Cyclosporine in the Management of Poststreptococcal Pustulosis

Poststreptococcal pustulosis (PSP) is a rare neutrophilic disease occurring after a streptococcal pharyngitis. In our report, we describe a mother and daughter who developed PSP and were successfully treated with low-dose cyclosporine.

Report of a Case | A woman in her 50s and her daughter, in her 30s, presented simultaneously with painful pustules on the palms (Figure, A) and a few on the soles. The eruption began 5 days after culture-proven group A streptococcal pharyngitis in the daughter and tonsillitis in the mother. Both were systemically well with no fever, arthropathy, or mucosal lesions. Both patients had already received a week of oral cephalexin therapy because pustules continued to develop. There was no medical or family history of psoriasis. The mother had type 2 diabetes mellitus.

Laboratory investigations revealed leukocytosis in the mother (leukocytes, 13.5 × 10^9/L; neutrophils, 9.1 × 10^9/L; eosinophils, 0.3 × 10^9/L) and daughter (leukocytes, 12.2 × 10^9/L; neutrophils, 7.2 × 10^9/L; eosinophils, 0.3 × 10^9/L) (normal leukocyte upper limit, 10.5 × 10^9/L). (To convert leukocytes, neutrophils, and eosinophils to cells per microliter, divide by 0.001.) Cultures of the pustules were sterile. There were no signs of renal involvement: urinalysis findings and serum creatinine levels were normal. Human leukocyte antigen typing and biopsy were not performed.

We elected to treat both patients with cyclosporine as an alternative to prednisone. This decision was based on the rapid action of cyclosporine, its direct suppressive effect on cytokines that influence neutrophils, and no hyperglycemic adverse effects when compared with corticosteroids. The patients were prescribed oral cyclosporine, 50 mg, twice daily for 3 days (approximately 1 mg/kg/d), then 25 mg by mouth twice daily for 3 days. Both patients reported marked reduction in pain within 48 hours and dramatic improvement of the lesions within 1 week (Figure, B). Because the lesions responded rapidly, we discontinued cyclosporine treatment after the initial course. There were no reported adverse effects of cyclosporine and no recurrences after 1 year of follow-up.

Discussion | Poststreptococcal pustulosis, aka pustulosis acuta generalisata, is a rare postinfectious disorder associated with streptococcal pharyngitis. To our knowledge, only 25 cases have been described worldwide. It typically presents with a symmetric eruption of sterile pustules predominantly on the hands and feet, though they can occur elsewhere. Laboratory abnormalities include leukocytosis and/or elevated acute-phase reactants. Both arthropathy and glomerulonephritis have been reported. Histopathologic examination reveals subcorneal collections of neutrophils.

Poststreptococcal pustulosis can resemble palmoplantar pustulosis, pustular psoriasis, acute generalized exanthematous pustulosis, pustular vasculitis, and subcorneal pustular dermatosis. These and other neutrophilic disorders have historically been treated with prednisone or dapsone. Our cases suggest a role for a short course of low-dose cyclosporine as an alternative to systemic corticosteroids for PSP and possibly related neutrophilic disorders. In a recent systematic review of chronic palmoplantar pustulosis, cyclosporine was effective in 48% of patients receiving 1 mg/kg/d compared with 19% receiving placebo (n = 58; P < .02) and 89% of patients receiving 2.5 mg/kg/d compared with 21% patients receiving placebo (n = 40; P < .001). While these results are promising, we cannot exclude spontaneous resolution.

Two published reports have found an association of HLA-B35 and HLA-A2; HLA-B35 has also been linked with pustular psoriasis. In addition, deficiency of the anti-inflammatory cytokine interleukin (IL)-36 receptor antagonist (gene,
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.

Figure 1. Generalized Anhidrosis on the Back of a Patient With Miliaria Profunda

A, Asymptomatic, nonfollicular whitish papules developed over the skin during active exercise. B, An admixture of starch-iodine powder was sprayed over the whole body after exercise using pressurized air through a spray gun, revealing generalized anhidrosis except for very limited areas (stained purple) on the lower back.

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.