AUTOIMMUNE BULLOUS DISEASES

EPIDERMOLYSIS BULLOSA ACQUISITA: A DIAGNOSTIC DILEMMA.

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Background: Epidermolysis bullosa acquisita (EBA) is a rare acquired autoimmune mechanobullous disease with diverse cutaneous affectation. Its exact aetiology remains elusive; however an association of autoantibodies targeted against collagen 7, which maintain adhesion between the basement membrane and the dermis, has been proposed.
Epidemolysis bullosa acquisita is rare; occurring in about 0.2 cases/ million people/ year. Common in middle life; mostly in African-Americans with no sex preponderance.
Diagnosis is confirmed by immunofluorescence microscopy (direct and indirect) which may not be readily available/accessible. Treatment options are variable.

Observation: A middle-aged Nigerian female presenting with widespread tense blisters, initially localized to mucosa - mouth and eyes; later extending to trauma-prone areas, fingers and scalp with secondary generalizations to trunk and skin folds.
No associated pruritus or urticaria-like lesions. Rash was not photo distributed. No associated hirsutism or features suggestive of SLE. She never had similar lesions in her childhood.
Skin biopsy revealed subepidemal blisters with neutrophil clusters. ANA and anti-dsDNA titres were not significant.
Investigation to confirm the diagnosis could not be done due to the scarcity and cost of immunoflourescence in our setting.
Case was initially managed with high dose steroids and azathioprine without success. Diagnosis was however reviewed to EBA (mixed type; which is rarely seen) and azathioprine was substituted with dapsone with significant and remarkable improvement and no relapses thus far.

Key message: Clinical diagnosis of EBA is difficult as a result of its similarity in presentation with many immunobullous disorders. Immunofluorescence microscopy to confirm diagnosis is not readily available especially in resource poor settings. A high index of suspicion and appropriate treatment may offer good prognosis. Index case was managed with dapsone with excellent outcome.