PAEDIATRIC DERMATOLOGY

INHERITED EPIDERMOLYSIS BULLOSA SIMPLEX: A KOEBNER TYPE

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Background: Inherited Epidermolysis bullosa (EB) is a grouping of rare genetic conditions characterized by extreme fragility of the skin and mucous membranes, which gives rise to the formation of blisters and ulcers following minor trauma. In EB simplex, trauma induced loss of tissue integrity consistently occurs within the basal layer of epidermal keratinocytes.

Observation: 9 days old neonate was hospitalized with chief complain of blisters on various parts of body since 3 days after birth. The baby was born to non consanguineous parents. The term male baby with birth weight of 2600 kg was born to second gravida mother by normal delivery. Systematic examination was normal. The blisters of variable sizes were present on his chest, back, abdomen, palm, elbow, knee extending below including dorsum of feet and buttock. The blisters were easy to break and contained relatively clear fluid, some blisters was broken and became erosion. Oral cavity was involved but conjunctiva, cornea and scalp were normal. There were no nails dystrophic and nikolsky was positive. Skin biopsy was performed. Biopsy showed subepidermal blisters that contains few neutrophils and eosinophils. In dermis there are lymphocyte infiltration, with neutrophil predomination and erythrocyte extravasation
On the basis of biopsy report and clinical features diagnosis of epidermolysis bullosa simplex was confirmed. Systemic antibacterial Cloxcilin was given to patient, after 10 days of treatment and skin erosion were treated with topical antiseptic, antibacterial agents and nonadherent dressings a significant improvement was observed.

Key message: There is presently no definitive cure for epidermolysis bullosa. The standart treatment is to alleviative the symptoms and provide supportive measures. Therapy is therefore focused on the prevention including avoidance of trauma, and halting the progression of skin lesions and complications.