Crescentic Glomerulonephritis in a Patient with Epidermolysis bullosa acquisita

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Dear Sir,

Epidermolysis bullosa acquisita (EBA) is a nonhereditary, autoimmune, adult-onset chronic subepidermal bullous disease with recurrent posttraumatic cutaneous blister formation and linear deposition of IgG along the basement membrane zone (BMZ) of the skin. EBA has been reported to occur with many systemic diseases usually of an autoimmune nature including inflammatory bowel disease, systemic lupus erythematosus, chronic thyroiditis, cryoglobulinemia, and vesicular cystitis [1]. In this report, we describe for the first time, to our knowledge, a patient with EBA who developed crescentic glomerulonephritis.

A 66-year-old woman presented in November of 1988 with a history of recurrent bullous lesions in both arms and feet for the preceding 6 months. A skin biopsy showed chronic changes including lymphohistiocytic perivascular infiltrates in the dermis and mild hyperkeratosis with linear deposits of IgG along the BMZ. Her serum, however, was negative for antibodies to the BMZ. The clinical presentation, the absence of family history, and the skin biopsy findings were compatible with the diagnosis of EBA.

She was noted to have microhematuria with over 50 RBC/high power field, a few granular casts, and proteinuria (1.2 g/day). Serum creatinine was 97.1 $\mu\text{mol/l}$. A renal biopsy revealed large cellular crescents in 4 of the 30 glomeruli studied with linear deposits of IgG and complement (C3) along the glomerular capillary walls by immunofluorescence microscopy. Her serum however was negative on an ELISA assay for anti-glomerular basement membrane (GBM) antibodies.

Because of lack of response to prednisone and dapsone, cyclophosphamide (150 mg, p.o. daily) was given for 1 year. Her cutaneous lesions responded dramatically and microhematuria stopped. The serum creatinine was stable at 132.5 $\mu\text{mol/l}$ in 1993 and she has had no skin blisters.

It is unclear why some patients with EBA develop a systemic illness with involvement of different organs. The EBA antigen has been identified as the carboxyl terminal of type VII collagen present in the BMZ [2]. Paller et al. [3] have documented the presence of EBA antigen in oral mucosa and esophagus but not in the kidney, urinary bladder or blood vessels. Circulating antibodies against BMZ have been noted in 22-88% of patients [1]. In some instances at least, antibody cross-reactivity may explain involvement of organs other than the skin in patients with EBA. Alternatively, patients could develop different antibodies resulting

in cutaneous and renal disease. In support of this is the observation of an increased incidence of HLA-DR2 allele in patients with EBA or bullous SLE or anti-GBM antibody mediated crescentic glomerulonephritis [4].

References

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