This new pattern is also different from condensing osteitis of the clavicle, which radiologically is characterised by sclerosis and enlargement of the medial clavicle and histologically by increased amounts of normal bone. The differential diagnosis includes osteoarthritis, osteomyelitis and osteoid osteoma, all of which may involve the lateral clavicle. Osteoarthritis of the acromioclavicular joint is characterised radiologically by osteophytes on the clavicular side and sclerosis on both sides of the joint. A history of infection would suggest osteomyelitis, although the initial radiographs may be normal. As the infection progresses, there is bony destruction and periosteal reaction. Osteoid osteoma presents as a central lucency surrounded by dense sclerotic bone on radiographs. CT may be needed to identify this classic pattern.

It may be that patients who develop aseptic necrosis of the lateral clavicle have some as yet undetermined predisposition, whether anatomical, physiological or both. Once the diagnosis is suspected, and after a trial of conservative measures for a reasonable period, we recommend excision of the lateral aspect of the clavicle as an effective treatment for this condition. The histological examination will confirm the diagnosis, thus allowing the patient to be counselled as to the risk of the development of contralateral necrosis.

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References


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We present a case of fatal ‘malignant’ necrotising streptococcal myositis in a previously healthy 39-year-old man. The infection was caused by Lancefield group-A haemolytic streptococcus (Streptococcus pyogenes). This case highlights the clinical features and the necessity of prompt aggressive treatment.

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We present a case of fatal malignant necrotising streptococcal myositis. This primary bacterial infection of skeletal muscle which is caused by Lancefield group-A haemolytic streptococcus (Streptococcus pyogenes) has an exceptionally high mortality and constitutes a surgical emergency. The paucity of discriminating clinical features in the early stages often leads to a misdiagnosis. This has prompted us to highlight the clinical patterns and the aggressive measures which may be required to avert a catastrophic outcome. If a patient presents with the prodromal symptoms of a fever and pain out of proportion to that expected and particularly if there are signs of a compartment syndrome, this diagnosis should be considered.

Case report

A previously healthy 39-year-old industrial chemist presented with a three-day history of an acutely painful and swollen left knee on which he was unable to bear weight. This had been preceded by a prodromal illness of generalised weakness, pyrexia, diarrhoea and vomiting which had lasted for five days. He had sustained no injury and had not travelled abroad. Examination revealed a fit man with a pyrexia of 38.5°C and in some discomfort. He had no skin rash and no clinical features of septicaemia. A provisional clinical diagnosis of deep-vein thrombosis (DVT) was made on account of a tender and swollen left calf with a difference in girth of 2 cm compared with the unaffected side, and a positive Homan’s sign. A ruptured Baker’s cyst was also considered as a differential diagnosis. Although his knee movements were a little restricted, there were no clinical features suggestive of septic arthritis. Plain radiographs of the left knee and calf were normal, with no evidence of gas in the soft tissues. The white cell count (WCC) and ESR were normal, and treatment with antibiotics was not initiated.

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Doppler ultrasound scanning showed no evidence of DVT, but the deep tissues and muscle planes were oedematous, raising the possibility of an inflammation of the deep muscles. After 14 hours he developed septicaemic shock and was dehydrated and anuric. He was transferred to intensive care for resuscitation. The left calf was tense with blistering of the skin and sloughs. High-dose intravenous penicillin and metronidazole were given. Emergency surgical exploration revealed extensive necrosis of the lower leg with an intact fascial envelope containing necrotic muscle in the quadriceps and adductor compartments. Immediate high transfemoral amputation was performed. Haematological investigations revealed a markedly raised level of creatine kinase (3370 units), metabolic acidosis and disseminated intravascular coagulation. Despite vigorous treatment he died approximately 36 hours after admission. Blood cultures and muscle specimens grew group-A haemolytic streptococcus. Specific anaerobic culture was negative, establishing the diagnosis of necrotising streptococcal myositis with toxic shock.

Discussion

Necrotising streptococcal myositis is a separate entity and particularly aggressive, when compared with milder variants of necrotising streptococcal infections such as pyomyositis and fasciitis. A review of the English literature reveals a few cases, but only two in the orthopaedic literature. Because of the high associated incidence of mortality which was reported in one series to be 85%, a high index of suspicion at its onset may present the only hope of survival.

The principal reason for a fatal outcome in this and other reported cases is misdiagnosis and delayed treatment. The combination of its rarity, the non-specific features at presentation, and the aggressive nature of the infection with an underestimation of its extent, usually delays the administration of antibiotics and surgical debridement, resulting in the patient’s demise.

From earlier described cases a certain uniformity in the clinical pattern of necrotising streptococcal myositis can be recognised. Previously healthy middle-aged patients predominate with a male-to-female ratio of 2:1. Although any muscle groups may be involved the limbs are commonly affected with a predilection for the proximal muscles of the lower limb. The disease is usually of spontaneous onset but there may be a history of trivial trauma. Misdiagnosis is frequent because the onset of symptoms is vague, non-specific and common to many musculoskeletal conditions. The unsuspecting clinician will thus be faced with initiating treatment. Parameters of infection such as the WCC and ESR may be normal, possibly because the speed of spread of infection is too great for the initiation of an inflammatory reaction.

The manifestation of necrotising streptococcal myositis can be classified into an early prodromal stage, a rapid intermediate stage and a variable late stage. The first may last for three to seven days, with non-specific ‘flu-like’ symptoms of general malaise, sore throat, myalgia, arthralgia, nausea, vomiting and diarrhoea with a disproportionate intensity of pain in the involved muscle compartment. There may be a pyrexia or scarlatiniform rash. Common misdiagnoses include simple viral illness, muscle injury and arthritis.

The rapid and aggressive intermediate stage is short and lasts for a few hours. There is an escalation of muscle pain, leading to restriction of movement, muscle spasm, limping, inability to bear weight and systemic toxic features. The site of inappropriate muscle pain progressively swells causing skin changes, from erythema to bluish discoulouration and finally, the formation of blisters containing serous non-purulent fluid. There may be an effusion in adjacent joints. During this stage common misdiagnoses include septic arthritis, rupture of a Baker’s cyst, DVT and cellulitis. Since the fascia covering the muscle is not breached the production of inflammatory products raises the intracompartmental pressure sufficiently to compromise the distal circulation and cause a compartment syndrome and infarction. In retrospect, our patient had early signs of this at presentation and it worsened rapidly. Differential WCCs, specifically looking for a shift to the left and neutrophocytosis, a massively raised serum level of creatine kinase and an urgent ultrasonogram of the involved limb, will help to exclude DVT and the formation of an abscess. The late stage is variable. It may last for a few days, often with a fatal conclusion, or many weeks with resultant severe morbidity and disability. The streptococcal toxic shock syndrome is a common complication leading to multiorgan failure and coagulopathy. At this stage, treatment by amputation represents an attempt to save life which is often not successful.

It has been reported that the in vitro sensitivity of group-A haemolytic streptococci to penicillin may not be effective in vivo. Animal studies reveal that penicillin sensitivity declines if treatment is not instituted within the first two hours of the onset of disease. Clindamycin may be more efficacious, but also has declining efficacy if treatment is delayed. A high index of suspicion at its early stages merits the immediate administration of antibiotics. Despite such treatment, however, further progression to the rapid intermediate stage may be inevitable because of ischaemic myonecrosis resulting from raised compartment pressure. This should prompt the clinician towards aggressive resuscitative measures, which includes the management of septic shock and early surgical exploration with sufficient debridement to save limb and life.

Our patient was characteristically a well-built young and healthy man who was misdiagnosed as having a DVT. The condition progressed rapidly to septic shock and death, despite a high transfemoral amputation and aggressive medical treatment.

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