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EPIDERMOLYSIS BULLOSA ACQUISITA

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A 20-year-old male started developing bullae following even minor trauma on the skin as well as the mucous membranes during the preceding 5 years. The bullae healed with atrophic scars. Involvement of the oral mucous membrane and the eyes, especially the right eye, was quite extensive and unusual. No other family member had similar complaint; neither was there association with any autoimmune or lymphoreticular disease.

Key word : Epidermolysis bullosa acquisita.

Epidermolysis bullosa acquisita (EBA) is relatively rare and non-hereditary form of sub-epidermal bullae on mechanical trauma involving the skin and mucous membranes. Though non-hereditary form of a cutaneous disease resembling dystrophic epidermolysis bullosa was described in 1897 by Fox,[1] the term acquisita was first applied by Kablitz[2] in 1904. Only three review articles[3-5] are available, and Roenigk et al[3] analysed 19 accepted cases of EBA until 1971 and proposed the criteria for the diagnosis. These included, (1) lack of family history, (2) adult onset, (3) clinical lesions consisting of bullae produced by trauma followed by atrophic scars, milia, nail dystrophy, varying degree of mucosal involvement, and (4) exclusion of other bullous dermatoses like bullous pemphigoid, cicatricial pemphigoid, porphyria cutanea tarda, bullous lichen planus etc. Association of autoimmune and / or lymphoreticular disorders was also frequently noticed. Ultrastructural changes consist of a sub-lamina densa split with amorphous granular deposits in the papillary dermis. We report a case of EBA with extensive involvement of the skin and mucous membranes.

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Case Report

A 20-year-old male electrician was admitted for evaluation of asymptomatic, recurring bullae with scarring of the skin and mucous membranes of 5 and 2 years duration respectively, along with dysphagia and constipation of one month duration. The illness started at the age of 15 years, primarily involving the areas subjected to trauma. Trauma was followed by bullae containing clear or haemorrhagic fluid, spontaneous rupture, ulceration and healing with atrophic scars. The nature of the trauma varied from pressure and friction of tight clothing to accidental injury. The course of the illness remained unchanged despite several therapeutic measures. Three years later, he developed irritation, congestion and watering followed by blisters in the eyes leading to progressive dimness of vision and photophobia. Right eye was involved predominantly. The patient noticed blisters and erosions in the oral mucosa and throat resulting in limitation in the movement of the tongue and progressive dysphagia to solid foods subsequently. There were no photosensitivity, recurrent infections, difficulty in phonation, haemoptysis, haematemesis, melena or difficulty in micturition. Defaecation was painful. There was no history of tuberculosis, diabetes and hypertension either in the patient or his parents and siblings. The patient was the last of the 4 siblings born of non-consanguinous

parents and no other member in the family had similar illness.

He was moderately built, anemic individual with average intelligence and free of any congenital abnormality. Systemic examination showed no abnormality. He had extensive hypopigmented atrophic scars confined predominantly to the extensor aspect of the knees, elbows, acral areas, genitals and the scrotum and scattered tense bullae containing both clear and haemorrhagic fluid on the extremities, trunk and genitalia. The skin on the hands and feet was thin with atrophic scars and the fingers were spindle shaped. Palms and soles were thickened. The skin of the face and scalp was totally spared. Urethral meatus was normal but glans showed atrophic scarring. Anal mucosa showed erosion and fissures. Ophthalmic examination showed congestion, symblepharon, haziness of the cornea, ectropion with loss of eyelashes and epiphora; right eye was involved more than the left. Oral cavity and throat showed haemorrhagic bullae with erosions, ankyloglossia and darkly stained teeth. Longitudinal ridging was seen on the finger nails while all the toe nails were absent with scarring of the nail beds.

Except for anemia (RBC 2.3 million/mm^3) and hookworm ova in the stools, the following tests were normal; total and differential white cell count, ESR, blood urea, blood sugar, VDRL, serum creatinine, electrophoresis, routine urine analysis, tests for porphyrins, x-ray chest, barium swallow and meal. Tzanck test showed no acantholytic cells. Biopsy of the bulla revealed sub-epidermal bulla containing neutrophils and a few eosinophils and sparse perivascular inflammatory cells in the dermis.

Comments

The diagnosis of epidermolysis bullosa acquisita was quite evident in our patient as it fulfilled the criteria proposed by Roenigk et al.[3] Most reported cases of EBA have shown relatively mild dystrophic changes, often limited to extensor surfaces, scarring, nail dystrophy and milia. Review of 19 cases by Roenigk et al revealed mucosal involvement in only one case. Although mucosal scarring has been reported in EBA,[3] scarring involving the ocular, oral and oesophageal mucous membranes of this magnitude simulating recessive epidermolysis bullosa dystrophica (REBD) has seldom been described. Lack of family history, adult onset of the disease with no deformity make the diagnosis of REBD untenable while an unusually severe variant of EBA simulating the mucosal features of REBD cannot be discounted dogmatically on clinical grounds alone.

The exclusion of cicatricial pemphigoid was not easy but simultaneous occurrence of the bullae following trauma in an adult, involvement of extensive areas of the skin and mucous membrane and dystrophy of the nails speak in favour of EBA.

Roenigk et al[3] emphasized the association of either an autoimmune of a lymphoreticular disease while others[5-7] noted the association of numerous other autoimmune diseases such as SLE, rheumatoid arthritis, thyroiditis, diabetes mellitus, inflammatory bowel disease, cryoglobulinemia and multiple endocrinopathies syndrome. However, Nieboer et al[8] could not find the high incidence of association of systemic diseases in their 4 patients, as also seen in our patient.

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