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Case Report

Internal Medicine Section

A Case Report of Staphylococcal Scalded Skin Syndrome with Toxic Shock Syndrome

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ABSTRACT

Toxins produced by bacteria can result into various systemic manifestations through the release of cytokines which leads to changes at the molecular level. This index case of an 18 years old adult immuno-competent male patient, presented with high grade ongoing fever for 3 days, had wound on left arm in deltoid region. there were diffuse maculo-papular rash predominantly in both palms feet and trunk, erythematous and blanchable on examination. Patient was treated with inj. Piperacillin- Tazobactam and Linezolid, i.v. fluids and nor-adrenaline infusion. Patient was diagnosed with concurrent Staphylococcal Scalded Skin Syndrome (SSSS) and Toxic Shock Syndrome (TSS). The two syndromes are caused by different toxins produced by different strains of *Staphylococcus aureus*. The source of infection was a pus discharging sinus on left arm. The findings were proven on skin biopsy and met the case definition criteria of TSS. He is currently asymptomatic after the treatment being given against Methicillin-resistant Staphylococcus aureus (MRSA) isolated from the pus. It is therefore possible for a single strain of bacteria being capable of producing different toxins, resulting in two different clinical syndromes having a concurrent presentation.

Keywords: Bullous skin lesions, Infectious diseases, Methicillin resistant staphylococcus aureus, *Staphylococcus aureus*

CASE REPORT

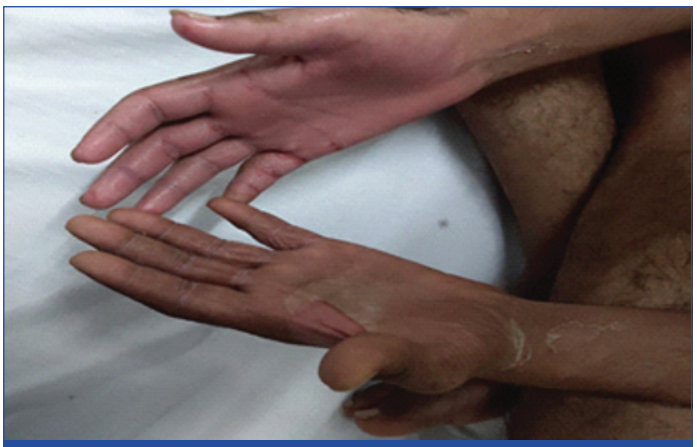
An 18-year-old male, not a known case of any systemic illness, presented to the Emergency Department with a history of high grade ongoing fever for 3 days, generalised erythematous rash initially involving both palms and progressed subsequently to involve the whole trunk and all the limbs. It was associated with generalised weakness and myalgia for last 2 days. There is a history of a bite by an un-identified organism (described by a hindi word 'keeda') 15 days back on left upper arm. The patient was not sure of the incidence and stated, that it might have occurred during sleep. There was no significant history of any drug intake in the past.

On examination, patient was conscious and the well-oriented, had cold peripheries, oral temperature of 102°F, low volume pulse with rate of 100/min, respiratory rate of 22/min and a blood pressure of 70/40 mmHg measured in right arm in supine position. There was a 1×1 cm wound on left arm in the deltoid region with overlying red colored scab. There was diffuse maculo-papular rash predominantly in both palms, feet and trunk; which were erythematous and blanchable on examination.

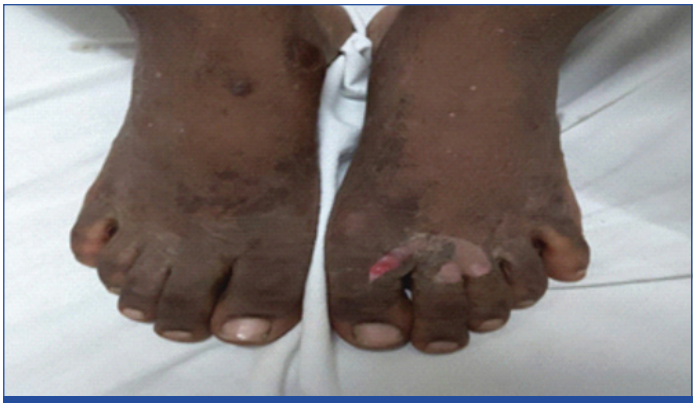
A provisional diagnosis of skin and soft tissue infection with secondary sepsis and septic shock was kept at this stage. Patient was treated with Injection Piperacillin-Tazobactam and Linezolid, intravenous fluids and noradrenalin infusion. Management was provided in intensive care unit for 7 days.

The scab on wound of the left arm was asymptomatic till day 5 of admission, when it suddenly ruptured to form a pus discharging sinus. The pus was sent for examination followed by subsequent surgical debridement and daily dressing. Pus culture report showed an isolation of MRSA sensitive to vancomycin, linezolid and tigecycline. The erythematous rash started to subside after day 6-7 of illness. On day 7, the patient was afebrile and blood pressure was 112/76 mmHg without inotropic support. At this stage, Injection tigecycline was added at a dose of 50 mg intravenously 12 hourly, to the above treatment.

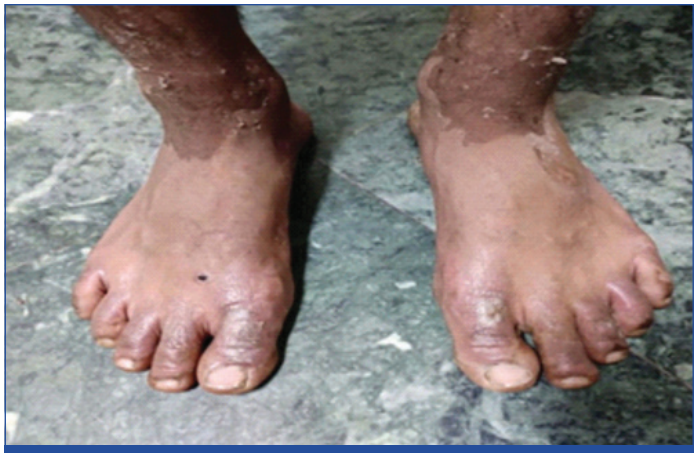
He was then shifted out to general medical ward on day 10, when he started developing fluid filled bullae only on palms and dorsum of bilateral feet (unlike the previous rash); Nikolsky's sign was positive [Table/Fig-1-3]. It was managed with liquid soft paraffin which was applied locally twice daily. Subsequently, whole of the back and trunk were also involved. Skin biopsy taken from the edge of the bulla on the dorsum of left foot, showed subcorneal



[Table/Fig-1]: Palm of left hand showing ruptured bulla with right hand showing post-desquamation skin.

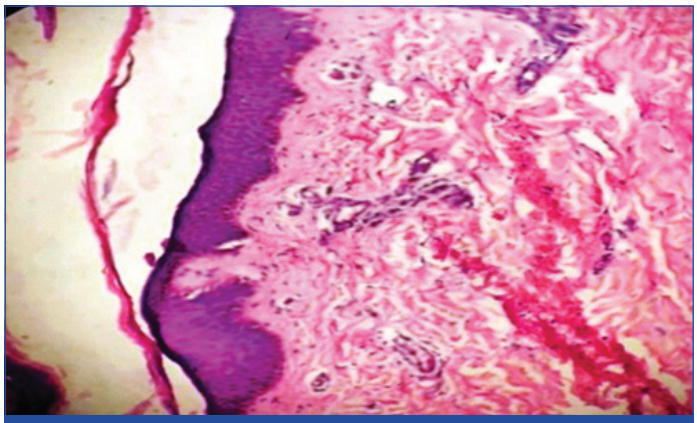


[Table/Fig-2]: Left foot showing similar ruptured bulla from the edge of which skin biopsy was taken.



[Table/Fig-3]: Bilateral feet showing shiny denuded skin after the rupture of bulla and subsequent desquamation.

acellular bulla with inflammation of the underlying papillary dermis [Table/Fig-4].



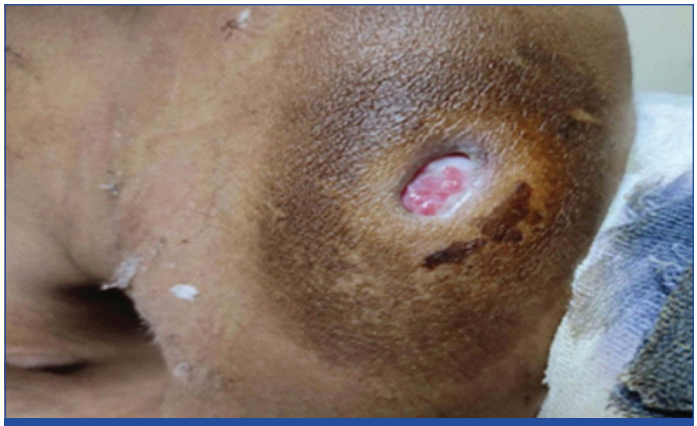
[Table/Fig-4]: Skin biopsy slide taken from the edge of the bulla on the foot (H&E stain at 40X) showing subcorneal cleavage producing an acellular bulla; with underlying inflammatory infiltrate in papillary dermis, confirming the diagnosis of Staphylococcal Scalded Skin Syndrome (SSSS).

The leukocyte count and kidney function tests started improving at around day 11 and the antibiotics were continued for a total of 14 days [Table/Fig-5]. The pus discharge from wound on the arm had also stopped by the time [Table/Fig-6].

	Day 1	Day 4	Day 8	Day 11
Haemoglobin(g/dL)	12.3	11.8	11.9	12.2
Total leukocyte count	27,000	18,800	15,370	5200
DLC (mm^3)	89% neutrophils 6% lymphocytes 4% monocytes	78% neutrophils	76% neutrophils	68% neutrophils
Platelet count (/dL)	60,000	28,000	50,000	1.1 lakh
Blood Urea (mg/dL)	70	66	40	32
Serum creatinine (mg/dL)	1.2	1.4	1.1	0.8
Total bilirubin (mg/dL)	0.8	-	-	-
AST(IU/L)	32	-	-	-
ALT (IU/L)	30	-	-	-
ALP (IU/L)	56	-	-	-
CPK-total (U/L)	664	430	220	114
CPK-MB (U/L)	34	14	20	-
HIV	Non- reactive	-	-	-
Dengue serology	Non- reactive	-	-	-
Peripheral smear for malarial parasite	Negative	-	-	-

[Table/Fig-5]: Sequential laboratory parameters for the patient.

He was completely asymptomatic with patchy areas of desquamation on back, and was subsequently discharged on day 15. Patient has been on regular follow-up since then with no fresh skin lesions or any other complaints.



[Table/Fig-6]: The wound on left arm where the insect bite occurred.

DISCUSSION

Approximately, 5% of *Staphylococcus aureus* strains produce exfoliative Toxins- A and B. The toxins have serine protease activity against Desmoglein 1, a type of adhesion glycoprotein of epidermis. Such dehiscence of the epidermis can result in SSSS in susceptible individuals [1-3]. SSSS mostly occurs in children under the age of 6 years [4,5]. It occurs rarely in adults, mostly seen in the setting of impaired kidney function, immuno-suppression and other comorbidities like diabetes mellitus [6]; with the source of toxins being endocarditis, arthritis, urethritis or the local infections involving conjunctivae/nares. A very high toxin load could explain its occurrence in an immuno-competent patient with normal renal function, as in the index case.

SSSS presents as diffuse blanchable erythematous rash initially involving skin folds and creases, followed by formation of sterile bullae and subsequent rupture; mucosa is spared. Nikolsky's sign is positive. It is associated with fever, skin pain and other constitutional symptoms. The index case had similar clinical presentation, with sparing of mucosa and findings being subsequently proven on skin biopsy.

Some strains also produce enterotoxins like Toxic Shock Syndrome Toxin-1 (TSST-1), which crosses the mucous membrane to enter systemic circulation and result in a disease with multisystem involvement, TSS [1]. TSS presents as rapidly developing hypotension, variable dermatological manifestations, multiorgan system failure in form of raised CPK levels, acute kidney injury, gastro-intestinal symptoms and the most common being profuse diarrhea. Staphylococcal TSS is defined in 2011 by the United States Centers for Disease Control and Prevention [7]. The index case presented with fever, hypotension, deranged renal function tests, raised CPK levels with the MRSA isolate on pus culture; hence, fulfilling the TSS criteria.

Concurrent presentation of SSSS with staphylococcal TSS is rare manifestation [8]. Only two such case reports have been published as yet, to the best of our knowledge [8,9]. A case of patient with liver cirrhosis presenting with concurrent SSSS and TSS was published in Japan in 1994 [9]. Another case report published in 2008 showed a patient with similar presentation, after the patient had undergone a tooth extraction. There was co-existence of the two syndromes in this immuno-competent individual, despite normal renal function [8]. Although in this case, various investigations to isolate the toxins responsible for the two syndromes could not be performed; the case definition of both the syndromes is essentially fulfilled. The case report highlights the co-existence of the two syndromes; with all the clinical findings fulfilling the case definition of each of the syndromes [1,7]. The findings were being subsequently proven on skin biopsy.

Management of both the syndromes requires intensive and targeted antibiotic therapy, surgical debridement of the wound and supportive

care including inotropes [10]. In this case too, the patient responded to the above lines of treatment and was subsequently discharged in a stable condition.

CONCLUSION(S)

It is possible for a single micro-organism to produce different toxins simultaneously to cause two concurrent syndromes. A case of fever with hypotension and diffuse erythematous rash can have innumerable differential diagnoses; a close look into the history, clinical examination and investigations can help with the effective management of the disease.

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