



PAEDIATRIC DERMATOLOGY

## ICHTHYOSIS BULLOSA OF SIEMENS SUPERIMPOSED WITH SCABIES: AN UNDERDIAGNOSED CASE

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**Background:** Ichthyosis bullosa of Siemens (IBS) is a rare autosomal dominant hereditary keratinization disorder which is distinguished clinically from bullous congenital ichthyosiform erythroderma (BCIE) by the absence of erythroderma, localization of dark grey hyperkeratosis on flexural sites, and areas of peeling of the skin known as the “Mauserung phenomenon”. Scabies is a common skin infestation caused by *Sarcoptes scabiei* presents with an intensely pruritic rash in predilection sites and typical burrows.

**Observation:** A 13-year-old girl presented with generalized erythematous macules, papules, pustules, blister, crust, and scales which was itchy especially at night since 3 weeks before consultation. She had history of scattered few flaccid blisters formation over her whole body since 1 day of age followed by erosion. The blisters came and went with decreasing number, with accompanying increased hyperkeratosis on flexor and extensor with keratoderma on palms and soles. There was no history of blisters or hyperkeratotic in family. Skin scraping from interdigital of hands and abdomen found *Sarcoptes scabiei* adult mites, and eggs. Histopathological examination from skin blister showed intraepidermal blister, vacuolar degeneration and keratohyaline granules in granular until spinous cell layers of epidermis with overlying basket weave hyperkeratosis and acanthosis. The patient was diagnosed with IBS and scabies and treated with twice application of topical permethrin 5% with 1 week interval which cleared the scabies. Then followed by topical 0,025% tretinoin and 100% glycerine twice daily with marked improvement of scales and hyperkeratosis.

**Key message:** Atypical scabies manifestation may include bullous lesions. Definite diagnosis is made if *Sarcoptes scabiei* mites or eggs are found. Ichthyosis bullosa of Siemens has milder clinical findings to those of BCIE. IBS diagnosis is missed due to its mild clinical manifestation and an overlapping scabies infestation. Management of IBS involves reduction of symptoms with application of glycerine which can normalize keratinization process.

