Unusual presentation of more common disease/injury

An elderly patient presenting with hip pain following a fall: an unusual presentation of necrotising fasciitis

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Correspondence to Evgenios Evgeniou, evgenios@doctors.org.uk**Summary**

Necrotising fasciitis (NF) is a rapidly progressive soft tissue infection involving necrosis of subcutaneous tissues. Early surgical intervention reduces mortality, but initial clinical findings are often non-specific and can delay the diagnosis. An 80-year-old patient, presented to our emergency department with pain in her left hip and mild bruising following a fall. An x-ray, requested to investigate a possible hip fracture, in fact demonstrated air in the subcutaneous tissues. She rapidly deteriorated and soon developed blood-filled blisters, crepitus and fixed staining of the skin. She underwent urgent debridement of involved tissues in theatre confirming the diagnosis of NF. The presence of subcutaneous emphysema on plain radiograph as in this case, is extremely specific to the diagnosis of NF. Although other imaging modalities can aid diagnosis these remain as an adjunct rather than a definitive diagnostic tool and should not delay surgical intervention based on clinical findings.

BACKGROUND

Necrotising soft tissue infections are rare and are rapidly progressive, often proving fatal. Although the incidence of necrotising fasciitis (NF) is low at approximately 500 cases/year in the UK,¹ death rates can be as high as 76%.² Early surgical intervention has been shown to reduce mortality;²⁻³ however, initial clinical findings are often non-specific and can contribute to delays in diagnosis. This case in particular demonstrates how the presentation can be extremely varied and atypical and therefore a high index of clinical suspicion is needed and any clues to the diagnosis must be noted and acted upon as a matter of urgency.

CASE PRESENTATION

An 80-year-old lady, with a background of insulin-dependent diabetes, presented to our local emergency department with a 1-day history of left hip pain. The pain was insidious in onset but increased in intensity throughout the day and culminated in the patients' leg giving way and being helped to the ground by her daughter. On admission the patient appeared pale and clearly in distress, although cardiovascular parameters were normal (blood pressure 104/60; pulse 82) and clinical examination only identified mild bruising to the medial aspect of the left upper thigh. Initial investigations were requested and the patient was reviewed 2 h later by the emergency team. At this stage, the attending clinician noted extension of the bruising down to the calf with blood-filled blisters and surgical crepitus. There was fixed staining of the skin on the posterior and medial aspect of the left leg.

INVESTIGATIONS

Initial blood tests showed a white cell count of $8.9 \times 10^9/l$, neutrophils of $8.2 \times 10^9/l$, C reactive protein of 51 mg/l and lactate of 4.9 which rose to 7.8 mmol/l within an hour (although the pH on arterial blood gas remained neutral as a result of respiratory compensation). Plain

x-rays demonstrated air in the subcutaneous tissue of the left thigh and hip (figures 1 and 2). Gram staining of debrided tissue sent for immediate histological review showed the presence of Gram-positive cocci, later confirmed as group A streptococcus. The NF was probably caused by the bacteria entering through broken skin attributable to the patients' poor diabetic control. However, due to the often rapid progression of NF, the source of bacteraemia and subsequent sepsis is sometimes difficult to confirm.

DIFFERENTIAL DIAGNOSIS

The provisional diagnosis, based on the history of trauma and pain in the hip was of a fractured neck of femur. However, with the rapid development of severe bruising, subcutaneous crepitus and blood-filled blisters, it became clear that NF was the likely diagnosis. Possible differential diagnoses were cellulitis or subcutaneous abscess; however, the above clinical findings, blood tests and x-rays made a diagnosis of NF almost unequivocal.

Figure 1 Anteroposterior plain radiograph of the left hip demonstrates gas in the subcutaneous tissues with no bony pathology noted.

Figure 2 Lateral plain radiograph of the left hip demonstrates gas in the subcutaneous tissues with no bony pathology noted.

TREATMENT

The patient was started on intravenous antibiotics and was taken directly from accident and emergency to theatre where she underwent urgent surgical debridement of the left leg. Skin from the medial, lateral and posterior aspects of the left thigh and medial and posterior calf were debrided until only healthy, bleeding skin edges remained. Resuscitation was initiated in the intensive care unit involving extensive inotropic support, however, the patients' lactic acidosis continued to rise and although further necrotic tissue was noted, she was deemed too unwell for further surgical intervention, particularly as the first debridement had been so extensive. The patient died of sepsis and multiorgan failure 24 h later.

OUTCOME AND FOLLOW-UP

Despite the gold standard treatment of broad spectrum intravenous antibiotics and extensive surgical debridement being initiated at an early juncture, the slight delay until clinical findings became more apparent meant that the systemic toxicity associated with group A streptococcus infection, had already taken hold. The patients' age and comorbid diabetic status also contributed to her poor prognosis and she died on the intensive care unit after 48 h of substantial resuscitation efforts.

DISCUSSION

NF is a life-threatening and rapidly progressing infection that spreads along fascial planes.⁴ It is usually caused by streptococcal organisms, but often a variety of both aerobic and anaerobic pathogens are grown from culture.^{5 6} In particular group A β -haemolytic strains are believed to cause a more profound systemic sepsis. Production of bacterial toxins and enzymes, such as hyaluronidase, cause rapid progression of the infection through the fascia,⁵ which undergoes liquefactive necrosis. As further bacterial proliferation and necrosis occur, occlusion and suppuration of blood vessels lead to evolving skin ischaemia.⁷

Although a history of trauma is usually present such as a scratch or bite,⁶ often no cause can be established. NF can occur following surgery, intravenous drug use, burns and childbirth, and can complicate soft tissue infections.⁸ Factors

that render patients susceptible to NF include any chronic disease or comorbid condition that could suppress the immune system including malignancy, malnutrition, obesity, immunosuppressive drugs or as in this case, diabetes.^{8 9}

Early signs and symptoms include fever, pain, swelling, erythema, tenderness and cellulitis.¹⁰ Crepitus, blisters and haemorrhagic bullae are late but more focal signs.^{3 11 12} The presence of subcutaneous emphysema on plain radiograph, as seen in this case report, is a very specific finding for NF.¹² One or more of crepitus, blistering and subcutaneous emphysema on plain x-ray can be seen in 85% of cases, and this triad of signs has been suggested as a screening tool.³ Soft tissue air on x-ray alone is not completely sensitive for infection, and NF can be present where subcutaneous emphysema is absent.^{3 13 14} Presentations are variable and while the symptoms and signs listed provide some diagnostic assistance, the most important point to note during clinical examination is severe pain disproportionate to local findings in association with systemic toxicity. Also of note, the skin involved becomes increasingly tense as the erythematous margins become indistinct, and the colour of the skin normally progresses from a red-purple to a dusky blue before the occurrence of bullae/blisters. Symptoms may develop over a period of hours to days and presentations are varied, some patients presenting already confused or agitated due to systemic shock thus increasing the diagnostic difficulty for the clinician.¹⁵

In addition to plain radiography, a variety of imaging techniques have been utilised to assist in the diagnosis of NF, particularly where clinical uncertainty exists.

- Ultrasound: demonstrates features of diffuse thickening of subcutaneous tissue and fluid accumulation along deep fascial planes, with specificities and sensitivities of up to 93% and 88%, respectively.¹⁶
- CT: CT findings in NF include thickening of fascia, subcutaneous emphysema, fluid collections and non-enhancing fascia indicating the presence of necrosis.^{17 18} It has also been shown to detect air missed on plain radiograph, particularly where present in deeper tissues.¹⁹
- MRI: Unlike CT, soft tissue air is less easily identifiable on MRI and can be mistaken for calcification or a metallic foreign body.¹⁸ However, MRI has high soft tissue contrast and can demonstrate the extent of tissue involvement in necrotising infections.^{18 20}

Although routine blood tests and various imaging techniques can help the clinician to differentiate NF from other conditions, the diagnosis is primarily based on clinical findings. If a high suspicion exists then additional tests such as CT or MRI should be avoided so as not to delay treatment which would subsequently worsen the outcome. Simple tests such as blood gas analysis showing an acidosis and high lactate levels are strongly associated with NF and will not delay surgical intervention.⁸

Immediate resuscitation and broad-spectrum intravenous antibiotics should be initiated alongside early involvement of the surgical team who should proceed directly to theatre for urgent debridement. Surgeons should make incisions to the deep fascia (this will often reveal the presence of 'murky dishwater fluid' in the wound), and all non-viable tissue including fascia should be excised. Further surgical exploration 24–48 h later is mandatory to ensure

that the infectious process has not extended. Repeated debridements may be necessary (as dictated by the condition of the wound) until the infection has been controlled adequately—in one study of 30 patients with NF in the central portion of the body (including the thighs), 6 (20%) patients died and the survivors each underwent between 2 and 4 debridements.⁹ Haemorrhage is not uncommon after debridement, and in cases of disseminated intravascular coagulation such bleeds can be difficult to control. Whole blood and clotting products should always be available for the patient prior to surgery.¹⁵ Early recognition of NF followed by urgent referral to surgeons, microbiologists and intensive care and subsequent prompt surgical intervention has helped to decrease the mortality from over 70% to approximately 20%.^{5 6} Aggressive debridement aims to excise all necrotic tissue up to healthy bleeding margins and urgent microbiological analysis of this tissue and of any ‘murky dishwater fluid’ can help to tailor antibiotics. Postoperatively patients should be managed on the intensive care unit where they often require physiological support from the ensuing overwhelming systemic compromise. Further debridement may be carried out in theatres after 24–48 h to ensure excision of all necrotic tissue. Once fully recovered from the infection, closure and reconstruction of the wound can be planned.

The presentation of an elderly patient with groin pain after a fall may have led to a preliminary differential diagnosis of a hip fracture in this case. However, the rapid progression to systemic compromise with significant skin changes, a high lactate and subcutaneous gas on plain radiography were all in fact classical features of NF emphasising that although other conditions must always be considered, NF will almost invariably have a typical presentation. There are several case reports of underlying pathology such as Ludwig’s angina, sigmoid perforation or psoas abscess presenting as NF^{21–23} but there are no case reports in the literature where NF has presented in an atypical manner which is reassuring for clinicians needing to make the critically important rapid diagnosis of the condition. The accident and emergency staff recognised very early that this was a case of NF and it is unlikely that debridement and intravenous antibiotics could have been instigated any earlier. Unfortunately the patients’ age and comorbidities together with the particularly rapid progression of the NF in this case contributed to the unsuccessful outcome.

Learning points

- Necrotising fasciitis (NF) is a rapidly progressive severe infection of the fascia that requires a high index of suspicion in order to make a prompt diagnosis and instigate rapid treatment.
- Certain laboratory tests and imaging can be useful in confirming the diagnosis of NF, subcutaneous air on x-ray being particularly specific for the condition. However, diagnosis is largely based on clinical findings and investigations should not delay surgical intervention when suspicion of NF exists.
- Early surgical referral and involvement of microbiology and intensive care teams are pivotal in ensuring a favourable outcome.

Competing interests None.

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