SHORT COMMUNICATIONS

Expanding Armor-Like Scales in a Middle-Aged Woman

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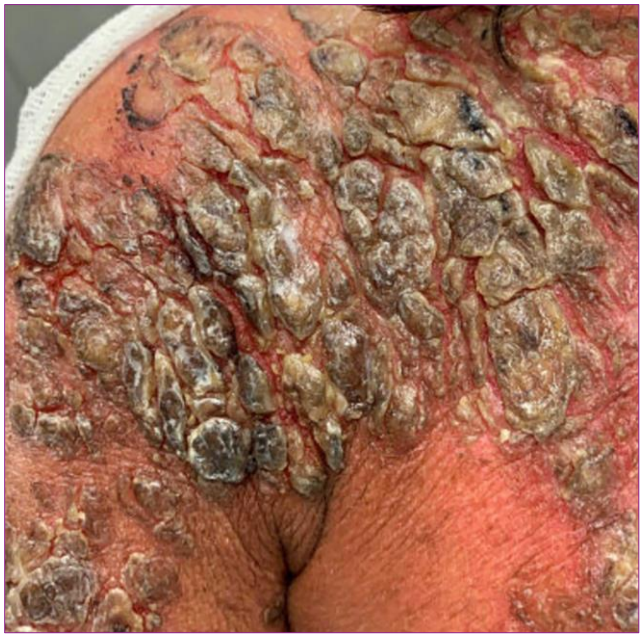
To the Editor: A woman in her 50s presented with a 7-month history of worsening pruritic papules and bullae on her face, trunk, arms, and axillae. Her medical history was significant for Meniere disease and hyperthyroidism. At an outside clinic, an initial skin biopsy from the arm showed intraepidermal acantholysis with dyskeratosis and she was diagnosed with transient acantholytic dermatosis (Grover disease). Treatments included triamcinolone ointment, doxycycline, antihistamines, and short courses of prednisone without clinical improvement. Over the following two months, her eruption worsened, and she developed painful oral mucosal erosions. Physical examination revealed vegetative scale-crusts overlying erosions on the face, arms, and chest in an armor-like pattern (Figure 1) as well as flaccid bullae on the back and erosions of the gingival and labial mucosae. A shave biopsy of the skin from the upper back was performed (Figure 2). Direct immunofluorescence showed intercellular epidermal deposition of IgG and C3. Indirect immunofluorescence was positive on monkey esophagus substrate in an intercellular pattern at a titer of 1:5120 and enzyme-linked immunosorbent assay (ELISA) quantified anti-desmoglein-1 (Dsg1) and anti-desmoglein-3 (Dsg3) antibodies at 620 (negative<20) and 176 units (negative<20), respectively.

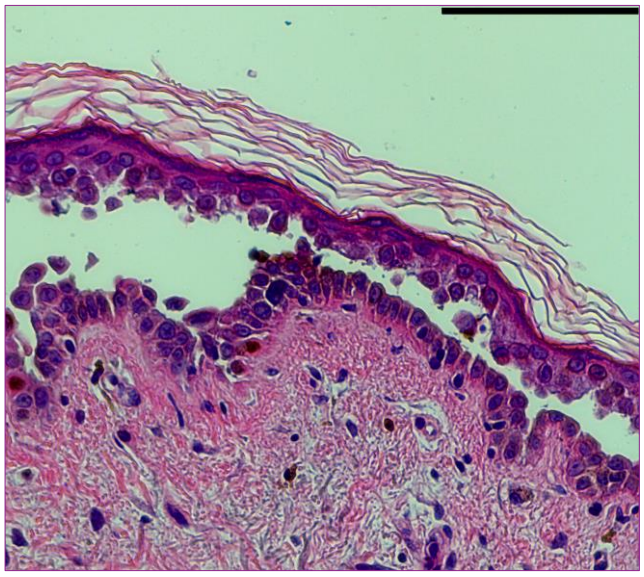
Figure 1. Armor-like vegetative scale-crusts on the right chest and arm.



Pemphigus vulgaris (PV) is an autoimmune blistering disease that affects stratified epithelia including the skin and mucosal surfaces. PV is characterized by loss of keratinocyte cell-to-cell adhesion secondary to circulating autoantibodies targeting the adhesive domains of desmosomal cadherins, Dsg1 and Dsg3. Binding of autoantibodies compromises tissue integrity, leading to intraepithelial acantholysis, which manifests in patients as bullae and erosions.[1] The diagnosis should be considered in patients with flaccid skin bullae, but PV may initially present with only

mucosal erosions or gingival sloughing. As in this case, vegetative scale-crusts can develop over long-standing erosions and, particularly when they are localized to intertriginous areas, the disease may be termed pemphigus vegetans.

Figure 2. Histopathology shows the characteristic tombstone pattern resulting from suprabasal epidermal acantholysis (H&E x40). Scale bar = 100 microns.



While a biopsy of Grover disease can show PV-like suprabasal acantholysis, mucosal erosions are not seen in this disease. Likewise, pemphigus foliaceus (PF) lacks mucosal involvement due to production of anti-Dsg1 but not anti-Dsg3 auto-antibodies[1; 2] and presents with more superficial skin erosions and crusting from subcorneal acantholysis. Bullous pemphigoid and linear IgA bullous dermatosis can feature both oral erosions and skin blistering, but they are more likely to produce larger and tense bullae due to sub-epidermal blister formation. Definitive diagnosis of PV depends on demonstration of typical suprabasal epidermal acantholysis along with detection of tissue-deposited or circulating auto-antibodies by immunofluorescence or ELISA, which measures antibodies recognizing Dsg1 and/or Dsg3.[2]

Treatment of PV is dictated by the extent of skin and/or mucosal involvement. Limited disease may be managed with high-potency topical corticosteroids while extensive disease necessitates initial high-dose systemic corticosteroids with transition to a steroid-sparing immunosuppressive agent.[2] Rituximab, an anti-CD20 monoclonal antibody, was approved by the U.S. Food and Drug Administration for moderate-to-severe PV.[3] Other off-label systemic agents that may be employed include mycophenolate, azathioprine, methotrexate, dapsone, and intravenous immunoglobulin.[2] Although relapse rates remain high in PV, rituximab can induce long-term remission[4] and ongoing clinical trials assessing comparative treatment efficacy should advance evidence-based practices for treating PV patients.

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