

## SHORT COMMUNICATIONS

### Pediatric Inflammatory Epidermolysis Bullosa Acquisita in a Patient with Oculocutaneous Albinism

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A 15-year-old Caucasian male with a history of oculocutaneous albinism (OCA) was referred to our dermatology clinic for a 4-year history of erythematous plaques and clear-yellow vesicles and bullae on his face, neck, upper chest, and extremities. The vesicles previously erupted once to twice a year, but recently have occurred more frequently and become less responsive to oral and topical steroids. During an acute episode, he would develop either a single or numerous, pruritic blisters on his face, trunk and upper extremities. The patient and his mother were unable to identify any triggers and due to the patient's history of OCA, he practiced strict photoprotection. Review of systems was negative and he denied any ocular or mucosal lesions. The patient's medications were mometasone furoate/formoterol fumarate dihydrate, cetirizine, and montelukast. Physical exam was remarkable for scattered erythematous scaly papules and polycyclic plaques on the chest, upper extremities, face and hands, as well as a few intact clear-yellow vesicles. Punch biopsies of the left chest and right arm were performed and histopathology revealed subepidermal bullae formation with neutrophils and a few eosinophils (Figure 1a). Direct immunofluorescence (DIF) of the

left chest punch biopsy was positive for linear and granular deposition of IgG, IgA, IgM, C3 and fibrin along the dermal-epidermal junction (Figure 1b). Serologic testing for complement fixing antinuclear antibodies (CANA), ANA, Ro (SS-A) and La (SS-B), anti-smith, anti-dsDNA, anti-RNP, anti-cardiolipin were negative; however, type VII Collagen antibodies were detected by ELISA at 7.6 U/mL. Complete blood count, CMP, ESR, CRP and complement levels were all within normal limits. A preliminary diagnosis of inflammatory epidermolysis bullosa acquisita (EBA) was made and the patient was started on dapsone 50 mg daily and prednisone 30 mg in conjunction with rheumatology. The patient's blisters resolved almost completely within several days with no residual scarring or milia formation.

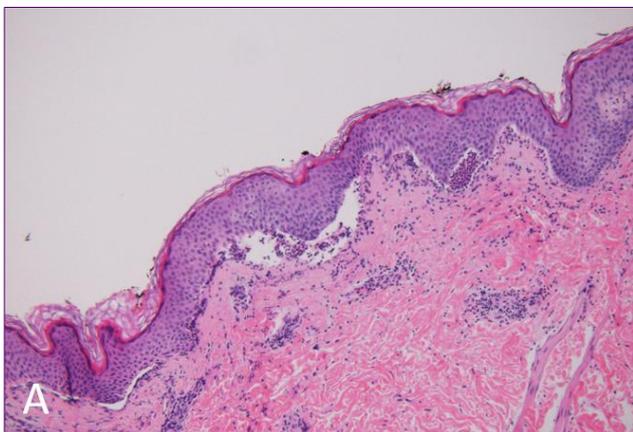
Epidermolysis bullosa acquisita (EBA) is rarely seen in the pediatric population and can have various clinical presentations. In addition to clinical manifestations of the disease, serologic, histologic and immunofluorescence findings are used to make the diagnosis. This case describes an unusual instance of pediatric inflammatory epidermolysis bullosa in a patient with

oculocutaneous albinism and multiple positive DIF reactants.

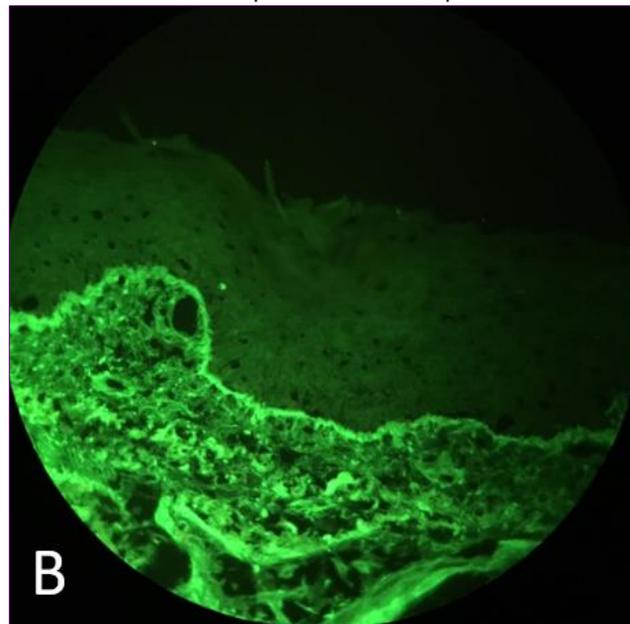
While deposits of IgG, IgA, IgM and C3 along the basement membrane zone (BMZ) have been reported previously, to our knowledge this is the first case of pediatric inflammatory EBA to demonstrate positivity of all the above reactants.<sup>1,2</sup> This pattern is more commonly found in cutaneous lupus, and while a diagnosis of bullous lupus was considered, this patient had no evidence of systemic lupus. However, this finding does suggest potential overlap in pediatric inflammatory EBA and bullous lupus.

UV radiation can cause damage to the BMZ and increase the transcriptional expression of type VII collagen antibodies in dermal fibroblasts.<sup>4,5</sup> We hypothesize that due to his history of OCA, our patient's increased sensitivity to solar radiation led to the exposure of BMZ antigens and development of type VII collagen autoantibodies. Ultimately, this resulted in the clinical manifestation of inflammatory EBA.

**Figure 1.** (A) Mid-chest with erythematous macules and bullae.



**Figure 1.** (B) H&E, 10x magnification biopsy specimen demonstrating subepidermal bullae formation with neutrophils and eosinophils.



**Conflict of Interest Disclosures:** Dr. Orlowski was active duty Air Force at the time of submission. The views expressed are those of the authors and are not to be construed as official or as representing those of the US Air Force or the Department of Defense. Dr. Orlowski was a full time federal employee at the time portions of this work were completed. They are in the public domain.

Dr. Hatch, Dr. Kissel, and Dr. Pavlidakey have no conflicts of interest to declare.

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