A woman with a painful and swollen hand

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CASE REPORT

An 86-year-old woman came to the emergency room complaining of a painful swollen right hand for the last 18 hours without a history of trauma (figure 1). At physical examination her blood pressure was 75/40 mmHg, heart rate 60 beats/min and body temperature 37.3°C. Sensibility of the digits was absent. Laboratory investigation showed leukocytosis, raised C-reactive protein, hyperlactataemia and renal insufficiency (serum creatinine 168 µmol/l). Creatine phosphokinase was still normal. During the first hours in the ER, the swelling progressed to the volar side of the right arm with purple colouring and formation of a blister (figure 2).

WHAT IS YOUR DIAGNOSIS?

See page 112 for the answer to this photo quiz.
The combination of acute severe pain, evident progressive swelling within a few hours, purple colouring of the right hand and forearm and the typical blister at the wrist suspicious for luminescent necrosis (figure 2) was suspected to be caused by necrotising fasciitis. Therefore, penicillin and clindamycin intravenously were added to the initially administered ceftriaxone. Although a small surgical incision at admission had not revealed necrosis, a more extensive exploration five hours later showed evident necrosis of the deep muscle loges of the forearm. Debridement was carried out. However, guillotine amputation appeared inevitable nine hours later. The patient died 27 hours after admission from intractable shock. Cultures from the fascia grew haemolytic streptococcus group A, which is the most commonly cultured pathogenic microorganism in necrotising fasciitis after Staphylococcus aureus.

Necrotising soft-tissue infections (NSTI) include necrotising forms of cellulites and fasciitis. NSTIs can be divided in three types. Type I mixed infection is caused by aerobic and anaerobic bacteria occurring most commonly after surgical procedures. Type II is caused by group A β-haemolytic streptococci, primarily affecting the extremities. Type III is associated with Vibrio vulnificus, which enters the subcutaneous tissue via puncture wounds from fish or marine insects. NSTI is a rare infection of the subcutaneous tissue and fascia with a high mortality rate of approximately 20 to 60%.

Establishing the diagnosis is not easy. Clinical findings suggestive for necrotising fasciitis include rapidly spreading oedema, numbness of overlying skin, severe pain out of proportion to skin findings, blister or bullae formation (which is rare in cellulitis and erysipelas), signs of toxic shock syndrome, mental status changes, and the presence of subcutaneous air, if caused by gas-producing organisms. A necrotising infection of an extremity is usually the result of an injury, however in some cases like our patient, no primary cause can be found.

Streptococci secrete exotoxins promoting the immune system's production of TNFα, and interleukins 1 and 6. Beta-haemolytic streptococci produce superantigens which activate CD4 T cells, which then activate complement and clotting factors, leading to shock and multiple organ failure. Broad-spectrum antimicrobial therapy should be administered empirically as soon as possible, and should cover Gram-positive, Gram-negative and anaerobic organisms. When a group A streptococcal infection is suspected, addition of clindamycin is highly recommended because it is believed to inhibit exotoxin production. Intravenous immunoglobulins may be a useful additional treatment in type II group A streptococcal NSTI complicated by toxic shock syndrome. Definite treatment involves early and complete debridement of the infected tissue. Delayed recognition, with consequent massive soft tissue loss and sepsis, remains a deadly pitfall in the management of necrotising fasciitis.

REFERENCES