



Case Series

Staphylococcal scalded skin syndrome – A case series with review of literature

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Abstract

Staphylococcal scalded skin syndrome (SSSS) is an acute dermatological illness which requires prompt treatment. It is a condition associated with widespread exfoliation of skin caused by *Staphylococcus aureus* (SA). The toxins elaborated by these gram positive microorganisms especially the exfoliative toxins A and B causes the SSSS. Literature review mentions that only 5% of SA produces these exfoliative toxins. The main route of spread of the toxins is by the hematogenous spread and the process results in extensive damage to the epidermis. This case series reports the SSSS in two children and highlights the significance of promptly diagnosing this serious pediatric dermatological illness.

Key words

Staphylococcal scalded skin syndrome, *Staphylococcus aureus*, Exfoliative toxins, Desmoglein-1, Nikolsky's sign.

Introduction

Staphylococcal scalded skin syndrome (SSSS) is a clinical dermatological condition associated with extensive desquamation caused by the gram positive pathogen *Staphylococcus aureus* (SA) [1]. SA elaborates many extracellular proteins which makes the SA virulent and the exfoliative toxin (ET) produced by them causes SSSS. ET, a major virulence factor of SA acts as enzymes-proteases to cause SSSS by targeting the protein desmoglein-1 (Dsg 1) which results in the

desquamation of the epidermis [2, 3]. Literature review highlights that SSSS is commoner in the paediatric population when compared with the adult population [4]. This article reported case series of SSSS and its significance.

Case series

Case - 1

A two and a half year old male child was seen in the out-patient clinic. He had redness of skin in the right side of neck, gradually extending with



associated high grade fever of two days duration. There was no past history of similar illness. There were few pyodermatous lesions in both the legs and the parents had applied antibiotic ointment. The skin lesions were itchy and not associated with conjunctivitis. Other symptoms such as running nose, cough, wheezing, discoloration of urine and dysuria were absent. He had normal milestones of development and was immunized to date. On general physical examination, the child was febrile-102⁰F, heart rate 92/minute, and respiratory rate 36/minute. Blood pressure was 100/60 mm Hg in right upper limb. All peripheral pulses were equally felt. There was no dehydration, but he was not very active. The child was irritable and was having body pain when he was held in the hands of the parents. He was noticed to have erythematous appearance of the skin in the right side of the neck in the submandibular region (**Photo - 1A**), similar lesions around the mouth, nose, and upper chest. There was perioral desquamation of the skin and also in the neck. Perioral crusting was seen. Nikolsky's sign done was positive on the upper chest. The oral mucosa was normal. There were multiple lymph nodes in the cervical and submandibular region. An initial diagnosis of SSSS was considered along with lymphadenitis. Laboratory investigations such as hemogram revealed Hb (12 g/dl), PCV (33.7%), total count (22000/ μ l), neutrophils (77%), lymphocytes (18%), eosinophils (3 %), monocytes (1.0%), basophils (1.0%), ESR (61 mm/hr), and platelets count (210000/ μ l). Urine examination was normal.. Blood culture sensitivity was send. The child was admitted and was given symptomatic treatment for fever and started on intravenous antibiotic Ampicillin and Cloxacillin combination (50 mg/kg/dose) 6th hourly for seven days, mupirocin ointment applied on the peeling ,desquamating skin lesions and pyoderma, he was well hydrated with home available fluids. In the next days the lesions were spreading to the

hands, thighs, legs and back. The child was ill and appeared toxic. All his systemic examinations were normal. The blood culture and sensitivity reported as sterile. The skin lesions gradually subsided by the sixth day and fever subsided. There was marked desquamation all over the body. The parents were reassured. Fortunately the child did not have any secondary infection. The child was discharged on the day 8 after seven days of intravenous antibiotics with advise to maintain good hygiene and continue oral antibiotic Ampicillin and Cloxacillin combination (50mg/kg/dose 6th hourly) for seven more days. Coconut oil bath was advised for the child. The child has come for follow up and is doing well.

Case 2

A three and a half year old male child was seen in the out-patient clinic. He had normal milestones of development and was immunized to date. His parents were feeling that he was crying on holding the left shoulder since the previous day. He was not playful and appeared lethargic. The child was afebrile, heart rate 82/minute, respiratory rate 36/minute. Blood pressure was 108/66 mm Hg in right upper limb. All peripheral pulses were equally felt but erythematous hue which was more in the face and chest was noticed. There was no itching and no conjunctivitis. He was not allowing lifting him in the arms and appeared suffering from generalized pain. The suspicion of an exanthematous viral fever was high but a possibility of an SSSS was suspected. He had a hemangioma in the left side of the neck with lot of miliaria lesions. There was no itching. The parents were not ready for admission and were advised to review the very next day in the outpatient. He was advised to be given paracetamol suspension. But he developed high grade fever and desquamating skin lesions in the neck by next day morning. He also had fluid filled blisters in the back of the neck and back.

These blisters were breaking when the child had pressure on his back during turning around in the bed and he was having pain. On general physical examination, the child was febrile-102⁰F, heart rate 92/minute, and respiratory rate 36/minute. Blood pressure was 108/66 mm Hg in right upper limb. All peripheral pulses were equally felt. There was no dehydration. He was in distress due to the generalised body ache and itching. Nikolsky's sign done was positive in the neck, on the upper chest and back. He was shown to the orthopaedic surgeon to rule out any synovitis in the left shoulder joint and was reported as mild synovitis and a sling was given in the left shoulder. The child was admitted and treated with the diagnosis of SSSS. Laboratory investigations such as hemogram revealed Hb (10 g/dl), PCV (32.7%), total count (19000/ μ l), neutrophils (74%), lymphocytes (21%), eosinophils (4%), basophils (1.0%), ESR (54 mm/hr), and platelets count (225000/ μ l). Results of the urine routine analysis indicated albumin (nil), sugar (nil), pus cells (1-2/hpf), epithelial cells (+/hpf), bile salt (negative), bile pigment (negative). Blood culture sensitivity was send. He was given symptomatic treatment for fever and was started on intravenous antibiotic Ampicillin and Cloxacillin combination(50 mg/kg/dose) 6th hourly for seven days, mupirocin ointment applied on the peeling ,desquamating skin lesions, he was well hydrated with home available fluids. In the next days the lesions were spreading to the upper and lower limbs, gluteal region and back. Perioral crusting increased. The child was ill and appeared toxic, with swelling and puffiness of face. He had an Ultrasonogram of the left shoulder joint and was normal. All his systemic examinations were normal. The blood culture and sensitivity reported sterile. The skin lesions gradually subsided by the sixth day and his fever subsided. There was markedly desquamation all over the body. The parents were reassured. Fortunately the child did not have any secondary

infection apart from the early synovitis of the left shoulder joint. The child was discharged on the day 8 (**Photo - 1B**), after seven days of intravenous antibiotics with advise to maintain good hygiene and continue oral antibiotic Ampicillin and Cloxacillin combination(50 mg/kg/dose 6th hourly) for seven more days. Coconut oil bath was advised for the child. The child had also come for follow up and is doing well.

Photo - 1A: Photo of child in case 1 on day 1 of admission with perioral crusting, desquamation in the neck and upper chest, erythematous appearance of neck and chest.

Photo - 1B: Photo of child in case 2 on day 8 after admission with redness of skin in the face and neck after desquamation, facial puffiness especially below eyelids.



Figure 1



Discussion

Staphylococcal scalded skin syndrome (SSSS) or Ritter's disease is an exfoliative dermatitis caused by the gram positive microorganisms, *Staphylococcus aureus* (SA) [1-4]. SA is virulent as it elaborates many extracellular proteins. They are toxic shock syndrome toxin 1, enterotoxins, and exfoliative toxins (ETs) [2, 3]. ET is also known as the epidermolytic toxins and has an important implication in the causation of serious dermatological illnesses. ET is the major virulent factor of SA. ET exists in three isoforms. They are ETA, ETB, and ETD. ET is actually glutamate-specific serine proteases enzymes. They are capable of specifically cleaving a single peptide bond in the extracellular region of human desmoglein 1 (Dsg 1), a desmosomal cadherin-type cell-cell adhesion molecule. Dsg 1 is present only in the superficial layers of the skin and this cleavage leads to the separation of the epidermis beneath the granular cell layer [2, 3]. The isomers of ET are serologically distinct molecules. SA strains with the isomer ETA cause bullous impetigo. SA with ETB is obtained from patients with SSSS. In patients with deep pyoderma ETD positive strains have been isolated [3, 4].

SSSS typically is seen in the paediatric population. Neonates are also susceptible for developing SSSS [5]. Many risk factors are mentioned to be associated with SSSS and are more pronounced in the adults. Patients with malignancies especially lymphomas, on immunosuppressant therapy, autoimmune diseases, Human immunodeficiency viral infection and renal diseases are the risk factors to develop SSSS [1-6]. Infants and young children are prone to SSSS because of their immature renal clearance mechanisms. This paves the way to ineffective clearance of ET from the body. This mechanism along with a lack of protective antitoxin antibodies causes SSSS in them [6]. There are characteristic clinical manifestations

of SSSS. The children are having low to high grade fever, with lethargy and malaise at the onset of SSSS. They are irritable, experience severe itching and are poorly feeding. This is followed by the erythematous appearance. There is formation of fluid filled blisters on the body which break on pressure and severe body ache. Both the children in the case series had these presentations. There is peeling of skin around the mouth (with crusting), neck, groin and the gluteal region. The mucosal areas are not involved and there can be puffiness of the face especially below the lower eyelids. The NS is done to assert the presence of SSSS in conditions of absence of desquamation. It should be performed by exerting tangential mechanical pressure on several erythematous zones and it is positive if mechanical pressure induces epidermal detachment [4, 7]. NS was positive in both the cases presented.

The differential diagnosis of SSSS includes many skin conditions such as toxic epidermal necrolysis (TEN) / Steven Johnson Syndrome (SJS), epidermolysis bullosa, bullous erythema multiforme, or listeriosis. Chemical and thermal burns are also included. Bullous impetigo (BI) is a localized form of SSSS which occurs only at the sites of infection with SA and is not generalized. Compared with BI, the skin lesions of SSSS are larger. The characteristic features of SSSS are perioral involvement, desquamation and absence of any mucosal involvement. In contrast, the TEN (or the Lyell's syndrome) is characterised by very severe involvement of visible mucosa and also the mucosae of respiratory, gastrointestinal, and urinary tract systems mucosae. NS is not specific for TEN or SJS [8, 9]. The skin lesions of SSSS are larger than that in BI. The diagnosis of SSSS is mainly clinical and the clinician should have high degree of suspicion especially in the early SSSS. In both cases presented, a clinical diagnosis could be made of SSSS. Clinical features along with a



positive Nikolsky's sign can clinch the right diagnosis. The mucosal surfaces in both the children were not involved and the diagnosis could be directed to a SSSS. The case series had children below five years and SSSS is characteristically seen in this age group in literature review [1-7, 10]. SSSS is generalised and routine blood investigations reveals a polymorphonuclear response. Blood culture and sensitivity is usually sterile and also the blister lesions are also often sterile as in the cases presented. PCR for toxin-encoding genes or random amplified polymorphic DNA analysis are being done in centres with the appropriate facilities. Once the clinical diagnosis of SSSS was made, treatment was started immediately in both cases with the intravenous antibiotics as is recommended for SSSS and other infections caused by SA [1-5, 7]. Here in both cases intravenous antibiotic Ampicillin and Cloxacillin combination (50 mg/kg/dose) 6th hourly for seven days was given followed by oral administration. It is very important to maintain the normal body temperature and protect from secondary infections as the denuded skin after the blisters burst and desquamate is a good portal for entry of microorganisms. Fluid loss can be corrected by proper rehydration orally. It is important to look out for complications of a SA infection and secondary infections such as pneumonia, empyema, septic shock, synovitis and others by doing appropriate investigations such as a Chest X-ray, Ultrasonogram [10]. In the cases presented both the children responded to the prompt treatment given and did not have any complications, except for a mild synovitis in the case 2. The prompt treatment for SSSS with appropriate antibiotic and supportive therapy has led to the mortality rate in children with SSSS not exceeding 5% [11]. The presence of complications can alter the mortality rate in these patients. In adults with SSSS the mortality rates are higher [6, 10, 11]. The extent of skin involvement also has a prognostic role as is

mentioned in some studies [8]. SSSS in patients after severe burns have been mentioned and stresses the importance of aseptic precautions and infection control in such patients [12].

Conclusion

This case study concluded that awareness of SSSS, especially among the Pediatricians and treating Physicians is very important. SA is a common cause of many infections and it is vital to diagnose SSSS very early itself. Every child with SSSS should be closely monitored for presence of various associated complications and secondary infections. Future studies on SSSS should be aimed at detecting its prevalence in different population and regional variations.

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