Fulminant necrotizing fasciitis with toxic shock syndrome and multiorgan failure caused by group A streptococcus (GAS): a case report

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Key words
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Background
Necrotizing fasciitis (NF) is a rare and life threatening infection of the skin and underlying structures. It is a progressive infection leading to soft tissue and muscular necrosis of the extremities, abdomen or perineum. NF is often very difficult to diagnose in the early stage of the disease. One of the most important clues is the pain that is often out of proportion to the injury itself [1]. It can often lead to shock with multiple organ failure with high mortality rate (30 %– 70 %) in spite of aggressive treatment [2].

Case report
A 47-year-old female patient with no co-morbidities was admitted to the Clinic for abdominal surgery for suspected appendicitis. Patient complained of severe right inguinal pain that spread to the right leg. Abdomen was tense, palpated in the area of the ileocecal region. There was a small pale skin lesion on the inside of the right thigh suggesting cellulitis. U idući 6 sati u bolesnice se razvio toksični šok s brzim napredovanjem nekroze mekog tkiva.
mg/L. Her renal and hepatic functions were within reference range.

An indication for an exploratory laparotomy was made by the abdominal surgeon. The patient was preoperatively conscious, hemodynamically unstable and respiratory stable. Intraoperatively no sign of acute appendicitis was found. Redness of the skin on the right thigh looking like cellulitis became more pronounced. The patient was postoperatively admitted to the Intensive Care Unit.

Postoperatively the patient continued to complain of severe pain in the right inguinal region spreading in the right leg up to the thumb of the right foot. She confirmed then that she did a pedicure a few days before. Over the course of a few hours the skin of the right leg was turning violet with the development of large fluid filled blisters (bullae) which is in favour of necrotizing fascitis. The patient was empirically started with clindamycin (900 mg every 8 hours), metronidazole (500 mg every 8 hours) and ciprofloxacin (400 mg every 12 hours).

General condition of the patient was also deteriorating in terms of developing toxic shock syndrome. She was conscious complaining of severe pain in her right leg and after she became tachypneic, and hemodynamically unstable we decided to secure the airway before urgent CT scan was made (Figure 1.). Surgical debridement with incisions was carried out urgently after CT diagnostic procedure.

Control blood investigation revealed total white cell count of 4.8×10^9/L, Neu 69.6%, Lym 21.5%, Mono 0.521%, Eos 0.539%, Baso 0.049%, red blood cell count of 11.2 mmol/l, hemoglobin 115 g/L, platelet 109×10^9/L, CRP 218 mg/L, serum calcium 1.23 mmol/L, CK 756 U/L, CKMB 461 U/L, aPTT 60 s, INR 1.64, lactates 6.9. Right leg incisions were widened by orthopedic surgeon.

Two days after the admission, necrotizing fasciitis was spreading to perineum and abdominal wall (Figure 2). The patient was still on mechanical ventilation with analgesia, FiO2 60%, hemodynamically unstable requiring high doses of vasopressor and inotropic support. Urinary output per hour was 40 ml.

Controlled post-transfusion laboratory blood testing revealed total white cell count of 3.92×10^9/L, Neu 87.8%, Lym 69.6%, Mono 7.9%, Eos 1.21%, Baso 1.68%, RBC 4.36×10^12/L, hemoglobin 115 g/L, platelet 109×10^9/L, CRP 218 mg/L, serum calcium 1.23 mmol/L, CK 11 756 U/L, CKMB 461 U/L, aPTT 60 s, INR 1.64, lactates 6.9. Right leg incisions were widened by orthopedic surgeon.

Two days after the admission the decision to perform right hip exarticulation was made. Despite all measures taken the patient’s condition continued to deteriorate and she developed multiple organ dysfunction syndrome.
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Discussion

Necrotizing fasciitis (NF) is a rare infection of the soft tissue that has high mortality rate and is often unrecognised in its early stage. [3, 4]. Its clinical presentation includes progressive infection of the skin and soft tissues presenting with soft tissue oedema, erythema, skin necrosis and bullae. Making adequate diagnosis can be often very challenging due to paucity of skin findings early in the disease. Because NF first starts in the deep tissue planes epidermal involvement may be minimal at the beginning. This can make it difficult to differentiate from non-necrotizing skin infections and cellulitis. The diagnosis is often extremely difficult and relies on a high index of suspicion. A striking sign which is characteristic for NF is severe pain disproportionate to physical findings. [5] At the site of minor trauma (such as a bruise, strained muscle, or sprained ankle), patients may develop deep infection such as necrotizing fascitis or myonecrosis within 24 to 72 hours, often with no visible break in the skin.

Necrotizing fasciitis type I is usually a polymicrobial infection involving mixed aerobic and anaerobic organisms. It is usually less severe, easier to diagnose and also it affects people with underlying disease. Type II infection is monomicrobial, usually caused by group A Streptococcus (GAS), and followed by Staphylococcus aureus. Unlike type I infections, type II is more aggressive, easily missed and may occur in previously healthy patients. Type III and Type IV infections are caused by marine-related organisms like Vibrio spp. and Candida spp. respectively [6]. Primary or idiopathic NF is principally a monomicrobial infection usually caused by group A streptococcus that most commonly occurs in the extremities [7].

Necrotizing fasciitis caused by GAS can lead to early onset of shock and organ failure defined as streptococcal toxic-shock syndrome (STSS) [8].

In our case the patient, with no underlying disease, was affected by GAS infection presenting at first as severe pain in the right inguinal region mimicking appendicitis. In the next 6 hours after explorative laparotomy she developed toxic shock with fulminant progression of skin and soft tissue necrosis spreading on her whole right leg. Reported pedicle days before could possibly be the cause of site infection. Mortality rate for patients who develop STSS can be higher than 50 % [9].

The treatment includes surgical debridement and early broad spectrum empiric antimicrobial therapy. The use of i.v. immunoglobulin is controversial [10, 11,12] but recent studies show that they can be beneficial in case conventional therapy has failed. [13, 14]

Conclusion

Group A streptococcus infection can lead to necrotizing fasciitis and streptococcus toxic shock syndrome in previously healthy patients with poor outcome. The presence of severe pain, erythema and fever require high index of suspicion for this rare but potentially fatal infection.

References