Bullous Pemphigoid

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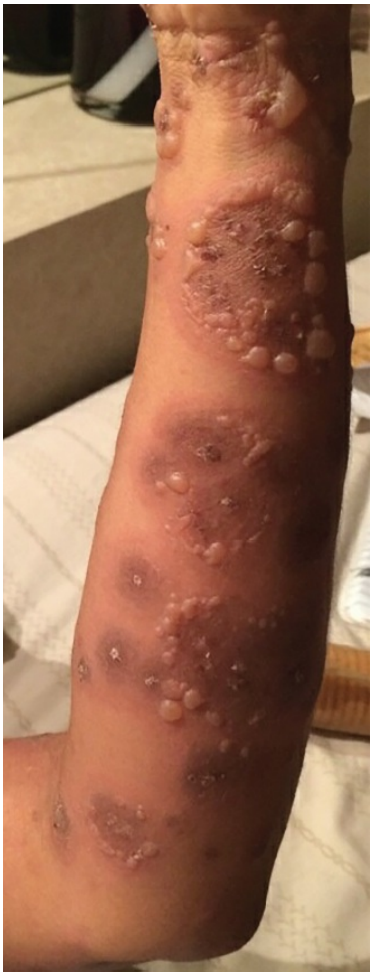
A 42-year-old woman with no past medical history and no medication use, who had undergone bilateral saline breast implantation with 5 subsequent repairs, presented 10 days after the last repair procedure with diffuse, blistering, pruritic and painful lesions on all surfaces (**Figure**). She was hospitalized and started on intravenous immunoglobulin and high doses of prednisolone. She then developed corticosteroid-induced hyperglycemia and was started on insulin.

After discharge, the woman's lesions gradually resolved, and the prednisone was tapered, resulting in normalization of glucose levels. Five months later, however, the pruritic lesions recurred extensively, and she was rehospitalized. The lesions again resolved after 4 days of similar treatment.



Physical examination. The patient had watery and painful lesions all over her body. Vital signs included the following: blood pressure, 114/82 mm Hg; pulse, 80 beats/min; respiratory rate, 17 breaths/min; height, 152.5 cm; weight, 52.7 kg; temperature, 37.8°C; and oxygen saturation, 100% on room air.

Diagnostic tests. A biopsy of the lesions was performed. Based on the biopsy results and clinical presentation, a diagnosis of bullous pemphigoid was made.

Discussion. Pemphigoid vulgaris is an autoimmune blistering condition involving immunoglobulin G (IgG) antibodies to the desmosomes of the stratum spinosum. Blister formation occurs between the stratum spinosum and stratum basale, creating thin, easily ruptured lesions with a positive Nikolsky sign. The mean age of onset is between 40 and 60 years.[1]

Bullous pemphigoid is another condition that also involves IgG antibodies, with mean age of onset at 80 years. Antibodies are formed against the subepidermal hemidesmosome, a component of the basement membrane zone adhesion complex, inducing damage via complement activation after binding to desmosomal proteins BP180 and BP230, causing neutrophil and eosinophil accumulation and proteinase release.[2] Conditions associated with bullous pemphigoid include multiple sclerosis, epilepsy, dementia, cardiovascular disease, and Parkinson disease. Medications such as gliptins, metformin, spironolactone, neuroleptics, and furosemide have been reported as the etiology in several case-control studies.[3] Additionally, rare cases of bullous pemphigoid have been seen following various surgical procedures, such as split-skin grafting.[4]

Clinical presentation of bullous pemphigoid involves pruritus or urticarial plaques that may precede the formation of tense bullae by weeks or months. The blisters may be widespread or localized and also may involve the oral or genital mucosa. Diagnosis is obtained with direct immunofluorescence and linear deposition of IgG and/or C3 along the basement membrane zone. Other immunoglobulins are less common but may also be seen. Enzyme-linked immunosorbent assay can also be utilized to detect antibodies to BP180 and BP230, with BP180 being more sensitive.[5]

Bullous pemphigoid is rare, with studies in Europe estimating an incidence of 6 or 7 cases per 1 million persons per year.[6] Few studies have been conducted in the United States, but incidence is estimated to be similar.[6]

This recurrent autoimmune skin disorder can be life threatening and requires lifelong corticosteroid or immunomodulator treatment. There is no evidence in the literature that saline breast implants are linked to bullous pemphigoid. However, due to the proximity of the surgical procedure in our patient, it is interesting to consider the possibility that breast implant surgery may have triggered this recurring autoimmune condition. It is uncertain whether removal of the implants will prevent future recurrence of the condition, but this is under consideration by the surgeon in this case.

References:

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