Drug-Induced Linear IgA Bullous Dermatosis

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Case Presentation:

Patient: J.T. is a 61 year-old Caucasian male.

History of Present Illness: Patient presented to Lehigh Valley Hospital with a several day history of a painful and pruritic rash that began in his inguinal creases and had progressed to his back, abdomen, buttocks, and upper and lower extremities. He was hospitalized 3 weeks prior for an acutely painful ischemic left foot and gangrenous first toe which resulted in femoral-peroneal bypass surgery. During that time he received numerous antibiotics including vancomycin, piperacillin/tazobactam, and levofloxacin. At the time of evaluation, vancomycin was the only antibiotic being given. He denied any history of a similar eruption in the past.

Medical History: Peripheral vascular disease, HTN, DM type 2, neuropathy, GERD, HSV1

Current Medications: Vancomycin, fluconazole, gabapentin, heparin, insulin (lispro and glargine), metoprolol, lisinopril, pantoprazole, aspirin

Physical Examination: Numerous shallow ulcers with surrounding erythema on the scrotum, penis, and perineum. Many flaccid vesicles and bullae, some arranged in linear, arcuate and polycyclic patterns on the trunk, buttocks and lower extremities. Small vesicles noted on the lips. Pink thin blanchable plaques scattered on the trunk and extremities.

Laboratory Data: CBC (8/29/12): WBC: 14.2 (4.5-11 thou/cmm), neutrophils 82% (40-70%), lymphocytes 9% (20-44%); Hgb: 11.2 (13.5-18.0 g/dL); Hct: 33 (40-54%); Pt: 459 (140-350 thou/cmm); CMP (8/29/12): WNL.

ANA profile, serum BP180 and BP230 autoantibodies, and serum transglutaminase IgA autoantibodies were ordered but not obtained by the patient.

Studies: Blood Cx (8/29/12): negative, MRSA screen (8/30/12): negative, HSV Cx IgG (8/30/12): negative.

Biopsy: Health Network Laboratories (S12-23570, 8/30/12)
Right posterior proximal arm: “Subepidermal bullous process with neutrophils lining up along the dermal-epidermal junction. Figures 1a-1c: Many vesicles and bullae. Some are arranged in arcuate and annular patterns, others in a more linear configuration, particularly adjacent to the scar from recent femoral-peroneal bypass surgery.

Images 2a-2b: Subepidermal bullous process with neutrophils lining up along the dermal-epidermal junction.

Discussion:

Linear IgA bullous dermatosis is an uncommon autoimmune blistering disorder in which IgA antibodies are directed against antigens in the basement membrane zone (BMZ) of the skin and mucosa. The most commonly identified antigens are proteolytic cleavage products of the hemidesmosomal protein BP180 and include a 97-kDa (LABD-97) antigen and 120-kDa (LAD-1) antigen. IgA binding to these sites, subsequent complement activation, inflammatory cell recruitment, and release of proteolytic enzymes lead to disruption of the BMZ and ultimately epithelial-dermal detachment. The resulting clinical lesions appear as clear or hemorrhagic vesicles and bullae of varying sizes that may coalesce to form annular or polycyclic arrangements – the so-called “cluster of jewels” pattern. Pruritic urticarial plaques may occur alongside the vesiculobullous lesions.

Linear IgA may be idiopathic or secondary to precipitating factors including drugs and, less frequently, systemic disease, trauma, or ultraviolet light. The drug-induced form is reported to occur anywhere between 24 hours to 29 days following administration of the medication. Vancomycin is the most commonly associated drug followed by amiodarone, diclofenac, captopril, clofazimine, and piroxicam. The pathophysiologic mechanism behind drug-induced linear IgA is unclear but it is suggested that the offending medication may cross-react with autoantigens of the BMZ and change their conformational structure or perhaps unmask antigens previously hidden from the immune system[1].

Biopsies of early lesions reveal subepidermal bullae with a predominantly neutrophilic infiltrate. Direct immunofluorescence (DIF) facilitates the diagnosis by demonstrating a linear pattern of IgG deposition along the BMZ. This is in contrast to the granular deposits of IgA seen in the papillary dermis in dermatitis herpetiformis. Deposits of C3 complement accompany IgA in most instances. Linear IgG or IgM deposits are also commonly involved along the BMZ. One study reported that IgG, IgM, and complement were seen in 20/31 cases[2]. Because bullous pemphigoid (BP) may also show linear IgA, C3, and IgG on DIF, differentiation can be problematic. BP usually involves a higher concentration of IgG over IgA and a more eosinophilic infiltrate. In these cases, clinicopathological correlation is necessary.

Drug-induced linear IgA bullous dermatosis has a very good prognosis, typically resolving within 2-6 weeks after discontinuation of the offending medication. Treatment involves topical corticosteroids in mild disease either as a sole agent or an adjunct to systemic therapy. The first line systemic agent for more involved disease is dapsone followed by sulfapyridine. Systemic corticosteroids and other immunosuppressive agents such as mycophenolic acid and cyclosporine can be helpful in patients not adequately controlled with first or second line medications. Dicloxacillin, erythromycin, and trimethoprim-sulfamethoxazole have been shown to be highly effective as well. Ulcerations may be treated with topical barrier creams and emollients to expedite healing.

References: