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Approach to Staphylococcal Scalded Skin Syndrome (SSSS)

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Introduction:

Hi everyone! My name is Jasmine and I am a third year medical student at the University of Alberta. This podcast was created with Dr Melanie Lewis a Professor and General Pediatrician from the Stollery Children's Hospital. This PedsCases podcast is designed to give you an approach to staphylococcal scalded skin syndrome (SSSS). It is a blistering skin infection that is seen mainly in infants and young children.

By the end of this podcast, our goal is to have covered the following learning objectives.

Learning Objectives:

1) Identify and describe the morphology of lesions seen in SSSS.
2) Discuss the pathophysiology and risk factors for the development of SSSS.
3) Review the differential diagnosis and the management of SSSS by presenting a clinical scenario.
4) Counsel caregivers on the treatment of SSSS.

Clinical Case:

Let's begin with a brief clinical case.
You are a 3rd year medical student on your pediatrics rotation, and you see your first patient of the day, Annie, a previously healthy 4-year-old female accompanied by her mother. She presents with a 2-day history of a generalized blistering skin rash associated with a fever up to 39.9 degrees Celsius, rhinorrhea and fatigue. The rash began under the axilla and progressed to her face, trunk and back. She attends daycare 3 times per week. Her immunizations are all up to date and she does not take any medications nor have any known allergies.

On physical examination, Annie appears to be fatigued while laying quietly in her mom's lap. You note a diffuse erythematous rash over her face, trunk and across her back. There is evidence of exfoliative desquamation over her cheeks, mouth and axilla. She has 3 flaccid bullae on her upper trunk region, and a positive Nikolsky's

sign (slight rubbing/pressure on the skin results in a bullae to get larger or a bullae to rupture and result in an eroded area). The exfoliative process affects approximately 10% of her total body surface. She had dry mucous membranes however there was no involvement of the mucous membranes.

Annie's mother appears concerned and asks you what may have caused the rash.

So what is SSSS?:

Staphylococcal scalded skin syndrome, also known as Ritter's disease, Staphylococcal Epidermal Necrolysis and Pemphigus neonatorum is a blistering skin disease caused by exfoliative exotoxins produced by some Staphylococcal bacteria that have spread through the bloodstream.[1] It primarily affects infants and young children under the age of 6, who have a decreased ability to clear the toxin renally and/or may lack the toxin-neutralizing antibodies.[1,2] The exfoliative, or epidermolysis exotoxins result in the development of tender, flaccid bullae within the epidermal layer. The severity can vary from local to generalized involvement and possible sequalae include hemodynamic changes, and electrolyte abnormalities, associated with loss of the protective skin barrier because of the blisters.

Pathophysiology and Risk Factors for the Development of SSSS:

Hematogenous spread of exfoliative toxins A and B produced by the phage group II stains of S. aureus (MSSA or MRSA) bacteria results in the blister formation.[1,3] The exotoxin cleaves desmoglein 1 a protein in desmosomes located in the stratum granulosum that results in splitting of desmosomes and subsequent separation of the epidermis that results in bulla formation. The infection may spread from other sites such as the nasopharynx or conjunctivae and have widespread effects.[1]
As mentioned, it typically occurs in children younger than 6 years. It may be seen in older children and adults, immunocompromised individuals and those with renal insufficiency are specifically at risk for the development of this condition.[1]

Clinical Features of SSSS:

There often is a prodrome of symptoms that may include fever, generalized malaise and skin tenderness. Depending on the site of the initial infection, the patient may have rhinorrhea, or conjunctivitis.[1-3]

Erythema typically manifests first in the head/neck region associated with possible facial edema and at intertriginous sites. Although the disease may only be localized, the generalized spread of the rash may also occur within 1-2 days, followed by the development of flaccid, sterile bullae in the epidermis yielding a positive Nikolsky sign.[1-3] These bullae can easily rupture and lead to areas of erosion, typically involving the flexural aspects initially. Crusting can occur around the perioral and periorbital region with associated fissuring.[3] Later progression includes desquamation or peeling of the skin followed by re-epithelialization. Depending on the severity of the disease and distribution, concurrent septic shock and/or hypotension may be present.

Overall, the diagnosis can be made clinically. Consider eliciting history of sick contacts, particularly with staphylococcus infected individuals. Given the natural progression of spread of SSSS, consider culturing the site of initial infection to isolate Staph species (this may include nasopharynx, conjunctivae, or umbilicus in neonates).[3]

Important differential diagnoses include:

Perhaps the most important differential diagnosis of staphylococcal scalded skin syndrome is Steven-Johnson Syndrome (SJS)/Toxic Epidermal Necrolysis (TEN).[1-3] Key differentiating factors include that TEN is usually drug induced, primarily seen in adults, and generally involves the mucous membranes. While SSSS, is primarily seen in a pediatric population following infection with a toxin-producing Staph aureus species, and results in a generalized rash with worsening erosions along the flexural aspect, and it does not involve mucous membranes. Notably, both conditions may elicit a positive Nikolsky sign. Histopathology can also help differentiate between these conditions. SSSS results in splitting in the granular layer of the epidermis whereas TEN involves dermal-epidermal separation in addition to the epidermal necrosis – resulting in comparatively more discrete, tense bullae, and it may be involve inflammatory infiltrates in the dermal layer.[1,3] Other differentials include iatrogenic causes such as a drug reaction, infectious etiologies such as a cellulitis, streptococcal scarlet fever, viral exanthem, bullous impetigo, toxic shock syndrome, or graft-versus host disease, inflammatory cause such as Kawasaki disease, autoimmune causes such as pemphigus foliaceus or an immersion burn or sunburn.[1-3]

What about some of the complications?

Complications associated with SSSS may include septic shock and associated hemodynamic instability. Due to loss of the protective skin barrier, patients may be susceptible to fluid and electrolyte imbalances, as well as superimposed bacterial and fungal infections, if large areas of the body are involved and/or they are immunocompromised.[1-3]

Treatment

In patients with mild cases, who are otherwise well and stable, with minimal body surface area involvement, treatment may be initiated with oral antimicrobials. Agents that target staphylococcal include cephalexin and cloxacillin, although SSSS may also be caused by MRSA. While clindamycin may reduce exotoxin production some strains may be resistant to clindamycin monotherapy.[1]

Patients presenting with extensive involvement or at risk of sepsis and dehydration would benefit from hospitalization for further diagnostic workup cultures, intravenous hydration, and initiation of parental antimicrobials with both MSSA and MRSA coverage. Some children may benefit from treatment in a burn unit to support wound care, regulation of temperature and nutritional support. Cultures from intact bullae are

of little benefit as the fluid is sterile.[3] In a hospital setting, consider treatment of S. aureus carriers with topical mupirocin in the nares.[1]

In addition, wound care with non-adherent dressings may be useful to promote healing of lesions and prevent loss of heat and/or exposure to infectious agents.[2]

While desquamation can continue for another 5 days, re-epithelization will soon follow allowing the skin to heal within a period of 1-2 weeks. Children generally have a good prognosis with a mortality rate </= 4%. Neonates and young infants may be particularly susceptible to complications and therefore would benefit from close monitoring.[3]

Indications for Referral:

- Burn Center
  - Consider assessment by your local burn center if at risk of dehydration, sepsis, extensive TBSA involvement or requiring intensive wound care.[1,2]
  - Follow your local burn center guidelines.
- Dermatologist
  - Early diagnosis and treatment is beneficial to prevent complications therefore if patient's present atypically or there is uncertainty about the diagnosis consider referral to a dermatologist and/or infectious disease specialist.[3]
  - Additionally, in patients who do not respond to empiric treatment consider referral.[3]
  - In patients with extensive disease or widespread involvement at risk of complications, refer for inpatient management for close monitoring and parenteral fluid resuscitation and antibiotic therapy.

Now going back to the case:
While examining Annie, you determine that she is unwell and may have SSSS and would benefit from emergent treatment therefore you promptly alert your preceptor. Your preceptor confirms your suspicion of SSSS. You assist your preceptor in writing a succinct summary of her presentation and past medical history and inform Annie's mom to bring her directly to the local emergency department for further investigations, intravenous hydration, and antibiotic therapy. Obviously if you thought Annie was unstable (low blood pressure, tachycardia or altered LOC) you would transport via EMS.

Conclusion:

Now to summarize, here are few key take home points:

1) SSSS is a relatively uncommon, however potentially serious skin infection seen in infants and young children that is characterized by an erythematous rash and flaccid, sterile bullae due to staphylococcal exotoxin production.
2) Children are at risk due to inability to clear the exotoxin and/or lack the toxin-neutralizing antibodies. Other risk factors include renal insufficiency and immunocompromised states.
3) On history, patients may have a prodrome of fever, irritability, and tenderness of the skin. Physical exam findings include a localized erythematous rash that begins in the face and flexural aspects that generalizes and results in flaccid, bullae that then erode. Crusting and fissuring may appear in the perioral and periorbital regions.
4) An important differential diagnosis includes SJS/TEN which characteristically result in more discrete, tense bullae and mucosal involvement.
5) Treatment includes antimicrobial therapy with anti-staphylococcal activity - in milder outpatient cases oral cephalexin or cloxacillin +/- clindamycin may be prescribed.
6) Early recognition and accurate diagnosis is imperative to prevent complications such as septic shock, dehydration and electrolyte imbalances. Consider referral to a dermatologist, infectious diseases specialist and/or burn center based on clinical suspicion or after non-response to empiric treatment.

Thank you for listening to our PedsCases podcast!

References:

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