IL36RN is associated with generalized pustular psoriasis. The defect appears to increase expression of IL-8, a neutrophil recruiter and a target of cyclosporine. Whether the IL36RN gene played a role in our cases is uncertain.

Poststreptococcal pustulosis is a rare neutrophilic disorder that occurs as a complication following streptococcal infections. We describe 2 cases in a mother and daughter that responded quickly to a 6-day regimen of low-dose cyclosporine. The rapid response suggests that short courses of cyclosporine may be used as an alternative to systemic corticosteroids in patients with PSP.

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In Vivo Imaging of Miliaria Profunda Using High-Definition Optical Coherence Tomography: Diagnosis, Pathogenesis, and Treatment

Miliaria profunda (MP) is a condition of sweat duct obstruction manifesting with generalized papular eruption and anhidrosis. Using imaging with high-definition optical coherence tomography (HD-OCT), we identified, in vivo, the depth of lesions in MP and the likely location of sweat duct obstruction. To our knowledge, this is the first time such an evaluation has been performed.

Report of Cases | Case 1. A man in his 30s presented with generalized anhidrosis for 5 months. When his body temperature was increased, “goose-bumps” appeared over his trunk and limbs, which resolved spontaneously within an hour of cooling. Findings of baseline skin imaging using HD-OCT (Skintell; Agfa Healthcare) were normal. However, after exercise testing extensive whitish papules appeared over his trunk and limbs (Figure 1). Starch-iodine testing revealed generalized anhidrosis.

Testing with intradermal carbachol, 0.01% at 1 mL (Miostat; Alcon Laboratories Inc), failed to stimulate sweat production in our patient, although it did stimulate sweat production in 5 age- and sex-matched controls. Visualization with HD-OCT was repeated at the previous locations after exercise testing, now with whitish papules present (Figure 2). Biopsy of a whitish papule revealed epidermal spongiosis and hyperkeratosis over the sweat orifice.
Aluminum hexachloride was applied to an age-, sex- and ethnicity-matched healthy volunteer’s chest, and serial HD-OCT images were taken daily. The aluminum salt was found in the sweat orifices in this healthy control, and underlying intraepidermal changes similar to the patient’s MP lesions developed.

Ultimately, isotretinoin, 10 mg 3 times per week, was prescribed for our patient. Three months later, a repeated exercise starch-iodine test revealed marked increased areas of sweating. Analysis of pixel color in pretreatment and post-treatment images revealed that hidrotic areas on the trunk (that were most important in lowering core body temperature) had increased from 8.4% to 92.6%.

Case 2. A man in his 20s developed MP as he was recovering from widespread pityriasis rosea. His HD-OCT findings were very similar to those in case 1. He opted for conservative management.

Discussion | The term miliaria profunda was coined to refer to the deeper level of sweat duct obstruction, at or below the dermoepidermal junction, in contrast to the superficial and deeper epidermis locations in miliaria crystallina and miliaria rubra, respectively.\(^2\) This location of sweat duct obstruction in MP was determined experimentally in a healthy volunteer whose terminal sweat ducts were destroyed using electrolysis.\(^2\)

Evaluation with HD-OCT in both of our patients localized the skin lesions to the epidermis. Dilated spiraling acrosyringium was identified, and the adjacent hyperrefractile (bright) substance likely represents macerated keratin (Figure 2), which can be correlated with the poorly demarcated whitish papules observed clinically and on dermoscopy. The lesions were also surrounded by a hyporeflective (dark) rim, which likely represents free fluid (water has a low refractive index) consequent to obstructed sweat outflow and correlating with the spongiosis observed in histologic analysis. This phenomenon was better observed via in vivo imaging than histologic analysis likely because of the dehydration process used in histologic slide preparation.

Aluminum hexachloride applied to a healthy volunteer to experimentally obstruct the sweat orifices produced HD-OCT changes similar to those seen in MP. In previous reports,\(^2-4\) surface hyperkeratosis and parakeratosis of acrosyringium have been noted, but these were postulated to be only an epiphenomenon. We also identified these hyperkeratotic plugs in our histologic analysis, and by correlating histologic with HD-OCT findings, we determined that the obstruction most likely occurred at the sweat orifices due to these hyperkeratotic plugs. Such plugs can occur subsequent to inflammatory skin diseases\(^5\) such as pityriasis rosea (case 2). Patient 1 was treated with isotretinoin because retinoids are known to reduce hyperkeratinization.\(^6\) The marked response to isotretinoin in patient 1 further supports the important role of hyperkeratinization in the pathogenesis of MP.

Miliaria profunda can be effectively diagnosed using HD-OCT. Contrary to current belief, the evanescent skin papules are intraepidermal, and hyperkeratotic plugs at the sweat orifices are the likely cause of obstruction. We hereby
propose that MP be renamed *miliaria alba*, following the pattern of the clinically descriptive names for *miliaria crystallina* and *miliaria rubra*.

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Birt-Hogg-Dubé Syndrome in an African Patient and a Novel Mutation in the FLCN Gene

Birt-Hogg-Dubé syndrome (BHDS), an autosomal dominant condition caused by mutations in the *FLCN* gene, is characterized by fibrofolliculomas, spontaneous pneumothorax, and renal cell carcinoma. Herein, we report the first case to our knowledge of BHDS in an African patient.

Report of a Case | A 53-year-old man from Somalia with a history of hypertension presented with multiple asymptomatic bumps on his face that appeared progressively over time. He denied having a personal or family history of similar lesions, renal cell carcinoma, spontaneous pneumothorax, or other pulmonary disease. Physical examination revealed diffuse flesh-colored, dome-shaped and pedunculated papules on the face, neck, upper back, chest, arms, and axillary folds (Figure 1). Histopathologic analysis of biopsy specimens taken from the left jawline (Figure 2) and left forearm were consistent with fibrofolliculoma and trichodiscoma, respectively. Genetic test results were positive for a novel nonsense heterozygous mutation p.Glu410Stop (E410X) in exon 11 of the *FLCN* gene. The patient was treated with shave removal of the lesions that were particularly disfiguring. At last follow-up, he was undergoing evaluation for renal and pulmonary disease.

Discussion | Birt-Hogg-Dubé syndrome was originally characterized by the triad of fibrofolliculomas, trichodiscomas, and acrochordons. The lesions usually appear after the second decade of life. Clinically, fibrofolliculomas and trichodiscomas are indistinguishable; both present as firm dome-shaped papules predominantly involving the face, scalp, and neck. However, skin findings may be absent in some patients. One of 2 major criteria proposed to diagnose BHDS is the presence of 5 or more fibrofolliculomas or trichodiscomas with 1 confirmed histologically.1

Histologically, fibrofolliculomas, trichodiscomas, and acrochordons were initially described as separate entities. However, many authorities now believe that all 3 are variants of a