Indian Academy of Pediatrics (IAP)

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Staphylococcal Scalded Skin Syndrome

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Staphylococcal Scalded Skin Syndrome

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Introduction

- ☑ Staphylococcal scalded skin syndrome (SSSS) is a blistering skin condition that occurs due to the epidermolytic toxin produced by certain strains of *Staphylococcus aureus*.
- ☑ It was first described by Baron Gottfried Ritter von Rittershain in the 19th century.
- ☑ It usually occurs in neonates, infants and young children aged <6 years of age, but may rarely occur in adults, who are immunocompromised or have renal dysfunction, diabetes, malignancy, or cardiac disease.

Synonyms

- ☑ Ritter disease
- ☑ Scalded skin syndrome
- ☑ Ritter von Rittershain disease
- ☑ SSSS
- ☑ Staphylococcal epidermal necrolysis
- ☑ Pemphigus neonatorum.

Etiopathogenesis

Certain strains of *Staphylococcus aureus*, especially type 71 of phage group II produce epidermolytic or exfoliative toxins (ETA and ETB), which are responsible for the skin manifestations seen in SSSS. These toxins which are serine proteases reach the skin via the blood circulation and target the desmosomal intercellular adhesion molecule, desmoglein 1 and cause cleavage of the epidermis at the level of the stratum granulosum layer. A localized effect causes bullous impetigo, while a systemic spread of the exotoxins causes widespread desquamation. Most organisms identified are methicillin-sensitive *Staphylococcus aureus* (MSSA). However, a significant proportion of infections, even those acquired in the community, are caused by methicillin-resistant *Staphylococcus aureus* (MRSA).

Children are more susceptible to SSSS because:

- ☑ Lack of antibodies to exotoxin
- ☑ Immature renal clearance of exotoxins.

Clinical Features

- ☑ The disease afflicts children under the age of 6 years but is most common between the age of 2 and 3 years. There is a usual history of a localized infection of the perioral region, umbilicus, perineum, nares, conjunctivae, or an upper respiratory infection. Endocarditis, septic arthritis, and pyomyositis may also lead to SSSS.
- ☑ Clinical manifestations of SSSS can range from localized blistering to generalized desquamation and erythroderma.
- ☑ Affected children are febrile and irritable a day or two before the typical skin lesions appear. They usually have rhinitis, conjunctivitis, malaise, and loss of appetite. Erythema appears first on the periorificial areas of the face, axillae, neck, and groin, thus indicating the predilection for the flexures.
- ☑ The skin becomes extremely tender, with the child becoming highly irritable and crying even on light touch.
- ☑ This is followed by the development of superficial and large fragile bullae that rupture to leave behind areas of desquamated, denuded, and raw areas of skin. The skin appears red and wrinkled, with diffuse peeling. Gentle pressure on the skin causes it to peel off (Nikolsky's sign). Extensive exfoliation of skin that follows within hours to days may result in fluid loss, electrolyte imbalance, thermal dysregulation, and increased propensity to develop secondary bacterial infections.
- ☑ Crusting occurs and when the perioral crusts get separated, radial fissures are seen around the mouth, which is referred to as the characteristic appearance of the face in SSSS.
- ☑ Palms, soles, and mucous membranes are not involved.
- ☑ Without treatment, the disease can be fatal. Mortality in children is below 5% with appropriate treatment. Sepsis, electrolyte imbalance, and dehydration are associated with poor outcomes.

Differential Diagnosis

Proper history taking and astute clinical examination will facilitate the correct diagnosis.

- ☑ Toxic epidermal necrolysis (TEN), which is the most important differential diagnosis of SSSS is usually drug induced and is characterized by the presence of severe exfoliation and mucosal involvement
- ☑ Epidermolysis bullosa
- ☑ Scalding burns/thermal burn/sun burn
- ☑ Nutritional deficiency dermatoses
- ☑ Graft-versus-host disease
- ☑ Neonates—bullous ichthyosiform erythroderma and diffuse cutaneous mastocytosis.

Investigations

- ☑ Diagnosis of SSSS is usually based on the characteristic clinical presentation.
- ☑ Confirmation of the diagnosis can be done by Gram stain and culture, which should be done from the primary pyogenic foci of the skin/nares/nasopharynx/conjunctivae, as the blisters in SSSS are caused by the epidermolytic toxin.
- ☑ Examination of frozen section of blister roof/denuded skin facilitates rapid diagnosis.

The tissue sample is snap frozen in liquid nitrogen and sections cut. In SSSS, cleavage is seen in the superficial layers of the epidermis at the level of the stratum granulosum, whereas in TEN, the cleavage is much below the dermoepidermal junction, with the presence of necrosis of the entire epidermis.

General Care

- ☑ Child with SSSS should be admitted in intensive care unit (ICU). No management on regular outpatient department (OPD) basis
- ☑ Incubators to maintain body temperature and humidity
- ☑ Management of fluid and electrolyte abnormalities
- ☑ Nutritional care
- ☑ Saline compression for the crusted areas three to four times per day (four-fold saline gauze place over the affected area and removed after 15 minutes)
- ☑ Banana leaf (autoclaved) smeared with liquid paraffin for laying the child and can be used as a dressing. Pressure relieving mattress should be considered
- ☑ *Nonadherent dressing on raw areas*:
  - Petrolatum impregnated gauzes
  - Mepitel dressing (silicone dressing, only if affordable)
- ☑ Contact isolation should be considered. Swab from anterior nares should be taken from suspected asymptomatic carriers.

Topical Treatment

- ☑ Mupirocin 2% ointment/cream for few areas like face/nares/paranasal and perioral region
- ☑ Liquid paraffin/bland emollients for desquamation phase (healing areas)
- ☑ Topical silver sulfadiazine is to be avoided.

Systemic Treatment

Child should be started on intravenous (IV) antibiotics based on the culture sensitivity. Early administration of antibiotics is necessary for treating the active focus and halting the progression of the disease. Penicillinase-resistant penicillin, first- or second-generation cephalosporins are the appropriate options.

First Line of Treatment

- ☑ IV cephalosporins (first/third generation)
  - Cefazolin 30 mg/kg/dose (maximum dose—2 g/dose) IV eighth hourly for 7–10 days
  - Cefotaxime 150 mg/kg/day IV eighth hourly for 7–10 days.

Management

Systemic Treatment

Second Line of Management

- ☑ IV flucloxacillin 50–100 mg/kg (maximum dose—2 g/dose) sixth hourly
  +
- ☑ IV clindamycin 10 mg/kg (maximum dose—600 mg/dose) sixth hourly

} for 7–10 days

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If child is stable after 7 days with IV antibiotics and skin lesions are showing desquamation, child can be discharged after 7 days with oral antibiotics for 1 week and subsequently followed after 1 week.

- ☑ Oral cephalexin 25–50 mg/kg/dose.

Note on management of MRSA-induced SSSS

- ☑ IV vancomycin 20 mg/kg/dose, eighth hourly (maximum dose—1,250 mg/dose) (>5 years with normal renal function tests) for 10 days

OR

- ☑ IV linezolid 10 mg/kg/dose, eighth hourly (maximum dose—600 mg/dose) for 10 days.

Patient Education

- ☑ Counseling about the disease and prognosis and the need for follow-up.
- ☑ Parents need to be reassured that the lesions will heal without scarring.
- ☑ Avoidance of multiple handlers of the child/minimum handling of the child should be ensured.
- ☑ Regular disinfection of the hands of caregivers/instruments used in the management of SSSS.
- ☑ Finger nails should be short to avoid staphylococcal contamination.

Further Reading

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