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Blistering Diseases in Pediatrics: A Case Series

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Abstract

Introduction: Vesicles and bullae are associated with various conditions in paediatric practice. These conditions may be Inherited, Drug Related, Infectious, Autoimmune and Miscellaneous. While Epidermolysis Bullosa (EB) is a type of Inherited Bullous disorder, Steven Johnson's Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are drug reactions characterized by epidermal detachment and necrolysis presenting with macules, papules, plaques and flaccid bullae. Being rare in paediatric practice and of acute onset, they pose both diagnostic and management challenges. Our three patients had an Acute presentation with Blistering skin lesions at the onset.

Key-words: Acute Blistering in Pediatrics, Stevens Johnson Syndrome, Toxic Epidermal Necrosis, Epidermolysis Bullosa Simplex, Drug Induced Blisters

Introduction

Acute Blistering lesions in Pediatrics present clinically as Vesicles or Bullae. Etiology may be: Inherited, Drug Related, Infectious, Autoimmune and Miscellaneous. Epidermolysis Bullosa (EB) is a type of Inherited Bullous disorder. Steven Johnson's Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are a type of Drug Related skin disorder with macules, papules, plaques and flaccid bullae. These conditions are rare in Pediatric practice. Thus, they pose both diagnostic and management challenges.

Case History:

Case 1: A 2 ½ year old male child with fever and cough reported after 48 hours of some home therapy with painful erythematous purpuric macules and targetoid lesions all over the trunk, face and back along with bullae and erosive lesions involving the oromucosal surface. Child had labial and periorbital oedema with conjunctival inflammation, inability to take oral feeds due to oral mucosal edema and had excessive drooling of saliva [Figure 1]. History revealed intake of Co-Trimoxazole, Ibuprofen and Levocetrizine. He had tachycardia, fever and polymorphonuclear leukocytosis. A diagnosis of Stevens Johnson Syndrome was entertained. All previously prescribed drugs were stopped. He was managed with IV fluids and Ryle's tube

feeding. Skin cleansing, oral hygiene and supportive care was given along with IVIG (total 2Gm/Kg IV), Antibiotics (Amoxycillin+Clavulinic Acid) and Syp.Prednisolone@2mg/kg. After 48 hours the child was afebrile, the oral and labial oedema regressed with crusting. The child was able to take oral feeds and IV fluids were stopped. After 5 days the edema regressed and child was on oral feeds along with oral medication. By day 7 he had recovered fully and discharged from hospital with a final diagnosis of SJS due to possibly Co Trimoxazole or Ibuprofen. Parents were advised not to give these drugs in future.

Case 2: A 12-year-old girl, a recently diagnosed case of Neurocysticercosis with complex partial seizures was started on Tablet Carbamazepine. After 1week she developed vesiculobullous lesions all over the body. She denied any drug intake other than Carbamazepine. On examination the child was afebrile. Vital parameters, neurological, systemic examination and hematological parameters were normal. Dermatological examination revealed erythematous macules and targetoid lesions with vesiculobullous eruption all over her face, trunk and limbs covering over 80% of her body surface area [Figure2]. In view of her skin lesions and history of Carbamazepine intake a diagnosis of Toxic Epidermal Necrolysis (TEN) was made. Carbamazepine was stopped immediately. She was started on Steroids (Tab Prednisolone @ 2mg/Kg) along with IVIG (Total dose 2gm/kg) and anticonvulsant Sodium valproate for seizures. Aseptic skin care, non-adherent antiseptic dressing and systemic antibiotics were given. The patient improved over the next 2 weeks and the bullae healed without scarring. Carbamazepine was stopped immediately.

Case 3: A term male baby was born with severe skin blistering especially around the elbow, ankles, trunk and back. Peeling appeared more superficial without bleeding suggestive of Epidermolysis bullosa simplex [Figure3]. The mother did not have any comorbidities, was not on any drugs, no history of consanguinity or similar cases in the family. The vesiculobullous lesions were distributed all over the body. Oral cavity, conjunctiva, cornea, nails, scalp and genitalia were normal. Systemic examination and hematological parameters were normal. When handled, new blisters and erosions appeared. Nikolsky's sign was positive. Gram stain and culture of bullous fluid was negative for Staphylococcus aureus. He was managed with aseptic supportive care and Nasogastric feeds since handling for breast feeding was causing extensive blistering. Parents took the baby home against medical advice.

Discussion

Stevens Johnson Syndrome and Toxic Epidermal Necrosis

The incidence rate in India of SJS and TEN is from 1.2 to 6/ million patient‑ years and 0.4 to 1.2/million patient‑ years, respectively.[4]

SJS and TEN occur by epidermal detachment and necrolysis predominantly of drug induced etiology as depicted in Table 2.

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Table 2: Drugs causing SJ and TEN in Childhood[5]

Drug Category	Drug Name
Antibiotics	Penicillin, Cephalosporins, Macrolides, Fluoroquinolones, Sulfonamides
Antiepileptic	Valproic acid, Lamotrigine, Carbamazepine, Phenytoin, Phenobarbitone
Non-Steroidal Anti-Inflammatory Drugs (NSAID)	Nimesulide, Paracetamol
Others	Allopurinol, Nevirapine, Ayurvedic and Homeopathic medicines

The appearance of SJS/TEN starts with symptoms of fever, headache, malaise, anorexia, myalgia, and sore throat for two or three days lasting up to ten or eleven days [6]. Then the mucosal and skin lesions appear. Mucosa, usually of the conjunctiva and oral, is affected before the skin. Flaccid Blisters and erosions appear on the face, trunk, limbs, and mucosa. Acute dysfunction of ocular, respiratory, cardiovascular, gastrointestinal, renal, and hematological systems occur. [5]

SCORETEN is a severity score to predict the mortality in cases with TEN and is calculated on day 1 and day 3 of the illness. About <5% of SJS patients die but mortality in TEN patients in Acute stage is >30%.[6]

SJS/TEN Treatment:

Remove the causative drug immediately, treatment of skin and eye lesions, supportive care,

Systemic adjunctive treatment: corticosteroids, IVIG, cyclosporine, anti‑TNF antibodies (infliximab and etanercept) and plasmapheresis.[4]

Epidermolysis Bullosa (EB) is a complex group of autosomal dominant disorders producing various degrees of skin blistering and shearing. It affects about one in 17,000 live births, with an estimated 500,000 cases worldwide.[1]

Epidermolysis Bullosa is of Four major types based on cleavage zones

•Epidermolysis Bullosa Simplex (EBS): Blistering in epidermis within basal keratinocytes

•Junctional Epidermolysis Bullosa (JEB): Blistering within lamina lucida

•Dystrophic Epidermolysis Bullosa (DEB): Blistering in the dermis/ sub lamina densa

•Kindler Syndrome: Collagen binding the Outer Epidermis and Inner Dermis is lacking in Epidermolysis Bullosa.[1] Blisters occur within or below the Basement Membrane zone or occur in both in Kindler Syndrome. Photosensitivity is one of the remarkable clinical features of Kindler's syndrome.[2]

EB causes pain and results in long lasting debility. Butterfly Children is the term for EB affected children as their skin is delicately friable like a butterfly's wing.[3]

Blisters are present on the scalp, elbows, hands, knees, legs and feet in EBS. The time of appearance of blisters is at birth or in the neonatal period in EB.[3]

The treatment for Epidermolysis Bullosa is only symptomatic. Non adherent dressing of Blisters or eroded skin is recommended.

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Figures

Figure 1: Clinical picture of Steven Johnson's Syndrome

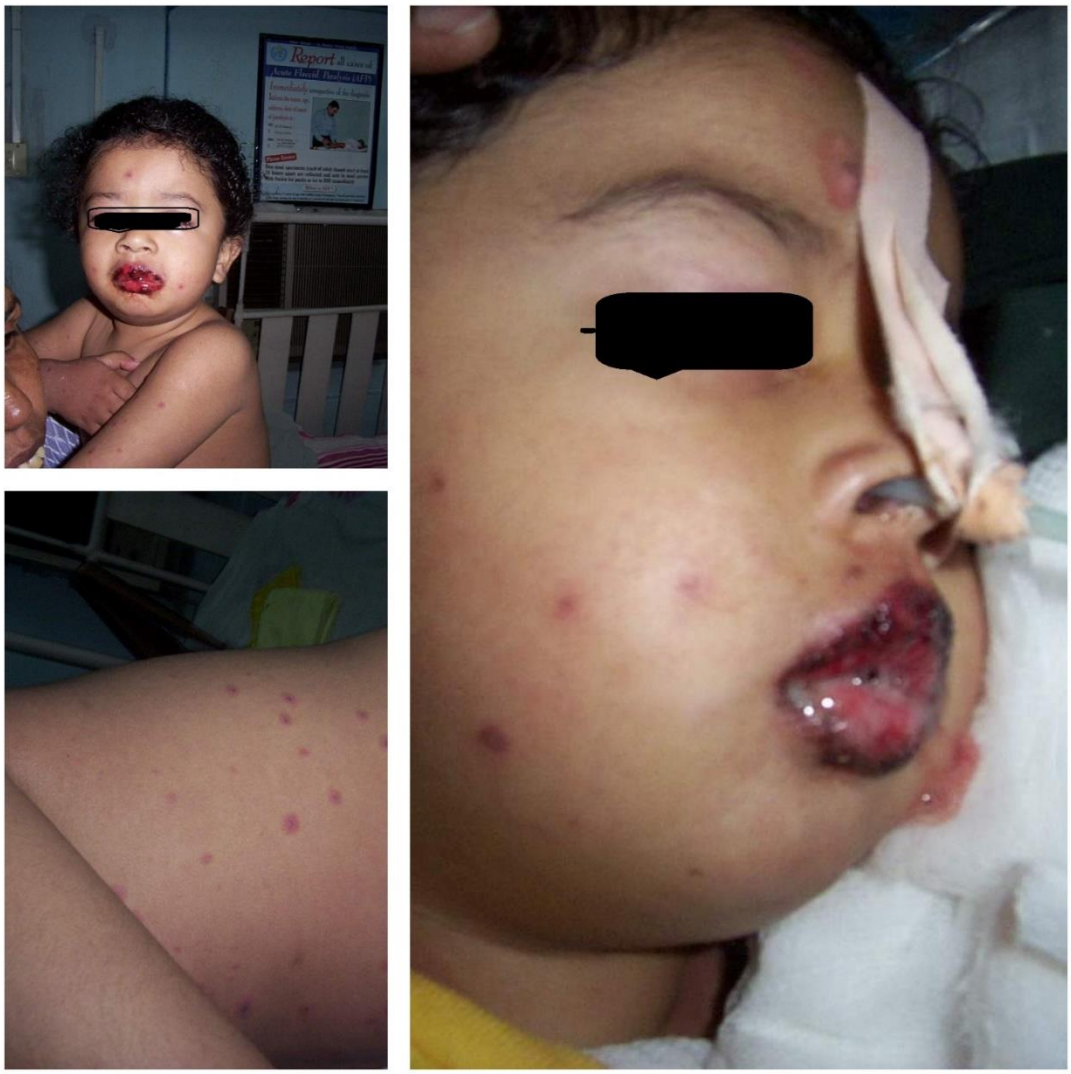

Figure 2: Clinical picture of Toxic Epidermal Necrolysis

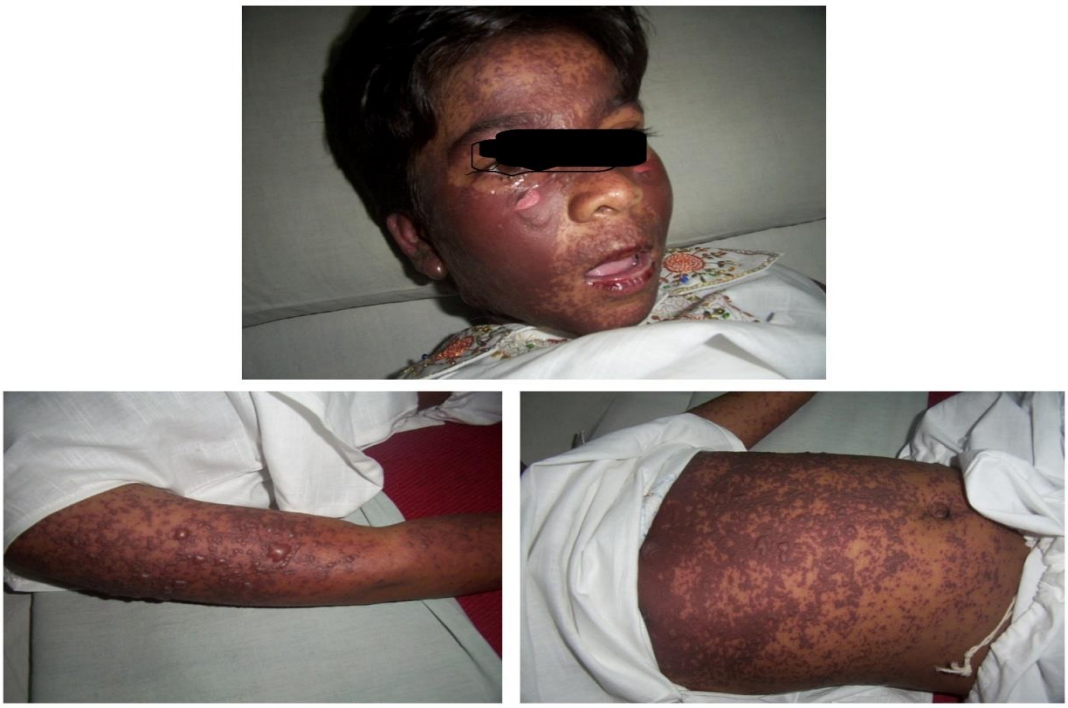

Figure 3: Clinical picture of Epidermolysis Bullosa Simplex