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Linear Immunoglobulin A (IgA) Bullous Dermatosi s in a Child

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Case Presentation

Patient: 8 year-old African American female.

History of Present Illness: The patient presents with a diffuse painful and pruritic bullous eruption. Approximately five days prior to the onset of the eruption, she had “hives” on her arms and symptoms of an upper respiratory infection including: myalgias, rhinorrhea, cough, and pharyngitis. She was afebrile and did not seek medical care at that time. She took guaifenesin and ibuprofen. She then developed blisters on the lower extremities and forehead. She had presented to the primary care physician and was started on empiric systemic corticosteroids, diphenhydramine, acetaminophen, and albuterol for presumed allergic reaction. She did not improve and the eruption continued to progress. Review of systems was positive for dysuria, eye pain, myalgias, rhinorrhea, and unproductive cough. She was treated with prednisolone and after ten days of treatment, we noted significant improvement in pain and pruritus as well as partial re-epithelialization of denuded lesions.

Medical History: Measles

Social History: She lives at home with her parents and two younger siblings; attends public school. No travel history. There are no pets in the home.

Current Treatment: Prednisolone taper starting at 40 mg daily, mupirocin 2 % ointment, hydroxyzine, diphenhydramine, ocular preparation of erythromycin

Physical Examination: Multiple tense vesicles and bullae (approximately 1-3cm in diameter) coalescing on the forehead, back, and buttocks. Multiple pink papules coalescing into thin plaques on the abdomen and bilateral upper and lower extremities. Two faint pink macules on the right palm. There is periocular and bulbar injection as well as crusting of the right nostril.

Laboratory Data: Staphylococcus aureus positive culture from right naris, Respiratory viral panel is positive for rhinovirus and enterovirus and negative for various strains of influenza, parainfluenza, coronavirus, metapneumovirus, adenovirus, bocavirus, chlamydomphila, and mycoplasma pneumoniae. VZV DFA, HSV/VZV cultures, ASO titer, ANA, anti-dsDNA, anti-SSA, anti-SSB, SM/RNP, anti-Smith, anti-SCL-70, CBC, CMP, and urinalysis were all WNL.

Biopsy: *Health Network Laboratories* (S16-37681, 10/20/16) 1) Back (vesicle): “Subepidermal vesicular dermatitis with neutrophils and eosinophils.” 2) Back (papule): “Early subepidermal vesicular dermatitis with neutrophils and eosinophils.” 3) Back-DIF: “Strong continuous linear basement membrane zone with IgA, weak patchy granular basement membrane zone with C3.”



Figure 1: Clinical image of patient's face demonstrating denuded bullae on the forehead.



Figure 2: Clinical image of patient's back demonstrating multiple coalescing tense vesicles and bullae.

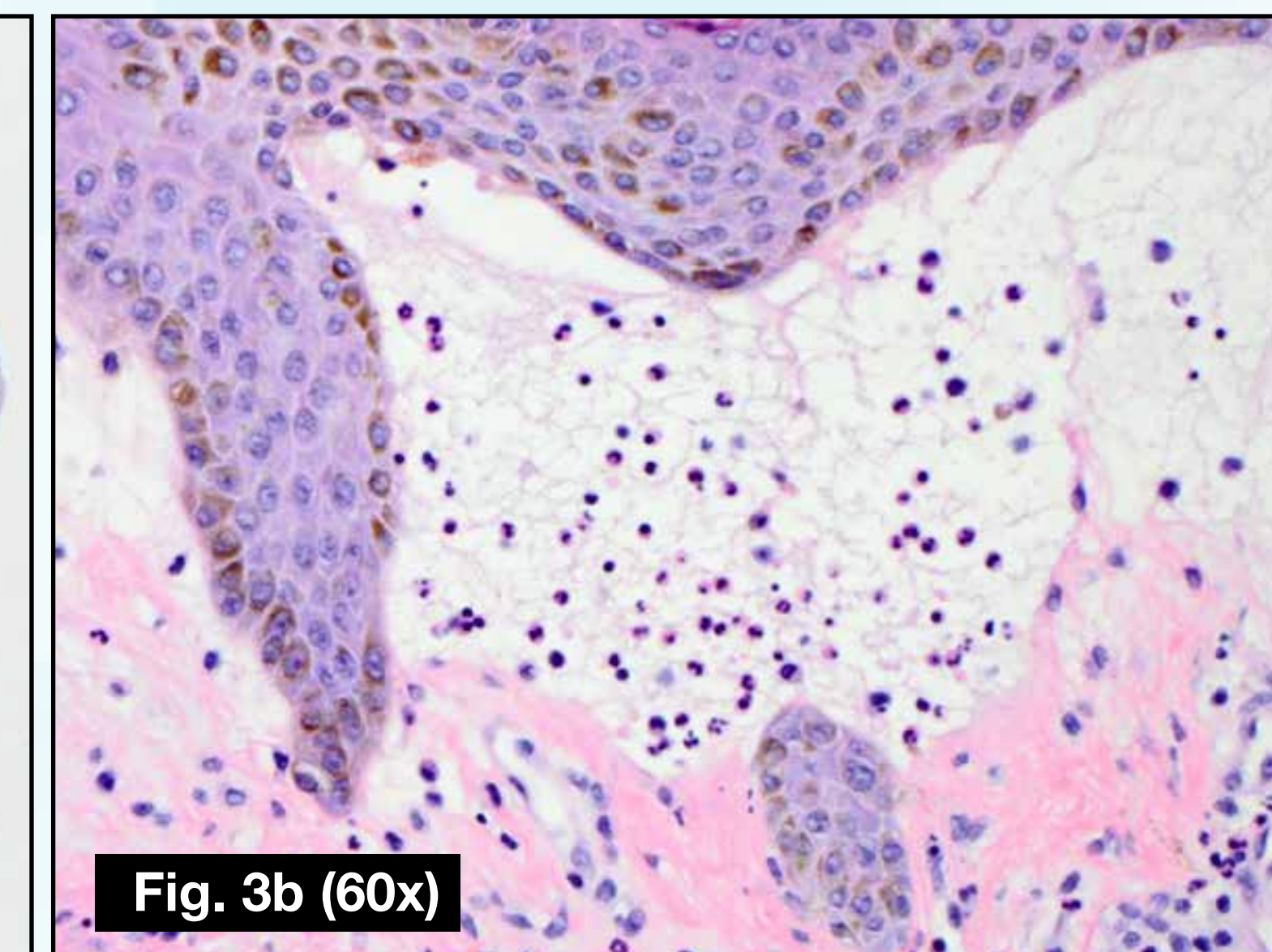
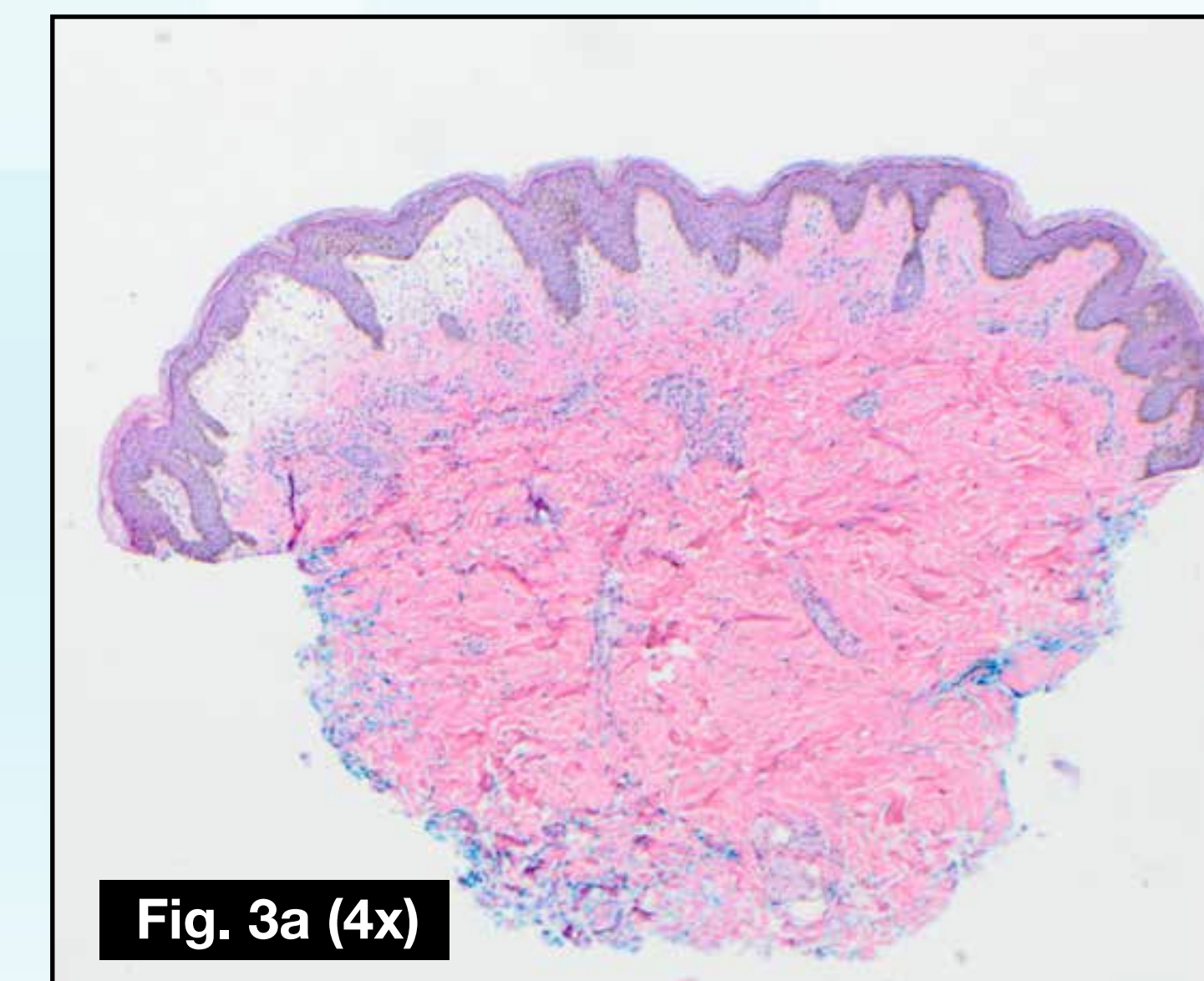


Figure 3a and 3b: Punch biopsy from back displaying a subepidermal vesicular dermatitis with neutrophils and eosinophils.

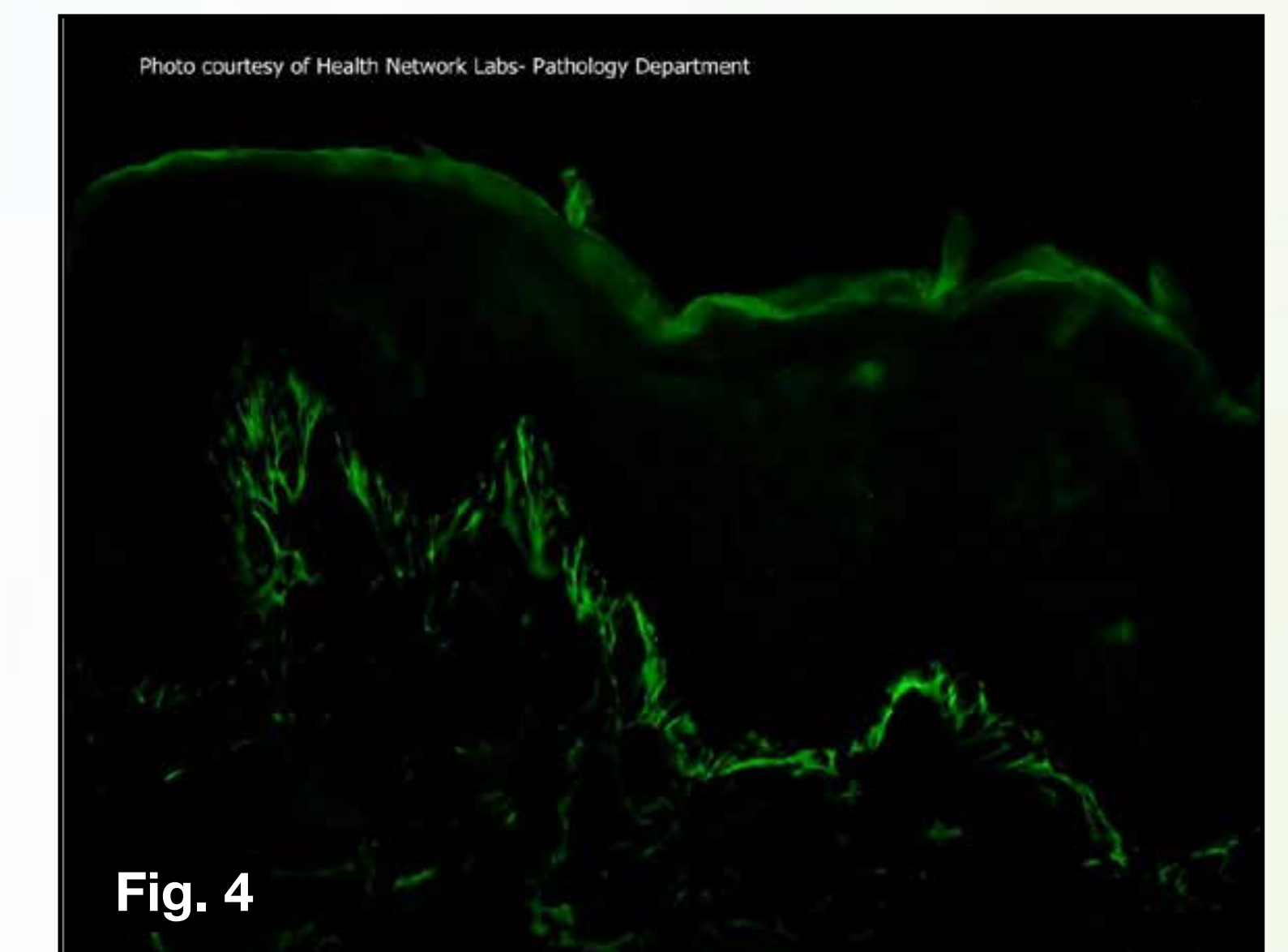


Figure 4: Direct immunofluorescence displaying strong continuous linear basement membrane zone with IgA.

Discussion

Linear IgA bullous dermatosis (LABD) is a rare acquired autoimmune subepidermal blistering disease. It is characterized by linear homogenous IgA deposition of the dermo-epidermal junction observed with direct immunofluorescence (DIF). There are two clinical presentations; one occurs in adults and the other in children, which is also known as chronic bullous dermatosis of childhood. It is the most common bullous dermatosis in children with a peak incidence at four to five years of age. In adults, the disease peaks in the teenage years and the sixth decade. The eruption may be incited by a medication, such as vancomycin or penicillins, but is often idiopathic. Some cases have also been associated with lymphoproliferative malignancies, infections, including upper respiratory tract infections, and autoimmune diseases such as rheumatoid arthritis, inflammatory bowel disease, and psoriasis. The significance of these associations is unknown, and their potential role in stimulation of the immune system has yet to be elucidated. Antibody deposition leads to activation of complement and neutrophil chemotaxis, which leads to loss of adhesion at the dermal-epidermal junction and blister formation.

The vesiculobullous eruption consists of tense bullae which are often grouped, giving the appearance of a “cluster of jewels.” Lesions usually occur on the lower abdomen, perineum, and face. Mucosal lesions can occur with oral and ocular involvement being the most common. Ocular involvement may resemble cicatricial pemphigoid and lead to blindness. Mucosal lesions may heal with scarring but cutaneous lesions generally resolve without scarring or milia. Histopathology displays a subepidermal blister with a neutrophil-predominant infiltrate in the superficial dermis, characteristically arranged in a linear array. The most preferred biopsy site is the back. Indirect immunofluorescence is used to detect circulating IgA antibodies. When using salt-split-skin technique, epidermal binding is generally observed. Western immunoblotting is more sensitive, and the most frequently detected autoantigens are the Laminin-1 (LAD-1), the 120kDa portion of BPAG2 (BP180). Cleavage of LAD-1 results in a second autoantigen, LABD97. Genetic associations between LABD and human leukocyte antigen (HLA) B8, HLA DR3, HLA DQ2, and HLA Cw7 have been reported.

The differential diagnosis of LABD includes other autoimmune bullous diseases such as dermatitis herpetiformis, bullous pemphigoid, epidermolysis bullosa acquisita (EBA), and bullous lupus. Dermatitis herpetiformis presents with group vesicles on the scalp, extensors, and sacrum and rarely involves the mucous membranes. DIF demonstrates granular deposits of IgA in the dermal papillae, not in a linear fashion, of the basement membrane zone. Bullous pemphigoid manifests with pruritic plaques and bullae and can have mucosal involvement. This generally affects persons in the sixth decade and DIF will display linear deposition of IgG and C3 along the basement membrane zone. EBA could be considered as a differential diagnosis, and purely IgA-mediated variants have been described. However, EBA resolves with scarring and milia and the DIF demonstrates linear IgG along the basement zone. Lastly, bullous lupus is an uncommon eruption in systemic lupus erythematosus. It presents with herpetiform vesicles to large hemorrhagic bullae and can involve mucosal surfaces. Histopathology will also reveal a subepidermal bulla, as in LABD, however DIF will show linear or granular band-like pattern of C3 and IgG, with or without IgM and IgA, at the basement membrane zone. Bullous impetigo could also be considered, but will have fragile bullae and a negative DIF.

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