EPIDERMOLYSIS BULLOSA ACQUISITA IN A YOUNG FEMALE

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A 20-year-old female developed recurrent blisters since 5 years. The blisters occurred following trauma and healed with scarring and milia. Indirect immunofluorescence on the sodium chloride split skin confirmed the diagnosis of epidermolysis bullosa acquisita. The immunoglobulins were localised to the floor of the sodium chloride split skin.

Key Words: Epidermolysis bullosa aequisita, Indirect immunofluoreseence

Introduction

Epidermolysis bullosa acquisita (EBA) is a rare chronic subepidermal blistering disease in which antibodies to type VII collagen are found. Flaccid, serous or haemorrhagic bulla develops at the sites of trauma and heal with scarring, milia and hyperpigmentation. Most patients are over the age of fifty, although EBA has beeen reported in children.¹

Case Report

A 20-year-old female developed continuous cough with expectoration, loss of appetite and weight. She was diagnosed as having pulmonary tuberculosis and referred for bullous lesions since 5 years.

The bulla was tense and both Nikolsky's sign and

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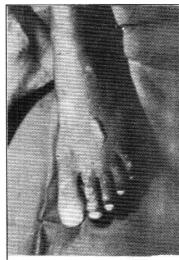


Fig.1. Bulla, milla, scarring

bulla spread' sign were positive. In addition areas of scarring, milia and postinflammatory hyperpigmentation were seen (Fig.1).

The patient d e v e l o p e d haematemesis, and endoscopy revealed a longitudinal ulcer in the oesophagus. Biopsy of an early bulla showed a sub-

epidermal bulla, filled with RBCs and transudate. Direct immunofluorescence showed a band of IgM, IgA, C3 and fibrin at the basement membrance zone. Indirect immun-

ofluorescence (IIF) showed fluorescence at the floor of the split skin.

Discussion

The diagnosis of EBA was made by the history of repeated episodes of bullous lesions which healed with scarring and milia. The presence of 'bulla spread' sign as observed in our patient has already been reported.² IIF on saline split skin helped to confirm the diagnosis by showing immunofluorescence along the floor of the split skin,³ and thus ruled out bullous pemphigoid where the band is seen along the roof of the split skin. We report this case as this is the first confirmed case of EBA in India in the younger age group.

References

- 1. Rubenstein R, Esterly NB, Fine JD. Childhood epidermolysis bullosa acquisita. Detection in a 5-year-old girl. Arch Dermatol 1987; 772-776.
- Srinivas CR, Balachandran C, Wojnarowska F, et al. Epidermolysis bullosa aquisita with positive immunoblotting on split skin and bulla spreading sign. Indian J Dermatol Venereol Leprol 1991; 57:287-288.
- 3. Gammon WR, Briggaman RA, Inman AO, et al. Differentiating anti-lamina lucida and anti sublamina densa anti BMZ anti-bodies by indirect immunofluorescence on 1.0M sodium chloride separated skin. J Invest Dermatol 1984; 82:139-144.

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