Staphylococcal Scalded Skin Syndrome

Staphylococcus aureus is a bacterium commonly found harmlessly colonising human skin and mucosa without causing any morbidity. Colonisation begins soon after birth. It can, however, predispose to infection, particularly when the bacteria have an opportunity to break through the skin. This is one of the most common causes of skin infection, giving rise to folliculitis, impetigo, cellulitis and abscesses. S. aureus may also cause a number of toxin-mediated, potentially life-threatening diseases, including staphylococcal scalded skin syndrome (SSSS).

SSSS is a disease characterised by red, blistered skin, which resembles a burn or a scald, hence the name. It can also be known as Ritter's disease.

Pathogenesis

SSSS is caused by certain S. aureus strains. Most commonly it is caused by those belonging to phage group II (types 3A, 3B, 3C, 55, or 71) but occasionally to groups I and III. They cause skin damage by releasing epidermolytic toxins. These are serine proteases which are spread by the circulation from a localised source, causing widespread epidermal damage at distant sites. They break the epidermal cell adhesion molecule, desmoglein 1, breaking up the skin by preventing skin cells sticking to each other. In SSSS the superficial epidermis becomes detached.

The same toxins that are responsible for causing SSSS also cause bullous impetigo. Some consider bullous impetigo to be a type of SSSS. There appears to be a relationship between the disease extent, the amount of toxin produced and whether the toxin is released locally or systemically. As a result there is likely to be a spectrum of disease and there are likely to be a number of milder cases of adult SSSS that go undiagnosed.

Epidemiology

SSSS typically affects neonates and children aged under 5 years, but may also occur in predisposed adults. It is thought that SSSS is less common in adults because of their higher concentrations of circulating antibodies to exfoliative toxin, acquired during childhood, and the adult kidney's ability to excrete the toxin. Neonates are conversely most at risk due to their immature renal clearance of toxins, and lack of immunity to them. For the same reasons, those with immune deficiency or poor renal function at any age are more at risk.

98% of cases occur in children under the age of 6. Incidence figures are estimated to be between 0.09-0.13 cases per million people.

Presentation

- The initial clinical features of SSSS are fever, generalised erythema, and skin tenderness. There may be a prodrome of sore throat or conjunctivitis.
- Extremely tender flaccid bullae (large superficial blisters), which are Nikolsky sign-positive (where gentle shearing force on intact skin causes the upper epidermis to slip, indicating a plane of cleavage in the skin), develop within 48 hours and commonly affect the flexures; occasionally, large areas of the skin may be involved.
- The bullae enlarge and rupture easily to reveal a moist erythematous base, which gives rise to the scalded appearance.

Differential diagnosis

- For many years SSSS and toxic epidermal necrolysis (TEN) were thought to be the same disease because of their clinical features. However, the layer of skin detachment differs, and TEN is most commonly due to a drug reaction. The two can be differentiated by skin biopsy.
• Blistering associated with S. aureus also occurs in bullous impetigo, in which the blisters are discrete and are not accompanied by generalised erythema.
• The responsible toxins also cause toxic shock syndrome but skin tenderness and Nikolsky's sign are not classic features.
• Staphylococcal scarlet fever affects mainly children of school age, and the generalised erythematous rash, classically without blisters or Nikolsky's sign, is followed on days 5-10 by desquamation.
• Pemphigus.

Investigations

• Diagnosis is usually clinical.[9]
• Take swabs for bacteriological confirmation and antibiotic sensitivities and to identify the primary focus of infection.
• Frozen sections from skin biopsies of the lesions can confirm the diagnosis.
• The superficial blisters are generally culture-negative, but S. aureus is usually grown from material obtained from the suspected site of infection - eg, umbilicus, conjunctiva, breast, surgical wound, nasopharynx or, occasionally (especially in adults), blood.
• Nasal swabs from the patient and immediate relatives should be performed to identify asymptomatic nasal carriers of S. aureus. In the case of outbreaks on wards and in nurseries, healthcare professionals should also be swabbed.[3]

Management[3, 9]

It is painful and distressing for the patient and parents, although most cases respond to antibiotic treatment.

• Hospitalisation is usually required.
• Supportive care and appropriate attention to fluid and electrolyte management usually ensure rapid recovery.
• Moist, bare areas should be lubricated with a bland emollient to alleviate pruritus and tenderness.
• Analgesia may be required. Paracetamol is usually first line. If pain is severe, an opioid infusion is preferred to non-steroidal anti-inflammatory drugs (NSAIDs) because the damaged skin is already prone to bleeding and renal excretion of the exotoxins makes maximised renal function important.
• Enteral nutrition must be commenced if oral intake is not possible.
• Physiotherapy is important because SSSS tends to affect limb flexures most severely and patients will voluntarily restrict movement because of discomfort.
• Topical therapy should constitute either fusidic acid as a first-line treatment, or mupirocin in proven cases of bacterial resistance.[10]
• First-line systemic therapy is oral or intravenous flucloxacillin.

Complications

Dehydration, cellulitis, sepsis and pneumonia are possible.[2, 9]

Children with severe SSSS (>50% body surface area) may need to be transferred to a tertiary paediatric burn unit for multidisciplinary care in an intensive care environment.

Prognosis

• Skin usually heals well without scarring.[6]
• Mortality is relatively low in infants (approximately 4%) but can be as high as 67% in adults, and depends on the extent of skin involvement and whether there are comorbidities.[9, 11]
• Outbreaks may occur, and particularly in neonatal units can be difficult to control.

Further reading & references

3. Staphylococcal scalded skin syndrome; DermNet NZ

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