01

Treatment of Benign and Malignant Melanocytic Lesions With the Long Pulsed Alexandrite Laser

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Abstract: Epidermotropic metastases of melanoma has been well described in the literature. However, exclusive epidermotropic malignant melanoma metastases, without systemic involement, are rare. Herein, we present two cases of persistent epidermotropic malignant melanoma successfully treated with the 755 nm long pulsed (3 ms) alexandrite laser. Decreased tumor burden could theoretically decrease the risk for systemic metastasis. Furthermore, treatement results in improved appearance and enhanced quality of life. Salvage therapy with high energy pigment targeting lasers is an option for treating malignant melanoma.
Congenital nevi may be large and disfiguring. Treatment is aimed at minimizing the risk of malignancy, and obtain an acceptable cosmetic result. Total staged excision with tissue expanders has many potential drawbacks. Less invasive means of ablation using Q-switched ruby and alexandrite laser therapy have been documented in the literature. Here we present a case of a congenital nevus treated successfully with the 755 nm long pulsed (3 ms) alexandrite laser.

02

Verruciform Xanthoma of the Earlobe in an Immunosuppressed Patient

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Abstract: Verruciform xanthoma is an uncommon mucocutaneous lesion of uncertain etiology. Originally thought to be limited to the oral mucosa, subsequent studies have shown its occurrence in other mucosal, as well as non-mucosal sites. Histologically, verruciform xanthoma is characterized by subepithelial foamy histiocytes associated with papillomatosis, parakeratosis and dyskeratosis. Subepithelial foam cells are lipid containing non-Langerhans cell histiocytes. A variety of etiologic factors have been proposed without much consensus including infectious (both bacterial and viral), degenerative, reactive-reparative, inflammatory, metabolic, reactive-multifactorial and immunosuppressive. Verruciform xanthoma of the external ear is exceedingly rare; only two cases have previously been reported. Herein, we report the first case of verruciform xanthoma occurring on the earlobe at the pierced-site in a patient with immune suppression and discuss the possible pathogenetic mechanism(s).

03

Costello Syndrome: Report of a case and review of the literature

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Abstract: Costello syndrome is a rare, distinctive, multiple congenital anomaly syndrome, characterized by soft, loose skin, palmar and plantar creases, loose joints, distinctive coarse facial features, and skeletal and cardiac abnormalities. Affected patients have a predisposition to develop malignancy, developmental delays, and mental retardation. Cutaneous papillomas are a unique skin manifestation of Costello syndrome. We present a patient with Costello syndrome to promote awareness of this rare syndrome and also highlight associated cutaneous findings.

04

Subcutaneous Interferon alfa for the Treatment of Pseudolymphoma

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Abstract: Cutaneous pseudolymphoma (CPL) is defined as reactive lymphoid aggregates in the skin simulating cutaneous lymphoma. It is classified based on both the histologic pattern and the predominate infiltrate present, being either B-cell, T-cell or a mixed population. CPL almost certainly does not represent a single disease state, but rather reflects an exaggerated local immunologic reaction to a stimulus. Reported treatment options include antibiotics, intralesional and systemic corticosteroids, cryosurgery, local radiation or surgical excision. There have been two reports in the literature identifying improvement with intralesional interferon. Overall, interferons offer antiproliferative, anti-inflammatory and antiviral effects. Interferon-alfa is FDA approved for use in hairy cell leukemia, malignant melanoma, AIDS-related Kaposi's sarcoma, condyloma acuminata, follicular lymphoma, chronic hepatitis B and chronic hepatitis C. It is also used off-label for many other cancers including cutaneous T-cell lymphoma. Interferon-alfa has been shown to have a direct inhibiting effect on the release of cytokines by TH2 clones in patients with idiopathic eosinophilia, and can inhibit Interleukin-4 production and increase expression of class I major histocompatibility complex molecules thus promoting recognition by cytotoxic T-cells. Herein, we report the first case of a patient who had complete clearance of her pseudolymphoma after starting systemic interferon alfa. Further studies are needed, but in the event of idiopathic and recalcitrant cases of CPL, we feel that it would be reasonable to consider subcutaneous interferon as a first line treatment option.

05

Follicular Lymphoma with Cutaneous Involvement: A Case Report

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Abstract: It is often difficult to separate out cutaneous nodules of B and T cell lymphoid dyscrasia, primary cutaneous B and T cell Non-Hodgkin lymphoma (NHL) or systemic B and T cell NHL presenting in the skin. We report the case of a 31-year-old man who presented in 2008 with a 4-5 year history of a pruritic, gradually progressive nodular eruption of the face. Skin biopsy demonstrated an atypical $CD20^+$ $CD79^+$ CD30+ lymphocytic infiltrate and a clonal immunoglobulin heavy chain gene rearrangement consistent with follicular (B-cell) lymphoma. Although the lesion location and indolent course were suggestive of primary cutaneous follicle center lymphoma (CFCL), evidence of both peripheral and mediastinal lymph node involvement at presentation precluded this diagnosis. In addition, molecular studies demonstrated the presence of t(14;18), a translocation characteristic of systemic follicular B cell NHL but uncommon in primary CFCL. The patient remains in remission as of July 2010 after R-CVP. This patient illustrates the need for assessment of representative skin biopsies by light microscopy, immunophenotyping, gene rearrangement plus/minus genetic studies to confirm the diagnosis of a specific subtype of B cell NHL and the need for systemic evaluation to distinguish between primary cutaneous and systemic lymphoma with cutaneous involvement.

06

Agminated blue nevi arising within a congenital melanocytic nevus: Treatment with 755nm alexandrite laser

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PURPOSE: We evaluated the response to alexandrite laser in a patient with agminated blue nevi arising within a congenital melanocytic nevus.
PATIENT AND METHODS: A 34 year old presented with a large (30 x 15 cm) congenital melanocytic nevus, punctated with multiple dark blue papules, on the right arm. Histopathologic examination of a papule revealed a dermis with heavily pigmented, fusiform, dendritic melanocytes embedded in a sclerotic stroma, consistent with a blue nevus. A long-pulsed alexandrite laser with a wavelength of 755 nm and a pulse duration of 3ms was used to treat the lesion. The patient was treated at a fluence of 50 to 55 J/cm2 and a spot size of 8mm. Treatments were performed at 2 month intervals.
RESULTS: Flattening of the blue nevi was observed after 6 treatment sessions. Side effects included mild blistering and moderate scabbing.

CONCLUSION: The 755nm alexandrite laser is a safe and effective treatment for agminated blue nevi arising within a congenital melanocytic nevus.

07

Cutaneous T-Cell Lymphoma Mimicking Langerhans Cell Sarcoma

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Langerhans cell sarcoma (LCS) and Cutaneous T-cell Lymphoma (CTCL) share similar clinical and diagnostic features. As in our case, these similarities make differentiating CTCL from LCS can be a unique challenge.

An 85-year-old white male presented to our clinic with a 2 month history of an erythematous, brown ulcerated plaque with a violaceous rim on his right hip. Punch biopsy showed diffuse superficial and mid-dermal lymphocytic infiltrate and admixed histiocytic-appearing cells exhibiting marked nuclear atypia and pleomorphism with mitoses. These cells exhibited immunoreactivity for S-100, CD1a, Langerin, CD4, CD68 and CD117a. EM was negative for Birbeck granules. T-cell gene rearrangement studies were positive. With the above findings, we made the diagnosis of tumoral CTCL with non-specific immunohistochemical staining with langerin.

Langerin is thought to be exclusive for dendritic cells of epithelium as it is felt to co-localize with Birbeck granules, specifically in Langerhan cells. However, our case suggests that positive langerin staining is not as specific for dendritic cells of the epithelium as currently purported. Langerin, in fact, may stain the malignant T cells of CTCL in rare circumstances.

08

Exploring the Impact of Androgens on Syringomas

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Abstract: Syringomas are benign neoplasms of eccrine glands, classically appearing in a periorbital distribution in young Caucasian women. Progesterone is believed to play a role in the pathogenesis of these neoplasms; however, the role of androgens is not well established. This poster describes the first documented case of classic syringomas in an African American man.

Dihydrotestosterone, a potent form of testosterone, appears to be the main androgen regulating eccrine structures. This hormone also demonstrates considerable ethnic variability, with the highest levels being found in African American men. We propose that the higher baseline levels of androgens in African Americans may protect against these neoplasms. We evaluated our patient's hormonal status to ascertain whether an androgen deficiency could explain his presentation. We found his serum testosterone and DHT levels to be below normal. We theorize that the absence of elevated androgens normally found in African American men may explain the unusual presentation of syringomas in our patient. Further, we will discuss differences in androgen levels by race and age as support for the observation that syringomas occur with less frequency in certain ethnicities and age groups

09

Febrile Ulceronecrotic Mucha-Haberman Disease

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Abstract: A 21 month-old female was admitted to the hospital with a seven day history of fever, difficulty breathing, and rash. The eruption began on the trunk and progressed to involve the face, neck, and groin, with concomitant development of oral ulcers, bleeding gums, and laryngeal edema resulting in intubation. Physical exam revealed numerous erythematous plaques, some with dusky centers, involving the trunk and extremities, sparing the palms and soles. Oral exam was significant for ulcers on the buccal mucosa and edematous, friable gums. Differential diagnosis included Stevens-Johnson syndrome and erythema multiforme. Skin biopsy was suggestive of pityriasis lichenoides et varioliformis acuta (PLEVA). A diagnosis of Febrile Ulceronecrotic Mucha-Habermann Disease (FUMHD) was made. She improved with treatment of methotrexate, prednisone, and topical corticosteroids.

FUMHD is a rare variant of PLEVA which presents as necrotic papules which evolve into large hemorrhagic ulcers often with associated crusts, bullae, or pustules. Systemic manifestations are extensive with a potentially lethal outcome. This case highlights the clinical presentation, histology, and treatment options of FUMHD in order to better characterize this rare entity

10

A Unique Case of Dyschromatosis Symmetrica Hereditaria with Down Syndrome

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Abstract: Dyschromatosis symmetrica hereditaria (DSH) is rare pigmentary disorder characterized by an irregular hypopigmented and hyperpigmented macules on the face and distal extremities. In this poster, we report a case of a female with both DSH and Down Syndrome. We also discuss the numerous gene-mapping studies which have pinpointed the etiology of DSH to be most commonly due to mutations that occur in the RNA deaminase gene (ADAR1 or DSRAD) [3,5] as well as the possibility of a spontaneous ADAR1 gene mutation in our case [1,2,3,4,5,6]. Care must also be taken to differentiate cases of DSH from similar diagnoses such as Dyschromatosis universalis hereditaria (DUH), Xeroderma pigmentosum (XP) and Acropigmentatio reticularis (AR)[3].

1. Suzuki N, et.al., Mutation analysis of the ADAR1 gene in dyschromatosis symmetrica hereditaria and genetic differentiation from both dyschromatosis universalis hereditaria and acropigmentatio reticularis. J Invest Dermatol. 2005 Jun;124(6):1186-92.

2. Zhang XJ, et.al., Identification of a locus for dyschromatosis symmetrica hereditaria at chromosome 1q11-1q21. J Invest Dermatol. 2003 May;120(5):776-80.

11

Digital Ischemia as Presenting Cutaneous Finding in a Patient with Limited Scleroderma

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Abstract: In scleroderma, auto-antibodies against endothelial cells initiate vascular damage with eventual fibrosis. A spectrum of skin and systemic symptoms ensue. We present a case of a six year old Caucasian female with anti-centromere antibody (ACA) positivity (1:640) and acute onset of digital ischemia of several fingertips. Vascular stenosis and segmental vascular occlusion were documented on angiography. There was no history of Raynaud's phenomenon and no other systemic symptomatology. ACA positivity has been mainly linked to limited cutaneous scleroderma or CREST Syndrome. Digital ischemia, however, is a rare manifestation of limited cutaneous scleroderma, more often presenting in diffuse scleroderma with additional systemic symptoms. Various reported cases in adult patients have associated ACA positivity with digital ulceration in the absence of other skin or systemic disease; all of these patients had a prior history of Raynaud's phenomenon. Thus, this case is unique in that there was no antecedent history of Raynaud's phenomenon and this clinical presentation is not well documented in the pediatric literature.

12

Cutaneous Mastocytosis in the setting of Aggressive Systemic Mastocytosis and Urticaria Pigmentosa

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Abstract: Mastocytosis is a proliferation and accumulation of mast cells in one or more organ systems that is characterized by multiple clinical variants. The current World Health Organization subdivides mastocytosis into seven categories: cutaneous mastocytosis, indolent systemic mastocytosis, systemic mastocytosis with associated (clonal) hematological non-mast-cell lineage disease, aggressive systemic mastocytosis, mast cell leukemia, mast cell sarcoma, and extracutaneous mastocytoma. The most common clinical manifestation is the involuted hyperpigmented lesions of urticaria pigmentosa that often present in childhood and spontaneously resolve. Many cases of indolent systemic mastocytosis have cutaneous involvement, but aggressive systemic mastocytosis rarely presents with cutaneous manifestations. Aggressive systemic mastocytosis is characterized by significant organopathy due to mast cell infiltration. We report a case of a patient with a 15-year history of urticaria pigmentosa who presents with new-onset cutaneous lesions of aggressive systemic mastocytosis. After treatment with midostaurin, an oral multi-targeted kinase and c-kit inhibitor, the patient had a decrease in mast cell burden, decreased tryptase levels, improvement in gastrointestinal symptoms, and resolution of the cutaneous lesions of systemic mastocytosis.

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Systemic Lupus Erythematosus Presenting with Diffuse Purpra and a Platelet Count of 3000

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Abstract:A 35 year-old African-American female was transferred to MCG with diffuse purpura and a platelet count of 3000, presumed to have idiopathic thrombocytopenic purpura (ITP) refractory to IV steroids, IVIg, and platelet transfusion. Clinical exam revealed diffuse palpable, purpura on the face and extremities. Punch biopsy showed diffuse RBC extravasation, superficial dermal neutrophilic infiltration, and perivascular and endovascular fibrin deposition--suggestive of vasculitis. Lab results showed proteinuria, ANA titer at 1:640 speckled, positive anti-dsDNA, low C3/C4, and elevated CRP. Extensive subclinical interstitial lung disease was also found on further investigation, and a diagnosis of SLE was confirmed. Rituximab and thromboplastin were begun as an inpatient with minimal improvement in platelet count. Cyclophosphamide 100mg daily was then added to her regimen; platelet count had risen to 97000 upon discharge. She has continued to improve on follow-up visits. The initial presentation of SLE is known to vary greatly. Common presentations include fever, myalgias, arthralgias, malaise, and rash. One must keep a low diagnostic threshold to consider SLE in unusual presentations such as severe blood dyscrasias and palpable purpura. This case represents that although extremely uncommon, a presentation similar to ITP can be the initial presentation of SLE with significant internal organ involvement.

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Two Cases of Necrolytic Acral Erythema in the absence of Hepatitis C Seropositivity

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Abstract: Necrolytic acral erythema (NAE) is a relatively new disorder, first described almost two decades ago as a cutaneous marker for hepatitis C virus. The pathophysiology of NAE is poorly understood but may be related to an underlying systemic or cutaneous metabolic deficiency. NAE may be classified under an umbrella of metabolic skin disorders including acrodermatitis enteropathica, biotin deficiency, essential fatty acid deficiency, pellagra, and necrolytic migratory erythema. A subset of NAE patients clinically respond to oral zinc supplementation regardless of their serum zinc levels. Review of the English literature revealed at least three reported cases of NAE occurring in the absence of hepatitis C. We present two additional cases of NAE without an hepatitis C association. Both patients presented with predominantly acral psoriasiform eruptions; clinicopathological correlation of characteristic NAE histologic findings such as epidermal pallor in the upper third of the epidermis with the acral eruptions supported the diagnoses. Hepatitis panels, HIV serologies, and serum zinc levels were negative or within-normal-limits for both patients. One of the two patients responded to oral zinc supplementation. Characterization of the full spectrum of NAE in both hepatitis C seronegative and seropositive individuals awaits further collective clinical experience with this new disorder.

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Cutaneous Metastatic Adenocarcinoma of Cervix to the Scalp

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Abstract: Cutaneous metastasis in cervical cancer is an uncommon event with a poor prognosis. Scalp metastasis in cervical cancer is exceedingly rare with few cases described in the literature. Here we report a case of cutaneous metastasis of cervical adenocarcinoma to the scalp that was clinically suspicious for keratoacanthoma but whose diagnosis was revealed through histopathology. The features of all eleven documented cases of metastatic cervical carcinoma to the scalp are outlined. Though unusual, the possibility of cutaneous metastasis should be considered in patients presenting with new skin lesions and a past medical history of cervical cancer.

16

Case Report: Acquired Spiny Keratoderma of the Hands and Feet

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Abstract: A 63 y/o female presented with itchy lesions on her palms and soles which had been present for more than 10 years. There was no involvement of other body sites and she had no history of other skin diseases. She had a history of diabetes, valvular heart disease and congestive heart failure. No family members had a similar skin condition. Family history was strongly positive for colon cancer.
Examination revealed numerous 1-2 mm, spiky, keratotic papules in a on the palms, fingers and soles (Figures. 1 and 2). Dermoscopic examination revealed multiple yellowish-white, keratotic protrusions (Figure 3).
Histopathology revealed a prominent symmetrical column of keratin extending from the granular layer to project above the surrounding orthokeratotic stratum corneum. Parakeratosis was present within the column as well as underlying hypogranulosis (Figure 4).
The clinical-pathological correlation was consistent with Acquired Spiny Keratoderma. Several treatment regimens have been described including physical removal with a blade, pumice stone, or dermabrasion or topical treatments including urea, salicylic acid, retinoids, Vitamin A, 5-fluorouracil, and tacalcitol.[1-4] Regular physical examinations and screening tests to identify associated maligancies should be recommended as there have been numerous reports of different associated malignancies.[4,5]
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Dermoscopic and Reflectance Confocal Microscopy Features of Cutaneous Metastatic Melanoma

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Abstract: Background: Reflectance confocal microscopy (RCM) is a noninvasive imaging modality that allows visualization of "optical sections" of skin with cellular resolution approaching that of histopathology. RCM features of cutaneous melanoma metastases (CMM) have not been previously described.
Methods: Clinical, dermoscopic, and RCM images along with real-time video capture were obtained from CMM lesions which were then excised and submitted for pathologic review.
Results: Dermoscopy of 30 lesions revealed disorganized globular (7) or homogenous with atypical vasculature patterns (23). RCM revealed features similar to primary melanoma: keratinocyte disarray, multiple pagetoid dendritic cells, cerebriform nests, and severely distorted architecture. Video capture revealed dynamic vascularity, leukocytes rolling along vascular endothelium, and vascular-tumor interface.
Conclusions: Superficial CMM are accessible by RCM and demonstrate features similar to those of primary cutaneous melanoma. Disordered vascularity appears to be a common feature of CMM. Further investigation is needed to fully characterize the tumor and vascular patterns of superficial CMM seen by RCM with the goal of generating and validating a set of diagnostic criteria

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78 Cases of Vulvar Melanoma: The Duke Experience

Title: 78 CASES OF VULVAR MELANOMA: THE DUKE EXPERIENCE
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Abstract: Malignant melanoma is the most serious form of skin cancer, and vulvar melanoma has an especially poor overall prognosis--5-year survival rates have been cited between 8-55% (mean 36%). To examine prognostic factors influencing survival, the Duke Melanoma and Tumor Registry Databases were queried for patients with a diagnosis of vulvar melanoma between 1970 and 2009. 78 subjects (73 Caucasians, 3 African-Americans, 2 race-not-reported) with a mean age of 60.4 years were identified. Median follow-up time was 11.0 years with 49 subjects experiencing melanoma-related mortality. Cohort survival rates are as follows: 84.7% (1-year), 49.3% (5-year), and 28.8% (10-year). Breslow thickness, lymph node status, systemic therapy, and surgery were examined for differences in survival distributions using the logrank test. In general, survival was inversely correlated with Breslow depth, extent of nodal involvement, and provision of systemic therapy. A higher survival rate was observed among those who received wide local excision. In summary, subjects with thinner melanomas amenable to surgical resection had a better prognosis than those with more extensive, metastatic disease at presentation.

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Clinical and Dermoscopic Features of 78 Scalp Nevi in Children

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Abstract: Publications on the dermoscopic features of nevi on the scalp in children have been scarce. The purpose of this study is to establish the typical clinical and dermoscopic patterns of scalp nevi in children younger than 18 years old to help optimize clinical care and management. We obtained and reviewed (by Pigmented Lesions Specialist, KCN) clinical and dermoscopic images of 78 scalp nevi in 34 healthy Caucasian children. The main clinical patterns included solid-colored (47.3% brown and 32.1% pink), 16.7% eclipse, and 3.8% cockade nevi. The most common dermoscopic pattern of scalp nevi is the globular pattern (47.4%) followed by the complex (reticular-globular) pattern (20.5%). Perifollicular hypopigmentation (98.7%) is a hallmark feature of scalp nevi. The majority (29.8%) of scalp nevi are located on the vertex scalp. Dermoscopy is a valuable, non-invasive tool in the evaluation of cutaneous melanocytic lesions in children and may decrease the number of unnecessary excisions.

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Imaging of Vulvar Tumors with the Reflectance Confocal Microscope (RCM)

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Abstract: Lesions located in the vulvar area are often difficult to diagnosis and treat; this may be due to clinician and patient aversion toward biopsy procedures. Reflectance confocal scanning laser microscopy is a broadly used optical technique that permits the non-invasive acquisition of histology-like thin “optical sections” within living tissue without causing patient discomfort. Our goal is to describe the utility of RCM imaging in two patients with vulvar tumors. RCM images revealed a dense distribution of bright, refractile dendritic cells in a patient with vulvar melanoma in-situ and multiple large, dark round structures of irregular diameter and slightly oblong shape, consistent with Paget cells, in a patient with vulvar Extramammary Paget’s Disease. For both patients, RCM images provided delineation from affected and non-affected tissue. RCM imaging may provide more information to aid in the diagnosis of vulvar neoplasms.

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Folliculitis Associated with Non-Epidermal Growth Factor Receptor Inhibitor Chemotherapy

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Abstract: Chemotherapy-induced folliculitis has been traditionally associated with the use of epidermal growth factor receptor (EGFR) inhibitors, such as cetuximab, gefitinib, and erlotinib. During a clinical trial looking at chemotherapy-induced alopecia, 3 of the 47 women enrolled complained of developing a pruritic scalp rash after receiving chemotherapy. On examination, all three women were noted to have erythematous papules and/or pustules surrounding scalp follicular openings. None of the women received anti-EGFR agents as part of their treatment; one woman received TAC chemotherapy (cyclophosphamide + docetaxel + doxorubicin HCl) for invasive ductal adenocarcinoma of the breast, the second woman received carboplatin and paclitaxel for invasive serous ovarian adenocarcinoma, and the third woman received TC chemotherapy (cyclophosphamide + docetaxel) for invasive ductal adenocarcinoma of the breast. As folliculitis has not previously been reported with the chemotherapy agents used in the treatment of these women, these cases represent a novel report of chemotherapy-induced folliculitis in the setting of regimens without EGFR inhibitors.