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Case report

STAPHYLOCOCCAL SCALDED SKIN SYNDROME

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ABSTRACT

Staphylococcal scalded skin syndrome (SSSS) is skin disease which primarily affects children from newborns up to 5 years of age. When occurring in adults it is accompanied by immunodeficiency. In children, SSSS is usually with good prognosis and resolves with treatment completely, with low mortality, while in adults regarding underline disease mortality is up to 50%. Differential diagnosis includes Steven-Johnson syndrome, Epidermolysis bullosa (EB) and toxic epidermal necrolysis. In this article, we present 8 months old infant with SSSS caused by Staphylococcus aureus strain sensitive to penicillinases. Therapeutic and diagnostic challenges are discussed, together with differential diagnosis and therapy. Successful results are achieved in our case due to the good response to antibiotic and rigorous wound treatment in sterile conditions.

Keywords: Staphylococcal scalded skin syndrome, SSSS,

BACKGROUND

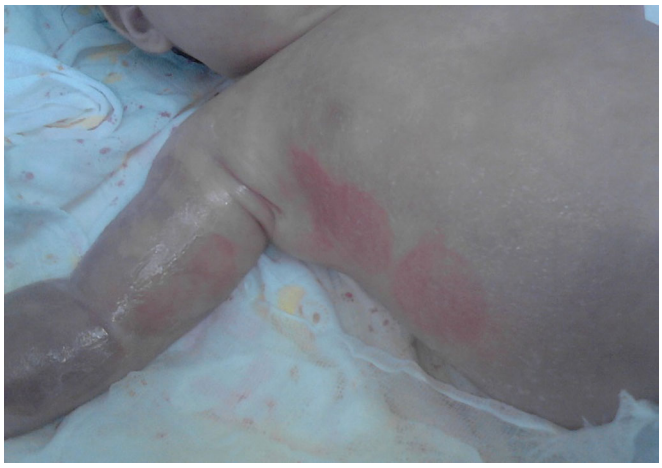
Several skin blistering disorders like Steven-Johnson syndrome, Toxic epidermal necrolysis, Epidermolysis bullosa, pemphigus and SSSS, may have a similar clinical presentation but they differ in etiology, morbidity and mortality, as well as treatment options. Thus, an accurate and timely diagnosis is very important.

CASE PRESENTATION

We present 8 month old female infant with the blistering disorder. The child had no previous history of recurrent infections, allergic disease or known contact with infected individuals. She was breastfed and received regular immunization.

She was admitted to our department with a history of persistent crying 5 days prior to hospitalization poor breastfeeding and fever. Skin lesions then started at the right forearm and right thorax with erythema and exfoliation with rapid progression on the back of the child. At physical examination, she was febrile, with large areas of the skin with erythema and exfoliation (figure 1). Exfoliation of the skin after rubbing was present. The affection of the mucous membranes was not presented. The infant appeared pale and had mucopurulent secretions in the nostrils. Laboratory tests show leucocyte count of 10,5x109 and C – reactive protein 0, 2 ml/l, within the normal range for the age. Microbiology bacterial testing was performed with a swab from the skin lesions and nasal and pharyngeal cultures. Intravenous blood was taken for hemoculture, proteinogram and blood electrolytes.

Fig. 1. Skin lesions on the right forearm and thorax.



Because of the skin lesions with exfoliation, child's age, positive Nikolsky's sign and no mucosal involvement SSSS was considered as working diagnosis and broad spectrum cephalosporin's where initiated, with intravenous fluids, antipyretics and analgesics (paracetamol), mupirocin locally on the exfoliated skin as well as sterile gazes. Skin swab was negative for bacteria, as well as haemoculture. Nose swab shows the presence of Staphylococcus aureus sensitive to penicillin and cephalosporin.

Another diagnosis was considered as TEN and impetigo contagiosa, but without the involvement of mucous membranes and relatively good general appearance, SSSS syndrome was the most likely diagnosis. Infant remains stable and started to feed regularly the second day of treatment.

Antibiotic treatment was continued for 10 days intravenously. Local treatment with sterile gazes and mupirocin continued until skin started to heal. Serum proteins were normal so there was no indication for plasma or albumins infusion. The infant was in good condition, with no skin lesions when discharged from the hospital at day 11.

DISCUSSION

SSSS is skin disorder which affects mainly children up to the 5 years of age and is more common in developing countries, but it can be spread also in neonatal unites as nosocomial infection [1, 2, 3]. First was described by von Rittershain in 1878 [4]. Exfoliation of the skin is caused by exfoliative toxins type A and B [5, 6] which are excreted only by the small portion of staphylococcal subtypes, mainly phage 1 Staphylococcal subtypes. Some of these subtypes are penicillinases resistant Staphylococcus aureus (MRSA) and are community acquired [7]. There are PCR methods for detecting exfoliative toxins but these methods are not available in many laboratories [8]. Exfoliation of the skin is due to cleavage of desmoglein 1 in stratus granulosum of the skin, where desmoglein acts as cell to cell anchored [9]. Skin lesions are superficial not reaching baseline membrane of the skin, with little or no cell infiltration on biopsy specimens.

Reason, why patients with SSSS are up to the age of 5 years, is because desmoglein 1 content in the skin of young children is relatively small compared with adult skin, and clearance of exfoliative toxins in children which occurs in kidneys are slow. One of the main characteristics of the syndrome is that staphylococci cannot be found in the lesions, but they can be found in other locations like nasopharynx or umbilicus, and their toxins spread by hematogenous dissemination reaching stratum granulosum of the skin.

In children, SSSS is a relatively mild disease and can be cured completely without scaring and complications, and has a mortality rate of 4%. In adults, the mortality rate is up to 50% due to underlined immunodeficiency.

Complications are rear, including pneumonia, sepsis and dehydration. Prompt diagnosis and timely treatment can prevent complications and leads to a halt of further exfoliation of the skin. Managing dehydration, electrolyte dysbalances, local wound care [10] and antibiotics are essentials for the treatment [11, 12, 13]. With rising of the methicillin-resistant strains of Staphylococcus aureus that excrete exfoliative toxins, it is important to change initial antibiotic treatment [14, 15] after the results of microbiological testing or if the further exfoliation of the skin continues. In our case, we have a good response to initial therapy as well as positive microbiologic culture from the nasal swab for methicillin-sensitive Staphylococcus aureus sensitive to cephalosporins. Some clinics suggest giving lactulose to small children with SSSS speculating that some amount of the toxin can be excreted via the digestive tract, bypassing immature kidneys, on the basis of findings that people with chronic constipation have elevated titers of antibodies against Staphylococcus aureus, suggesting higher bacterial load. In our opinion giving laxative to not constipating child with dehydration who has loss of fluids through the skin is not acceptable treatment especially with lake of evidence. Further investigation is needed to make this therapeutically option widely acceptable.

SSSS mimics other blistering and exfoliating skin diseases, as Steven-Johnson syndrome and toxic epidermal necrolysis, where there is involvement of mucosal membranes. Because of the lesions which are deep and go beyond baseline membrane loss of fluids, proteins and electrolyte disbalances are more common. Positive Nikolsky sign is one of the characteristics of the SSSS,

which can be found in other blistering skin diseases like pemphigus vulgaris, Epidermolysis bullosa, and toxic epidermal necrolysis. The diagnosis is often made by exclusions if there is exfoliation, positive Nikolsky's sign, negative skin swab, positive nasopharyngeal culture for staphylococci and lack of mucosal involvement. Biopsy of the skin is useful diagnostic tool but was not performed in our patient.

CONCLUSION

This case report highlights SSSS syndrome and its diagnostic and therapeutic challenges. This syndrome is relatively rear in a developed country, but is still common in the developing world and has a significant rate of morbidity. Raising the community acquired methicillin resistant strains of staphylococcus aureus is something that should be taken in consideration when staring initial therapy. Other therapeutically options as lactulose should be considered in some cases, but there is a lack of evidence of its positive effects due to relatively small number of cases described.

REFERENCES:

1. Paranthaman K, Bentley A, Milne LM, Kearns A, Loader S, Thomas A, et al. Nosocomial outbreak of staphyloccocal scalded skin syndrome in neonates in England, December 2012 to March 2013. *Euro Surveill.* 2014 Aug 21; 19(33). pii: 20880. [PubMed] [Crossref]

2. Davidson J, Polly S, Hayes PJ, Fisher KR, Talati AJ, Patel T. Recurrent Staphylococcal Scalded Skin Syndrome in an Extremely Low-Birth-Weight Neonate. *AJP Rep.* 2017 Apr;7(2):e134-e137. [PubMed] [Crossref]

3. Kouakou K, Dainguy ME, Kassi K. Staphylococcal Scalded Skin Syndrome in Neonate. *Case Rep Dermatol Med.* 2015; 2015: 901968. [PubMed] [Crossref]

4. von Rittershain G. Die exfoliative Dermatitis jungerer Sauglinge. Zent-Ztg Kinderheilkunde. 1878;2:3-23

5. Ladhani S, Joannou CL, Lochrie DP, Evans RW, Poston SM. Clinical, Microbial, and Biochemical Aspects of the Exfoliative Toxins Causing Staphylococcal Scalded-Skin Syndrome. ClinMicrobiol Rev. 1999; 12(2): 224-42.

6. Lamand V, Dauwalder O, Tristan A, Casalegno JS, Meugnier H, Bes M, et al. Epidemiological data of staphylococcal scalded skin syndrome in France from 1997 to 2007 and microbiological characteristics of Staphylococcus aureus associated strains. *Clin Microbiol Infect.* 2012 Dec; 18(12): E514-21. [PubMed] [Crossref]

7. Doudoulakakis A, Spiliopoulou I, Spyridis N, Giormezis N, Kopsidas J, Militsopoulou M, et al. Emergence of a Staphylococcus aureus Clone Resistant to Mupirocin and Fusidic Acid Carrying Exotoxin Genes and Causing Mainly Skin Infections. *J Clin Microbiol.* 2017 Aug;55(8):2529-2537. [PubMed] [Crossref]

8. Ladhani S, Robbie S, Garratt RC, Chapple DS, Joannou CL, Evans RW. Development and Evaluation of Detection Systems for Staphylococcal Exfoliative Toxin A Responsible for Scalded-Skin Syndrome. *J Clin Microbiol.* 2001 Jun; 39(6): 2050-2054. [PubMed] [Crossref]

9. Hanakawa Y, Schechter NM, Lin C, Garza L, Li H, Yamaguchi T, et al. Molecular mechanisms of blister formation in bullous impetigo and staphylococcal scalded skin syndrome. *J Clin Invest.* 2002 Jul;110(1):53-60. [PubMed] [Crossref]

10. Baartmans MG, Dokter J, den Hollander JC, Kroon AA, Oranje AP. Use of skin substitute dressings in the treatment of staphylococcal scalded skin syndrome in neonates and young infants. *Neonatology.* 2011; 100(1):9-13. [PubMed]

11. Patel GK, Finlay AY. Staphylococcal scalded skin syndrome: diagnosis and management. *Am J Clin Dermatol.* 2003; 4(3):165-75. [PubMed] [Crossref]

12. Aydin D, Alsbjørn B. Severe case of staphylococcal scalded skin syndrome in a 5-year-old child - case report. *Clin Case Rep.* 2016 Mar 12;4(4):416-9. [PubMed] [Crossref]

13. Handler MZ and Schwartz RA. Staphylococcal scalded skin syndrome: diagnosis and management in children and adults. *J Eur Acad Dermatol Venereol.* 2014 Nov; 28(11): 1418-1423. [PubMed]

14. Lamanna O, Bongiorno D,

Bertoncello L, Grandesso S, Mazzucato S, Pozzan GB, et al. Rapid containment of nosocomial transmission of a rare community-acquired methicillin-resistant Staphylococcus aureus (CA-MRSA) clone, responsible for the Staphylococcal Scalded Skin Syndrome (SSSS). *Ital J Pediatr.* 2017 Jan 6;43(1):5. [PubMed] [Crossref]

15. Braunstein I, Wanat KA, Abuabara K, McGowan KL, Yan AC, Treat JR. Antibiotic sensitivity and resistance patterns in pediatric staphylococcal scalded skin syndrome. *Pediatr Dermatol.* 2014 May-Jun;31(3): 305-8

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