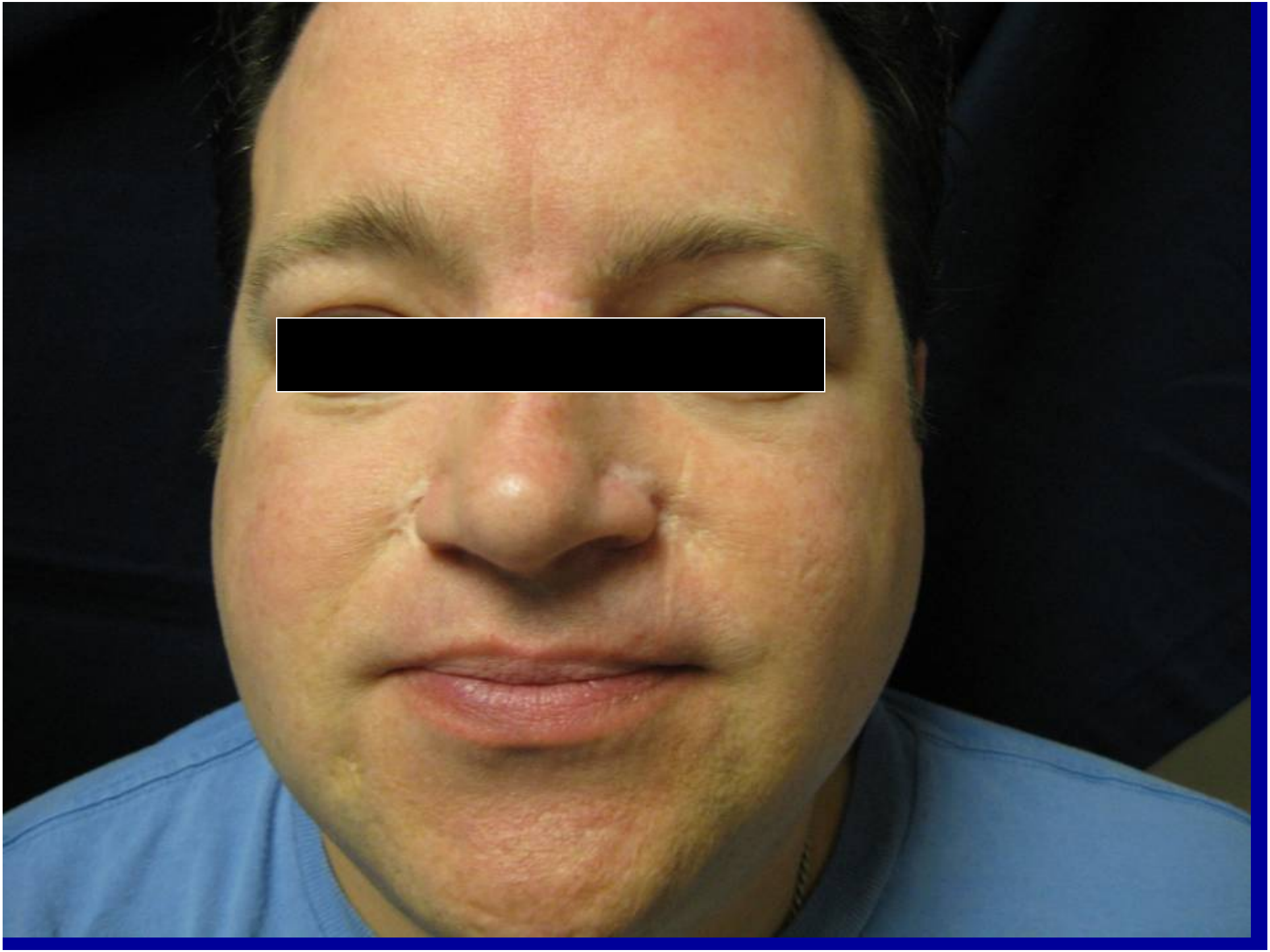

Case #32

SH

Membranous Aplasia Cutis Congenita

CASE # 32
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: SH

Area of Interest: Vertex of Scalp

History: One month old Caucasian girl with lesion of interest present at birth.

PMH: Full term birth, no complications at delivery.

FH: No family history of skin disease.

Physical Exam: 1 x 1.5cm oval patch on the vertex of the scalp characterized by a central clear membrane surrounded by a ring of thickened and elongated hairs.

Diagnosis: Membranous Aplasia Cutis Congenita

Treatment and Course: The patient was initially evaluated one week after birth by a dermatologist. Ultrasound revealed only a lack of epidermis.

Discussion: Aplasia Cutis Congenita is a broad term that encompasses nine different congenital types of skin aplasia. The types are classified by location, inheritance, shape, and associated findings. Scalp lesions are classified as either membranous or non-membranous. Membranous lesions are characterized by their smaller oval shape, location on the vertex, membranous atrophic appearance, and possible presence of a hair collar sign. Non-membranous lesions are larger with irregular borders. In either case a hair collar sign can signify an underlying cranial defect. Etiology is unknown, but suspected to be abnormalities at ectodermal fusion lines. Some scalp lesions are familial, however, most are sporadic in nature. Prognosis depends on the depth and size of the lesion of concern. In our patient, the lesion did not involve deeper structures so no further treatment was necessary.

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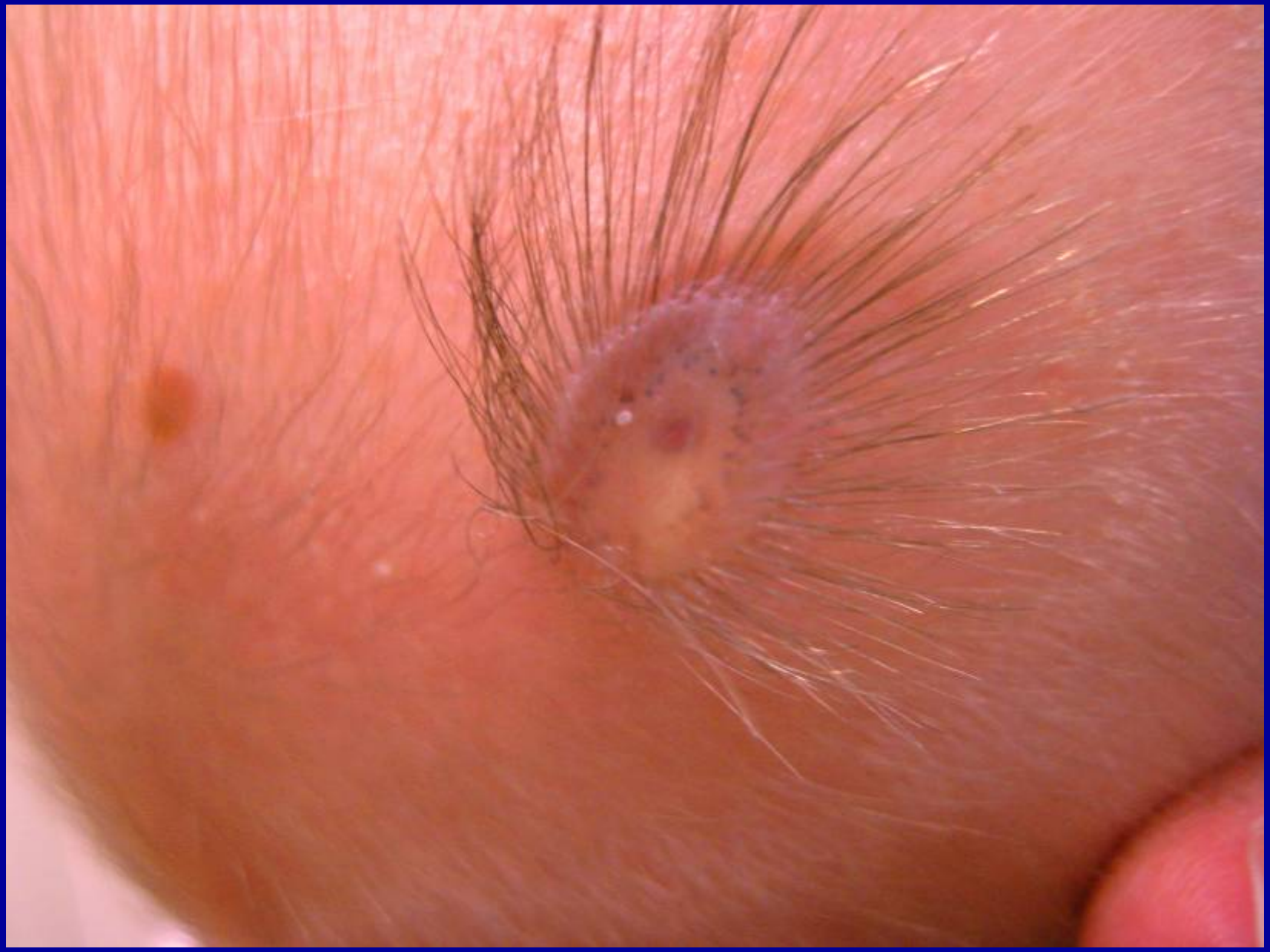

Case #33

YC

Phakomatosis Pigmentovascularis II

CASE # 33
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient: YC

Area of Interest: Left arm, chest, and leg.

History: Two year old Hispanic female presented to Dermatology for evaluation of vascular and melanocytic lesions. Patient also with incidental eczema at the time.

PMH: No noticed limb deformities or neurologic abnormalities.

Physical Exam: Erythematous, blanching, vascular patches located on the left upper extremity, left upper chest, left upper back and left lower extremity extending to the knee. Also seen was some hyperpigmentation of the left lower sclera and on the left zygoma.

Diagnosis: Phakomatosis Pigmentovascularis II

Treatment and Course: The patient was seen initially on referral in our resident clinic. At this point the diagnosis was made and a thorough history and physical exam were done to rule out history of seizure or neurologic deficiencies. She is seen by ophthalmology due to the nevus of Ota with scleral involvement. She was also seen in Peds Derm clinic were limb length and circumference measurements were done and will be followed in the future. If any limb deformities should arise she will be seen by orthopedics.

Discussion: Phakomatosis Pigmentovascularis describes a syndrome of a large vascular lesion in association with a significant pigmented nevus. Currently there are two classification systems that have been published. The first system described used PPV type I through V with an unclassified group, as well as an A and B subgroup for each group. Subgroup A included only cutaneous findings while B also had extracutaneous involvement. The newer classification intended to simplify the system and make it easier to use for the clinician. It contains four groups, one being reserved for unclassifiable lesions. Our patient falls under type II of the traditional classification or phakomatosis cesioflammea of the new classification. The most common pigmented nevus found in phakomatosis cesioflammea is a nevus of Ota or mongolian spot. Associated extracutaneous findings, such as neurologic abnormalities or ocular melanosis, may be present. Investigation should be done to rule out extracutaneous findings and if seen appropriate referrals should be made. If no further

associated abnormalities are seen observation is recommended. For cosmetic concerns laser treatment of the lesions is a possibility.

Comparison of Classification Systems for Phakomatosis Pigmentovascularis	
IIa, IIb	Cesioflammea
IIIa, IIIb	Spilorosea
Va, Vb	Cesiomarmorata
IVa, IVb	Unclassifiable

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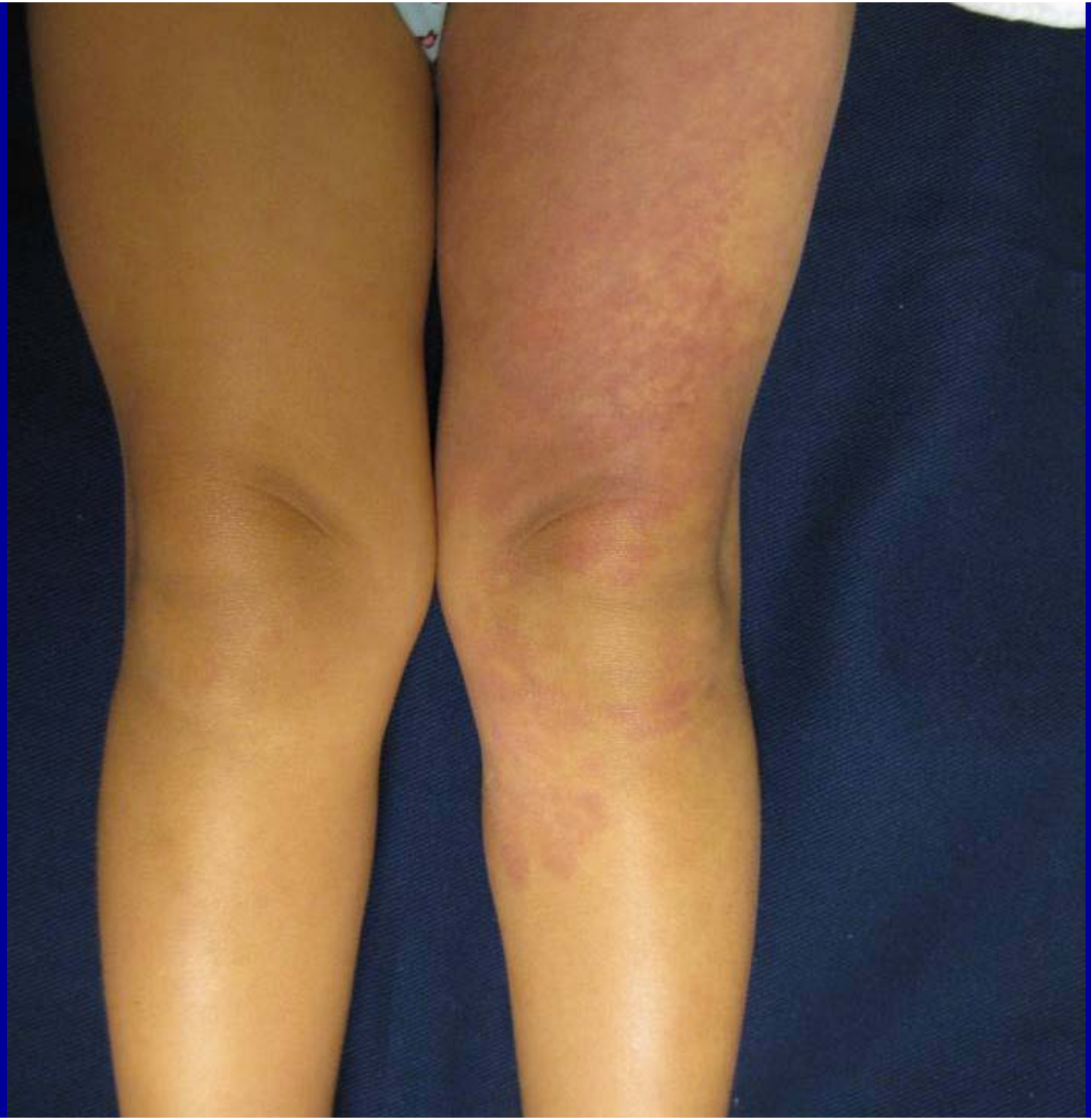

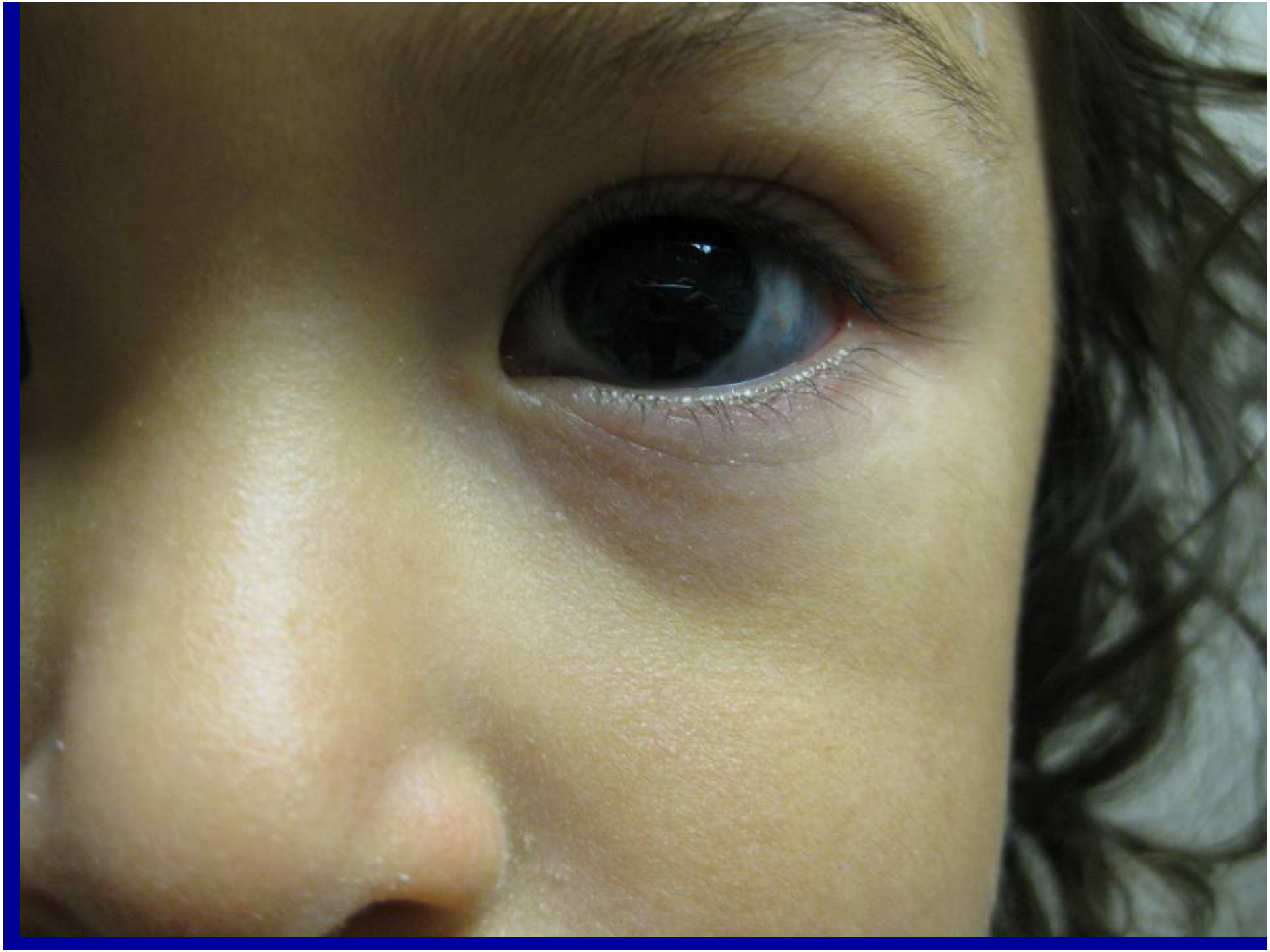

Case #34

SN

Cutaneous T-cell Lymphoma

CASE # 34
THE SOUTHEASTERN CONSORTIUM FOR DERMATOLOGY
October 16-18, 2009

Patient:	SN
Areas of Interest:	Face and chest
History:	50 year old black woman with an essentially asymptomatic eruption on her face and chest that began several years ago. Several biopsies in the past have been non-diagnostic. The cutaneous eruption seems to be slowly spreading, and the patient denies any systemic complaints.
Past Medical History:	Hypertension
Medications:	Hydrochlorothiazide
Laboratory:	ANA, CBC, CMP wnl
Physical Exam:	On her face, there are multiple scaly hyperpigmented papules and superficial plaques predominantly on the left. Periorally there is some fine scaling. On her upper chest, predominantly in the V of the neck there is a poikilodermatous, somewhat atrophic, hyperpigmented patch. This eruption appears to spare the sun protected areas of her body. No adenopathy.
Pathology:	There is slight epidermal acanthosis with overlying areas of focal parakeratosis. Within the acanthotic epidermis there is widening of intercellular spaces as well as basilar layer pigmentation. Filling and expanding the papillary and reticular dermis is an atypical and dense lymphoid infiltrate composed of a monomorphic population of monocytoid appearing lymphocytes. The cells show enlarged and angulated nuclei that demonstrate nuclear hyperchromasia with abundant pale cytoplasm. Lymphocytes line up along the basal keratinocytes in a linear fashion and show epidermotropism forming small Pautrier's microabscesses. Special Stains: CD3 positive, CD4 > CD8 positive, CD20 negative
Diagnosis:	Cutaneous T Cell Lymphoma (Mycosis Fungoides)
Treatment and Course:	This case of Mycosis Fungoides is unusual in its distribution and clinical appearance. Given its atrophic appearance and photodistributed location, a photosensitive dermatosis such as lupus or dermatomyositis was originally suspected. However, biopsies are more

consistent with CTCL. Thus far, because of her localized disease, topical corticosteroids have been prescribed alone, with the consideration of starting Plaquenil if it is unresponsive.

Comment:

Cutaneous T cell lymphomas represent 65% of all cutaneous lymphomas and are categorized by the World Health Organization-European Organization for Research and Treatment of Cancer. Mycosis Fungoides represents the most common form of CTCL and is characterized by a clonal expansion of T helper cells that classically evolve from patches into plaques and tumors. Classically, the lesions begin as nonspecific patches on the buttocks and lower trunk, and are immunophenotypically CD3+, CD4+, CD7-, CD45RO+, and CD8-, although aberrant phenotypes are not unusual. In the United States the incidence of CTCL is approximately 5 cases per million population per year, and is more common in elderly black men. Treatment for MF is typically stage dependant and is highly variable from institution to institution.

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