

Images in Allergy

The Striking Appearance of a Rare Skin Condition: “Half-Half” Blisters (Subcorneal Pustular Dermatitis)

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A 70-year-old white man was referred to the allergy/immunology clinic because of a 1-year history of recurring pruritic lesions on the torso and extremities. The past medical history was significant for hypertension, hyperlipidemia, benign prostatic hyperplasia, and mild memory loss. No significant family history was reported. The skin examination revealed multiple vesicular/pustular lesions with clear fluid in the upper half and purulent (yellow) fluid in the lower half (“half-half” blisters) (Figure 1). The healing lesions showed postinflammatory hyperpigmentation. Before the visit, the patient was evaluated by a dermatologist and had a skin biopsy, which showed a cell infiltrate with lymphocytes, histiocytes, and eosinophils. The total IgE level was elevated, which prompted an Allergy and immunology referral. The rest of the laboratory workup, including complete blood cell count with differential, complete blood cell count, thyroid-stimulating hormone, and serum protein electrophoresis, did not reveal a cause for the symptoms. The patient was prescribed an oral corticosteroid taper and cephalexin for 14 days, which resulted in a complete resolution of his symptoms. However, symptoms recurred after the completion of therapy. The patient continued to be followed closely by his dermatologist.

The diagnosis is based on the striking clinical appearances of the lesions and the immunofluorescence studies. In most of the cases, subcorneal pustular dermatosis (SPD) histopathology is characterized by neutrophilic inflammation but strikingly in our case the patient presented with IgA pemphigus subtype usually present in a pemphigus-like intercellular pattern, either in the entire epidermis or confined to its upper layers. This explains the predominance of lymphocytes, histiocytes, and eosinophils in our patient who did have the IgA pemphigus subtype of SPD.¹ Although in our patient the total IgE level was elevated but it is a nonspecific finding for SPD. SPD has also been associated with IgA monoclonal gammopathy and myeloma.²⁻⁵ But in our case serum protein electrophoresis was normal in our patient with no M-spike.

Immunofluorescence studies for epidermal intercellular IgA staining should be performed periodically in patients with relapsing disease. Positive immunofluorescence studies may develop years after the initial diagnosis of SPD. Immunofluorescence studies help to identify patients with IgA pemphigus subtypes of SPD.³⁻⁵ IgA pemphigus subtype was eventually confirmed in our patient, 2 years after the initial presentation. SPD presents with intraepidermal deposition of IgA antibodies to structural proteins such as desmoglein 1. Desmogleins are structural proteins necessary for cell-to-cell adhesion.⁴⁻⁷ These are distinct from the desmoglein proteins targeted in other pemphigus variants, for example, pemphigus vulgaris (desmoglein 3 and 1) and pemphigus foliaceus (desmoglein 1).⁸ When these proteins are targeted, the structural framework linking skin cells together breaks down and vesicles or bullae are formed. Because desmoglein 1 is located in the stratum corneum of the skin, loss of function results in small superficial vesicles and pustules that desquamate.⁸

In most cases, SPD presentation is confined to the skin. However, SPD can be associated with other cutaneous and systemic conditions, such as Raynaud phenomenon, Sjögren syndrome, systemic lupus erythematosus, and neutrophilic dermatoses (eg, pyoderma gangrenosum and Sweet syndrome), paraproteinemias (predominantly IgA monoclonal gammopathies [monoclonal gammopathy of undetermined significance]), and multiple myeloma. In the presented case, serum protein electrophoresis was normal with no M-spike, and therefore, no evidence of monoclonal gammopathy of undetermined significance. Skin biopsy revealed a chronic inflammatory process with abundance of lymphocytes, histiocytes, and eosinophils. Immunofluorescence studies for epidermal intercellular IgA staining should be performed periodically in patients with relapsing SPD.¹ Positive immunofluorescence studies may develop years after the initial diagnosis of SPD. Immunofluorescence studies help to identify patients with IgA pemphigus subtypes of SPD. To emphasize the recommendation above, IgA pemphigus subtype was eventually confirmed via immunofluorescence in the presented patient, 2 years after the initial presentation. There are currently no official diagnostic criteria for SPD. The patient's symptoms resolved with the initial treatment of oral steroids. By the time the biopsy results confirmed SPD, the patient was already under the care of his dermatologist who decided not to pursue dapsone treatment. The patient had a transient remission of symptoms with an oral steroid taper, but SPD recurred on therapy completion. At the latest follow-up, 2.5 years after the presentation, the patient had no symptoms of SPD.

SPD is a rare disease entity with a visually striking clinical presentation of “half-half blisters.” Patients with SPD should be evaluated for associated diseases such as gammopathies. Although SPD is generally a chronic and relapsing condition, sustained remission is possible.

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FIGURE 1. Subcorneal dermatosis “half-half blisters.” Blisters contain both clear and purulent (yellow) fluid.

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