AUTOIMMUNE BULLOUS DISEASES

EPIDERMOLYSIS BULLOSA ACQUISITA MIMICKING BULLOUS PEMPHIGOID TYPICAL HISTOPATHOLOGY: A GOOD CLINICAL RESPONSES TO METHOTREXATE

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Background: Epidermolysis bullosa acquisita (EBA) is a rare and chronic bullous subepidermal disease which develops on the skin and mucosas and initiates frequently in adulthood. The etiology is related with antibodies against type VII collagen. Treatment is usually disappointing.

Observation: A 72 year-old man presented with a 4 months history of tense blisters and erythema on his trunk and extremities that left milia after healing. His family members had no similar conditions. No mucosa lesions were observed. Laboratory studies revealed high levels of thyrotropic hormone and anti-thyroperoxidase antibody. The biopsy revealed subepidermal blister formation in the papillary dermis with eosinophils' infiltration. Direct immunofluorescence showed immunoglobulin (Ig)G and C3 along the basal membrane zone and salt split skin technique showed immune complex deposition in the blister roof. The patient was diagnosed with Epidermolysis bullosa acquisita coexisting with Hashimoto's thyroiditis and was treated with prednisone (60 mg daily) and topical betamethasone for one month with great clinical improvement. Despite slow withdrawal of corticosteroid therapy, there was lesions' recurrence. It was introduced dapsone (200 mg daily) and tetracycline (2g daily). The screening to immunosuppressive therapy showed the purified protein derivative (PPD) positive (21mm) but the thorax computed tomography was normal. He had a 6 months latent tuberculosis treatment with isoniazid. After three months of tetracycline, it was suspended because there was no clinical response. Azathioprine (150 mg daily) was used for the following three months persisting with poorly controlled disease. Later, it was introduced methotrexate (10 mg weekly), colchicine (1,5g daily) and pentoxifylline (800 mg daily) with maintained remission.

Key message: EBA often presents with neutrophils and plasmocitos bullae infiltration and the best responses are with systemic corticoid therapy, differently of our patient wich biopsy resembled bullous pemphigoid wich is rich in eosinophils and was controlled with methotrexate.