AUTOIMMUNE BULLOUS DISEASES

A CASE OF PARANEOPLASTIC EPIDERMOLYSIS BULLOSA ACQUISITA?

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Background: A 58-year-old Greek female presented with a 5-year history of a widespread mucocutaneous blistering eruption. She had been treated for 4-years with clobetasol ointment and oral corticosteroids and had developed steroid-induced diabetes mellitus and suspected adrenal insufficiency. Her condition remained poorly controlled. She had widespread erosions, tense bullae, post-inflammatory hyperpigmentation and scarring with milia. There were erosions in the mouth and she gave a history of dysphagia. Our differential diagnosis included mucous membrane pemphigoid, epidermolysis bullosa acquisita and porphyria cutanea tarda.

Observation: Bloods showed an elevated LDH 635U/L and protein 89g/L (negative monoclonal banding) with an 8am cortisol of 23nmol/L. Urine demonstrated significant proteinuria with a urine protein:creatinine ratio of 150mg/mmol. She had a negative porphyrin screen and her ANA was speckled 1:80 with an atypical ANCA.

Skin biopsies identified subepidermal bulla formation, an epidermal and upper dermal neutrophilic infiltrate and rare eosinophils. Direct-immunofluorescence demonstrated IgG and C3 deposition along the basement membrane zone.

She was commenced on dapsone and prednisolone was tapered to 10mg. She remained very symptomatic and continued to develop new blisters with her options for further steroid-sparing agents limited by her suspected renal disease and pending imaging. She was commenced on monthly intravenous immunoglobulin for three days. Initially she improved but subsequently continued to flare requiring temporary increases in her steroid dose.

Indirect-immunofluorescence was sent for salt-split skin examination which identified a linear deposition along the dermal side of the basement membrane. Type VII collagen antibodies were sixteen times normal.

Key message: A diagnosis of epidermolysis bullosa acquisita was made on the basis of the international bullous diseases group consensus on diagnostic criteria. Imaging revealed diffuse intrahepatic, extrahepatic and pancreatic ductal dilatation and a solid left renal lesion concerning for malignancy. She is awaiting investigation with an endoscopic ultrasound.

Could this be a paraneoplastic EBA?