Epidermolysis Bullosa Acquisita: A case presentation

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Case Presentation

Chief Complaint: Sores on my body

History of Present Illness: Patient is a 54 year old thin Hispanic male who presents to clinic with complaints of sores on his body of a two year onset. These sores start off spontaneously as blisters on his chest, arms, and feet, and are somewhat painful. He denies any other systemic symptoms, and is otherwise healthy. His main concern is not being able to work due to the painful sores on his feet.

Past Medical History: None

Medications: None

Family History: Noncontributory

Social History: Lives at home with family, works in a factory, although has been unable to work since onset of this condition. Denies alcohol/tobacco/illicit drug use

Allergies: NKDA

Physical Exam: Multiple areas of sclerotic plaques in various stages of healing on forehead, chest, posterior upper extremities, dorsal feet, and bilateral toes, with some plaques exhibiting central ulceration and others with hyperkeratotic crust. No mucosal or fingernail changes noted.

Laboratory tests: CBC, CMP, ANA, HIV screen within normal limits

Histology:

H&E: ulcer with inflamed fibrosing granulation tissue

DIF: Subepidermal bulla with linear IgG and C3 along DEJ

IIF: IgG on dermal side of DEJ

Patient Course/Treatment: Considering laboratory, histologic, and clinical findings, the patient was diagnosed with Epidermolysis Bullosa Acquisita, inflammatory type. He was prescribed triamcinolone 0.1% ointment for active lesions as well as a tapered course of oral prednisone and referred to a tertiary center for further management.

Discussion

Epidermolysis bullosa acquisita (EBA) is an autoimmune mucocutaneous blistering disease which usually occurs in adults. The disease is characterized by the production of IgG autoantibodies which target type VII collagen, the primary collagen found in anchoring fibrils of the lamina densa (1,2). Anchoring fibrils serve to attach the epidermal basement membrane to the dermis (3), and the loss of these fibrils triggers an inflammatory cascade and activating complement. This leads to destruction of essential matrix proteins in the DEJ and causes the BP-like inflammatory EBA variant. Our patient correlates with this subtype of EBA, with lesions in a distribution beyond the characteristic sites of trauma. Most noninflammatory EBA patients also have mucosal involvement, and severe cases may be complicated by alopecia, nail dystrophy, or hand/finger fibrosis - the so-calledimoto deformity (4). Our patient did not have mucosal involvement, further aligning with inflammatory EBA.

The evaluation of EBA begins with a thorough history and exam of suspicious lesions and is confirmed with biopsy. A biopsy taken from lesional skin can reveal the subepidermal site of separation, with a cell poor infiltrate. The BP-like EBA displays a more robust inflammatory infiltrate, as consistent with our case. A biopsy taken from perilesional skin can be used for direct immunofluorescence, which will reveal deposition in the dermal-epidermal junction (DEJ) primarily of IgG as well as possibly complement. IgA, IgM, Factor B and properdin (1,2,4).

Additionally, indirect immunofluorescence may be performed on split skin separated at the lamina lucida of the basement membrane zone. In EBA, antibody deposition is seen most prominently on the dermal surface. This is in contrast with bullous pemphigoid or linear IgA bullous dermatosis, which contain antibodies primarily on the epidermal surface (2,4).

The treatment for EBA currently relies on pharmacotherapy, with variable results. Treatment should begin with agents that can be tolerated for chronic use with minimal side-effects. Colchicine is frequently used as a first-line therapy. Dapsone can also be used either alone or in conjunction with other drugs such as colchicine or corticosteroids. Prednisone (0.5 to 1.5 mg/kg per day) has been frequently used, though its side-effect profile and lack of efficacy limits its use to second-line or in combination with other therapies (4,5).

Immunosuppressive medications have been used in refractory disease, including rituximab, azathioprine, cyclophosphamide, mycophenolate mofetil and cyclosporine. These are often used in conjunction with steroids or other anti-inflammatory medications such as those described above (6). IVIG has been used for widespread or refractory disease, either alone or combined with other agents (7-9).

As with many chronic and relapsing dermatologic conditions, patient education is first and foremost in the management of EBA. Patients should be counseled to take precautions against minor trauma. They should cleanse their skin gently, avoid scrubbing, and seek medical treatment early should infection be suspected.

References