Pruritic Photosensitive Rash

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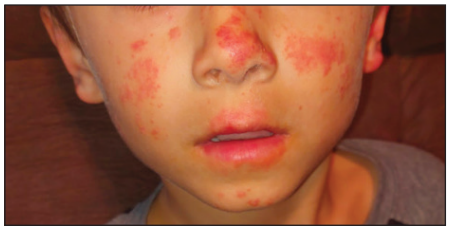

A 6-year-old boy presented via telemedicine for evaluation of a recurring rash that first presented on the face 9 months prior to presentation and waxed and waned throughout the fall and winter seasons for about 5 months. His mother noted that on a warm and sunny day 5 months after its first appearance, the patient was at a dog park and developed the rash on the face and hands—the only areas that had been exposed to the sun—later that evening. The patient reported pruritus but no associated burning or stinging. He was evaluated by an allergist 1 month later and was treated with oral cefazolin and hydrocortisone ointment 2.5% for suspected impetiginized dermatitis without improvement. The rash persisted until evaluation by our clinic 2 months later. Photographs showed erythematous scaly plaques and papules scattered on the cheeks, nose, upper and lower lips, and vermilion borders, as well as the dorsal aspect of the hands. He also had conjunctival erythema, which his mother reported was particularly worse in the summer months and associated with photophobia. His mother also noted increased tear production when in the sun. There was no mucosal involvement. The patient had no notable medical history and was not taking any medications. His mother had a history of polymorphous light eruption that recently was treated with hydroxychloroquine but without benefit.

WHAT'S YOUR **DIAGNOSIS?**

a. actinic prurigo
b. acute lupus erythematosus
c. allergic contact dermatitis
d. erythropoietic protoporphyria
e. polymorphous light eruption

PLEASE TURN TO PAGE E43 FOR THE DIAGNOSIS

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THE **DIAGNOSIS:** Actinic Prurigo

Actinic prurigo is an idiopathic photodermatosis triggered by UV exposure that primarily affects sun-exposed areas of the skin.[1,2] It typically presents as pruritic papules, plaques, and nodules, with most patients also experiencing oral tingling and pain.[3] In more severe cases, it can progress to include conjunctival disease, scarring, and cheilitis.[1] A study of ocular findings among children with actinic pruritus reported that photophobia was one of the most important features,[4] which was present in our patient. The face, especially over the zygomatic arches, nasal bridge, and lower lip, commonly is affected.[1] Secondary lichenification or eczematization may occur.[5] In our patient, the combination of conjunctivitis, cheilitis, and an eruption on sun-exposed skin were crucial in making the diagnosis.

Most cases present in patients younger than 10 years. It most commonly is seen in American Indians in North America, Central America, and South America.[2] After the diagnosis was considered in our patient, the family was asked about their ancestry and confirmed that both of the patient's maternal and paternal grandparents were of American Indian descent. There also is a strong genetic component; the HLA-DR4 allele variant is present in 90% of cases, especially DRB1*0407, which is seen in 60% of cases.[1,6] In our patient, testing revealed HLA-DR4, DRB1*04 positivity. We further hypothesized that his mother's photosensitive rash may have been actinic prurigo as opposed to polymorphous light eruption, which could explain the lack of response to hydroxychloroquine.

The diagnosis of actinic prurigo usually is made clinically. A skin biopsy typically is not necessary but would show hyperkeratosis, spongiosis, and acanthosis with a lymphocytic perivascular infiltrate. Biopsies of the lip classically show lymphoid germinal centers in the lamina propria, which can help distinguish actinic prurigo from polymorphous light eruption.[1]

In our patient, the differential diagnosis included polymorphous light eruption, connective tissue disease such as lupus erythematosus or dermatomyositis, porphyria such as erythropoietic protoporphyria, and allergic contact dermatitis. Polymorphous light eruption was ruled out by the oral and ocular manifestations, which are not atypical for this diagnosis. The patient's laboratory results displayed unremarkable antinuclear antibodies, creatine kinase, aldolase, and extractable nuclear antigens, which made connective tissue disease unlikely. Furthermore, a porphyria screen for total plasma porphyrins and whole blood protoporphyrin was negative, which helped rule out porphyria. Allergic contact dermatitis seemed less likely given the lack of improvement with topical steroids. Overall, the clinical presentation in a patient with relevant family ancestry and HLA-DR4 positivity supported a diagnosis of actinic prurigo.[7]

To manage the condition in our patient, strict photoprotection was recommended as well as the application of triamcinolone ointment 0.025% to the affected areas twice daily until the skin symptoms improved. For acute flares, other treatment considerations include topical tacrolimus, oral antihistamines, and oral corticosteroids. Some success has been reported with cyclosporine and azathioprine. For severe disease, thalidomide is the recommended treatment; it is effective in pediatric patients at dosages of 50 to 100 mg daily, but the dose has not yet been standardized for this age group.[8,9] Many adult patients initially are controlled with 100 to 200 mg daily, which can be tapered down to a dosage of 25 to 50 mg weekly with few adverse effects; however, the overall substantial side effects of thalidomide limit its use in both pediatric and adult populations.[1,2] Newer studies have suggested promising results with dupilumab, especially when actinic prurigo presents with high IgE levels or eosinophils on histology.[7,10] In our patient, the IgE level was normal.

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