Group A Streptococcus necrotising myositis of the limbs secondary to cavitating pneumonia

Manveer Rahi 1, Malcolm Will2

SUMMARY
Necrotising myositis is a rare complication of Group A Streptococcus infection requiring early and aggressive surgical management to prevent mortality. However, early diagnosis is difficult due to non-specific initial presentation and a low index of clinical suspicion given the paucity of cases. We highlight these challenges and present a case of a 22-year-old woman presenting with cough, fever and severe limb pain refractory to analgesia during the COVID-19 pandemic. We outline potential confounding factors that can delay intervention and offer diagnostic tools that can aid clinical diagnosis of necrotising myositis. In reporting this case, we hope to raise awareness among clinicians to avoid these pitfalls.

BACKGROUND
Group A Streptococcus (GAS) can cause a spectrum of necrotising soft tissue infections including necrotising fasciitis and, in exceptionally rare circumstances, necrotising myositis.1 Necrotising myositis has a reported survival rate of <1% without surgical intervention.2 Most reported cases of GAS necrotising myositis are due to direct trauma and upper respiratory tract infection. However, to the best of our knowledge, this is the first reported case study of GAS necrotising myositis presenting secondary to a cavitating pneumonia and parapneumonic effusion.

CASE PRESENTATION
A 22-year-old woman presented to accident and emergency (A&E) with a 10-day history of cough, fever, malaise and severe pain in her left upper arm and right leg refractory to analgesia. On examination, temperature was 38.2°C, heart rate was 150 bpm, respiratory rate was 20 and saturations 95% on room air. There was erythema, swelling and tenderness over the upper arm, axilla, distal hamstrings and proximal gastrocnemius. She had preserved movement of the upper limb, although internal rotation was very painful. Peripheral pulses and sensation were normal.

The patient had no significant medical history and was on 6 monthly Depo-Provera injections only. She had no risk factors for immunosuppression and no significant family history.

INVESTIGATIONS
Initial blood tests showed: haemoglobin 157 g/L, white cell count 26.2×10⁹/L, lymphocytes 0.78×10⁹/L, C-Reactive Protein (CRP) 464 mg/L, lactate 4.4. COVID-19 testing was negative. The admission chest X-ray showed opacification in the right upper-mid zone (figure 1). X-rays of the left arm and right knee showed no bony changes or free gas. A CT chest–abdomen–pelvis scan was performed on admission, which showed a cavitating right upper lobe pneumonia and parapneumonic effusion (figure 1). There was extensive soft tissue stranding and thickening of the left shoulder girdle, chest wall and upper arm musculature leading to suspicion of necrotising soft tissue infection (figure 1).

On admission to the intensive care unit (ICU), a transthoracic echocardiogram was performed, which showed normal left ventricular function and no evidence of infective endocarditis. An ultrasound Doppler of the right leg and left arm was performed, which showed no evidence of collection, deep vein thrombosis or Baker’s cyst. Joint aspirates were sent for culture and sensitivity, which showed no growth of organisms and no evidence of septic arthritis. On day 2, admission blood cultures grew GAS.

On day 11 of admission, the patient required extensive myofascial debridement for necrotising soft tissue infection. Fibrofatty pathology specimens were sent following debridement in theatre. The two specimens were taken from the upper right arm and showed fibrinopurulent exudate on the specimens when viewed macroscopically. On microscopic inspection, the specimens were principally comprised of fat with some associated muscle and fibrous tissue. The tissue showed areas of extensive necrosis with large numbers of associated neutrophils. These areas were surrounded by organising granulation tissue, fibrosis and patchy mixed inflammatory cell infiltrate. There was no evidence of granulomatous inflammation or atypia seen.

The following day, the patient underwent thoracotomy and decortication of empyema. Effusion cultures from theatre resulted in no growth of organisms and no evidence of infective endocarditis. The decortication report showed full-thickness infiltration of the sections by neutrophils and cosinophils with granulation tissue, haemorrhage and a reactive fibroblastic proliferation. The features were consistent with inflammatory changes in keeping with the clinical picture of empyema. There was no evidence of malignancy on pathology specimens.

DIFFERENTIAL DIAGNOSIS
On admission, the patient was reviewed by the orthopaedic team and although the Laboratory Risk Indicator for Necrotising Fasciitis (LRINEC) score was 8 (CRP 464, white cell count 26.9×10⁹/L,
Hb 157 g/L, Na 130 mmol/L, creatinine 58 μmol/L, glucose 5.9 mmol/L suggesting necrotising soft tissue infection, they felt this was misleading in the context of a cavitating pneumonia. The working diagnosis for the patient was cavitating pneumonia with possible septic emboli. Initial blood culture results together with the patient’s deterioration despite 10 days of appropriate antibiotic treatment prompted further imaging. The subsequent CT scans confirmed the diagnosis of necrotising myositis on day 10 of admission.

TREATMENT
The patient was transferred to the ICU from A&E with a lactate of 4.4 and hypotension refractory to significant fluid resuscitation with intravenous plasmalyte, requiring treatment with intravenous human albumin solution and intravenous norepinephrine infusion. Additionally, she was treated with intravenous immunoglobulin for presumed toxin producing organism. She was initially treated with piperacillin/tazobactam, flucloxacillin, linezolid and metronidazole for a cavitating pneumonia. On day 2, blood cultures grew GAS. Antibiotics were subsequently rationalised to intravenous benzylpenicillin and clindamycin. On day 6, she required an urgent chest drain insertion for massive pleural effusion with significant mediastinal shift.

Ten days after admission, the patient remained feverish with persistently raised inflammatory markers despite antibiotic therapy. Increasing erythema and blistering was noted on the left arm, with the patient requiring increasingly large doses of opiate analgesia for pain control. As such, a CT scan of the right leg and left arm was undertaken, which showed reactive fluid in the fascial tissue planes (figure 1) raising the suspicion of GAS necrotising soft tissue infection. The following day, the patient underwent surgical exploration and due to significant myonecrosis and fat necrosis required fasciotomies of all compartments as well as extensive muscle debridement (figure 2). Histopathology results from theatre showed a pattern indicative of necrotising soft tissue infection (figure 3). The wounds were left open, and the patient underwent further debridement 2 days later. The following day, in order to treat all source of sepsis she underwent a right thoracotomy and debridement/decortication of empyema. The next day, primary skin closure was achieved at all fasciotomy sites.

OUTCOME AND FOLLOW-UP
The patient continued to make good progress and the inflammatory markers improved following the surgeries. She was discharged from intensive care on day 34 and left the hospital on day 40 having completed rehabilitation.

DISCUSSION
This rare case highlights the challenges in early diagnosis of GAS necrotising myositis especially in the context of concurrent severe pneumonia. Non-specific early symptoms and signs together with the rarity of the disease contributed to the delay in diagnosis and treatment. An additional factor in this case was that the patient’s presentation was suspicious of COVID-19 infection and she had been advised to self-isolate, which led to a delay in presentation. However, on the patient’s transfer to our department, one of our cardiothoracic ICU physicians had seen

Figure 1 (A and B) A cavitating pneumonia and parapneumonic effusion on admission chest CT. (C) A reactive fluid collection under the oedematous left deltoid muscle, indicating muscle inflammation (α) and extensive fat stranding and inflammation (β). (D) A collection of reactive fluid in-between the superficial and deep fascial planes of the right leg.

Figure 2 Extensive fasciotomy incisions were performed in left arm (A,B) and the right leg (C).

Figure 3 Histopathology slides (×100 magnification) of left arm specimen from initial fasciotomy showing necrosis (α), fibrosis (β), neutrophil infiltration (γ) and small vessel thrombosis (δ) indicative of necrotising soft tissue infection.
a similar case previously and raised the suspicion of GAS necrotising myositis as a diagnosis.

Non-specific flu-like symptoms are often reported in the early stages of GAS necrotising myositis. Skin changes and muscle pain can be falsely attributed to other infective conditions such as cellulitis or septic arthritis. Pain that is refractory to opioid analgesia is often indicative of extensive soft tissue necrosis and the development of compartment syndrome and may precede toxic shock. Early aggressive surgical management is the mainstay of reducing mortality from severe GAS necrotising myositis, and delays in treatment are associated with high mortality.

GAS necrotising myositis remains a clinical diagnosis, however biochemical and imaging investigation provides diagnostic value. The LRINEC score was developed as a diagnostic tool using biochemical markers to risk stratify the patient for necrotising soft tissue infection. LRINEC has greatest sensitivity in patients with a score ≥6, with a positive predictive value of 92%. However, specificity for necrotising soft tissue infection is low and scoring can be confounded by the inflammatory response created from concurrent infections as seen in this case.

Radiological imaging such as CT and MRI can provide valuable information on the extent of soft tissue inflammation and guide the surgical approach. MRI is recognised as the most sensitive modality for necrotising soft tissue infections but is often difficult to access in the acute setting. Therefore, CT is more widely used with sensitivities reported between 80% and 85% for necrotising soft tissue infection. Radiological signs of thickened, oedematous fascia and deep fluid collections are typical for necrotising myositis. Importantly, unlike in clostridial myonecrosis, or gas gangrene, free gas is not a radiological feature of GAS necrotising myositis.

Within this case, there were several important markers that prompted a change in clinical course towards the diagnosis and treatment of necrotising myositis. In spite of a prolonged course of antibiotics, the patient continued to be febrile and have persistently raised inflammatory markers at day 10. This raised the concern of inadequate sepsis source control and thus prompted the clinical team to seek an additional focus of infection to the cavitating pneumonia. Despite the early orthopaedic review, when necrotising soft tissue infection was felt to be unlikely, there was a need to review the working diagnosis through additional CT imaging in light of enduring musculoskeletal symptoms of pain refractory to strong opioid analgesia and skin blistering. The decision to undertake surgical exploration was made with a high degree of suspicion for necrotising myositis due to the constellation of symptoms, the radiological finding of fluid between fascial planes and fever despite appropriate antibiotic treatment for GAS pneumonia.

Recognition of GAS necrotising myositis requires a high degree of clinical suspicion in patients with non-specific soft tissue infection and an acute inflammatory response. Delayed diagnosis is common among reported cases, which contribute to high mortality. Confounding factors, such as concurrent infections, can make diagnosis more difficult and contribute to delay in investigation and intervention. Biochemical results and radiological imaging offer valuable indicators of necrotising myositis that aid clinical decision making. This report highlights the need for clinicians to be aware of this rare condition and the need for early, aggressive surgical management to achieve good clinical outcomes.

Learning points

- Suspect necrotising myositis in patients with non-specific soft tissue infection and an acute inflammatory response, which is unresponsive to medical treatment.
- Delayed diagnosis of necrotising myositis contributes to high mortality.
- The Laboratory Risk Indicator for Necrotising Fasciitis score and CT scan are valuable tools to aid clinical decision making in suspected necrotising myositis.
- Early surgical management is required to achieve good clinical outcomes.

Contributors

MR and MW were involved in the clinical care for the patient during the reported presentation. MR collated the investigation results, figures and planned the report. MR wrote the initial draft of the report. MW and MR edited the drafts. All authors contributed to final version of the manuscript.

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ORCID iD

Manveer Rahi http://orcid.org/0000-0002-0747-8723

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