Psoriasis induced toxic shock-like syndrome mimicking hemolytic uremic syndrome

Rafay Khan, Nneka Iroka, Hiyam Ibrahim, Nikunjkumar Patel, Shuvendu Sen, Abdalla Yousif

ABSTRACT

Introduction: Over the past decade the incidence of invasive group A streptococcal infections causing streptococcal toxic shock syndrome (STSS) or toxic shock like syndrome has been increasing. Group A streptococcus (Streptococcus pyogenes) can cause several different conditions including simple cases such as pharyngitis to severe cases involving bacteremia.

Case Report: A 37-year-old female with history of dilated cardiomyopathy and psoriasis presenting with multiple episodes of watery diarrhea, multi-organ dysfunction, septic shock with an unclear underlying etiology later found to be secondary to toxic shock-like syndrome.

Conclusion: The mortality rate associated with toxic shock-like syndrome has been found to be high and a patient normally presents with signs of high fever, hypotension, liver injury, acute renal failure, coagulopathy, and at times soft tissue infection. It can be a difficult diagnosis to make as it can involve multi-organ dysfunction and as cultures are not readily available at time of presentation; its awareness needs to be differentiated to that of other conditions such as hemolytic uremic syndrome (HUS).
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Keywords: Hemolytic uremic syndrome, Psoriasis, Toxic shock-like syndrome, Streptococcal pyogenes

INtRODUCTION

The mortality found in patients with STSS has been found to be increasingly rising with a documented percentage ranging anywhere between 30–85% [1–2]. These patients normally present with signs of fever, hypotension, hepatic dysfunction, kidney injury, and coagulopathy. However, patients with hemolytic uremic syndrome can present in a similar fashion involving symptoms of fever, anemia, thrombocytopenia, acute renal failure, and at times also neurological changes. However, the exact pathogenic mechanism and underlying theory behind these two conditions has not been fully detailed and explained. Thus, it can become difficult for a physician to diagnose and manage these patients as cultures are not readily available upon admission. Furthermore, no cases have reported similar findings in Ashkenazi Jews with a long-standing history of psoriasis and whether a correlation or genetic link exists between these conditions has yet to be examined.
CASE REPORT

A 37-year-old Ashkenazi Jewish female with a past medical history of dilated cardiomyopathy and gastric bypass presented to the emergency room with complaints of generalized weakness for the past few days. She stated that she had a cheeseburger from a fast food restaurant three days prior, and soon thereafter had about five episodes of watery diarrhea per day. The diarrhea began four hours after eating at the fast food restaurant. Her mother who also ate from the same restaurant did not experience the same symptoms. The patient’s only other complaints were multiple episodes of dry heaving associated with nausea, but denied any abdominal pain, vomiting, constipation, melena, or hematochezia. She stated that she had skin lesions throughout her body for several years and was diagnosed with psoriasis and had thus been treated accordingly. She states her skin lesions (Figure 1) have been biopsied in the past and diagnosed as psoriasis. Although they were evident on examination were not new, unchanging, and were distributed throughout her upper and lower extremities. Previously, the patient has been placed on methotrexate, however for the past year she has only been using topical corticosteroids. She denies irritation or any manipulation of the lesions.

On physical examination her blood pressure was 60/34 mmHg, heart rate of 122, respiratory rate of 22, temperature of 104.1, and was saturating 99% on room air. Pertinent findings on physical examination demonstrated a lethargic middle-aged female, with dry oral mucosa, decreased skin turgor, with tachypnea and vesicular breathing. She had weakness in all limbs about a 4/5 in strength and significant anasarca with +3 pitting edema in all extremities. Extremities also had the presence of a diffuse discoid rash with multiple individual widely separated lesions without active bleeding or discharge.

Initial laboratory data showed hemoglobin of 8.7 g/dL, hematocrit of 26.9%, white blood cell count of 9.8x10^3/μL, platelets of 1.5x10^4/μL, and a bandemia of 37%. Basic metabolic panel had sodium of 123 mEq/L, potassium of 4 mEq/L, chloride of 84 mEq/L, bicarbonate of 18 mEq/L, blood urea nitrogen of 50 mg/dL, creatinine of 3.1 mg/dL, and glucose of 102 mg/dL. Chest radiography was unremarkable and urinalysis was positive for a few white blood cells.

The patient was given three liters of intravenous fluid resuscitation which failed to improve her blood pressure. Code sepsis protocol was initiated and patient was admitted to the intensive care unit. Initially, the patient received one dose of vancomycin and was continued on renal dosing of piperacillin/tazobactam. Her clostridium difficile cultures returned negative. Patient was started on norepinephrine drip to maintain her mean arterial blood pressure.

In lieu of these findings, an underlying cause of hemolytic uremic syndrome with associated multi-organ dysfunction syndrome could not be excluded. However, it remained difficult to determine the exact cause as acute tubular necrosis may have been secondary to shock but thrombocytopenia would otherwise coincide with a picture of hemolytic uremic syndrome (HUS). Abdominal ultrasound failed to demonstrate any signs of significant abnormalities or hydronephrosis. Plasmapheresis for management of HUS was withheld at this time as the diagnosis remained unclear.

Blood cultures returned significant for gram-positive cocci in chains and an infectious disease consultation was placed. Initially without subculture, it was believed to be secondary to enterococcus, which could be a translocation of bacteria from the gut as she presented with diarrhea. Further laboratory investigations revealed an INR of 1.2, PTT of 43, elevated fibrinogen of 531, increased fibrin degradation products at greater than 20, a reticulocyte count of 0.5, and a sedimentation rate of 80. Due to these findings, a diagnosis of HUS was considered to be less likely.

Final subculture returned growing group A streptococcal bacteremia and after further analysis it was determined that it may be secondary to her psoriasis, as there was no findings of pharyngitis or any other source. She was switched at this time to Penicillin G along with clindamycin. Left upper extremity computed tomography was conducted to rule out any underlying fasciitis or gangrene at an area of prominent skin lesions and swelling. CT scan suggested skin thickening and subcutaneous induration of the dorsal-lateral forearm.

Over the remainder of her hospital course, the patient showed signs of improvement with the use of antibiotics. Both her platelet count and creatinine had normalized prior to discharge. Patient would be monitored as an outpatient while completing her course of intravenous antibiotics.

![Figure 1: Lower leg skin lesions demonstrating psoriasis.](image-url)
DISCUSSION

First reported in the 1980s, invasive streptococcal infections were labeled as STSS due to its similarities with staphylococcal toxic shock syndrome (STSS) due to its clinical features including septic shock and multi-organ failure [3]. The diagnostic criteria of STSS were formulated in 1993 and involved the presence of Streptococcus pyogenes, hypotension, along with multi-system involvement involving at least two organ systems [3, 4]. These organ systems can involve coagulopathy, pulmonary, skin or soft tissue necrosis, renal, or hepatic involvement. Due to the involvement of multiple organ systems, it can easily be confused with HUS as it can also involve kidney injury, fevers, anemia, and thrombocytopenia. Thus, as demonstrated in this case it is vital for a physician to keep both in their differentials. The difficulty arises as the patient’s blood cultures require time for analysis. The rapid progression of the disease and the severity of the condition along with its complications require the rapid treatment with antibiotics and as the management differs greatly from HUS, it is important to properly differentiate the two conditions early on.

The mechanism in which STSS arises still remains a relative enigma as many different etiologies vary different from one another have been documented and studied. In contrast to STSS where the site of infection is often a trivial focus or asymptomatic colonization; STSS are associated with severe streptococcal focal infection and bacteremia [5]. The most common underlying causes may involve the skin or soft tissues but can be secondary to the upper respiratory tract such as from pharyngitis or even stem from the female genital tract [6]. Most cases have documented the emergence of STSS from either pharyngitis, skin necrosis, or a portal of entry from rashes such as cellulitis. However, it is unusual to find entry of these organisms causing bacteremia from diagnosed psoriasis, especially in patients with chronic psoriasis that has been treated long-term.

Although the pathogenic mechanism is not fully understood, it is thought to be secondary to streptococcal pyrogenic exotoxins, which are type of superantigens that stimulate proliferation of T-lymphocytes with induction of inflammatory cytokines [6]. These superantigens have an affinity to T cell receptors, which result in increased induction in the number of T cells. It has been studied that it has the capability to produce at least four serologically different types of exotoxins [7–8]. They are however very similar in structure and mechanism of action. Overall, this mechanism may result in hypotension and multi-organ dysfunction. Psoriasis has not been well linked in literature to the formation of STSS, and through this discussion it will demonstrate the importance of properly managing it early but may still be complicated by STSS. Whether there is a link between STSS arising from psoriasis and a genetic link to the Ashkenazi Jewish population has not been well addressed previously in literature.

Due to the severity of the condition, prompt recognition of the severity of its progression requires early diagnosis and management. Although the condition has a fulminant nature and complicated course, full recovery is manageable with proper intervention.

CONCLUSION

Through this case discussion, we illustrate an unusual presentation of streptococcal toxic shock syndrome (STSS) secondary to bacteremia erupting from chronic psoriasis and resulting in a picture, which may easily be confused with hemolytic uremic syndrome (HUS).

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