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Necrotising fasciitis

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Necrotising fasciitis is one of a group of highly lethal infections that cause rapidly spreading necrosis of fascia and subcutaneous tissues, sometimes involving muscles and skin. They were previously known by such names as hospital gangrene, gas gangrene, and Fournier's gangrene and are now referred to by the generic term "necrotising soft tissue infections." We review the clinical features and highlight the potential pitfalls in diagnosis.

Methods

We searched Medline and the Cochrane Library using terms such as "necrotising fasciitis" and "Fournier's gangrene." We also drew on our own experience. The overall quality of evidence is weak to moderate.

How common is necrotising fasciitis?

About 500 cases of necrotising fasciitis a year occur in the United Kingdom. Although rare, the infection occurs often enough for most emergency department doctors and general practitioners to see a case in the course of a working lifetime. The key to successful treatment lies in early diagnosis and appropriate management.

This condition is catastrophic if missed. Even with surgery, mortality is 20-40%. ^{2 3} Delay in diagnosis increases mortality, ⁴⁻⁶ and those who survive need more extensive surgery, reconstruction, and often amputation. With early diagnosis outcome is much improved ^{7 8} and significant long term disability is reduced or prevented.

What are the clinical features?

Necrotising fasciitis is notoriously difficult to diagnose. The initial symptoms are non-specific up to the point when the patient rapidly deteriorates, and septicaemia develops, often accompanied by shock or confusion. However, this clinical course is often slower than might be expected. Fever or pain develops first, so the patient often presents initially to primary

care or the emergency department. The pain may seem to be disproportionate to the clinical findings.

Cellulitic skin changes may develop next. The presentation may mimic haematoma, bursitis, phlebitis, sciatica, cellulitis, septic arthritis, or deep venous thrombosis. The classic textbook picture of haemorrhagic bullae, crepitus, and skin necrosis often does not occur until day 5 or later. The patient may seem systemically well until relatively late. Table 1 shows the frequency of signs and symptoms at presentation. Figure 1 shows necrotising fasciitis of the scrotum.

The patients who present the greatest diagnostic difficulty are those presenting with pain but without fever or systemic signs. Pain is caused by tissue necrosis, but the nerves can also be infarcted as perforating vessels to the tissues are occluded by thrombus during the necrotic process. This can result in exquisite pain and tenderness but also in sensory loss to the overlying skin. The area may be tender or tense. Pain is often very severe, preventing weight bearing or use of the limb but may be mild until late in the process. People who inject drugs often present without systemic signs. ¹⁶

Even in patients with systemic signs, the severity of the skin infection is often not apparent initially. The skin may look normal, or there may be erythema suggestive of cellulitis. In true fasciitis there will be no ascending lymphangitis, but this may be present in other, more superficial necrotising soft tissue infections.

How is necrotising fasciitis diagnosed?

Necrotising fasciitis affects all age groups but is particularly rare in childhood. It is more common in patients with diabetes, chronic hepatitis, and malignancy (particularly leukaemia) and in people who inject drugs. Iatrogenic immunosuppression also increases the risk. Intra-abdominal malignancy or sepsis can lead to necrotising fasciitis of the abdominal wall. Varicella infection is a recognised risk factor in children. ¹⁸ Any puncture wound or surgical procedure can introduce infection, including such minor procedures as acupuncture or intramuscular injection.

Learning points

Necrotising fasciitis is a lethal and rapidly progressive soft tissue infection, which can occur in healthy young patients

People with diabetes, those who inject drugs, and patients with haematological malignancy are particularly at risk

Diagnosis requires a high index of suspicion. Consider necrotising fasciitis especially when the presentation is "not quite right" or the patient is not responding to treatment

Early surgical exploration of the soft tissues has little morbidity and may be the only means to reach a definitive diagnosis and expedite treatment

In established necrotising fasciitis, surgery gives a 60-80% chance of survival. The earlier the first exploration and subsequent debridement, the less extensive the resection and postoperative morbidity is likely to be

However, about 25% of cases occur in patients without comorbidity or precedent trauma.

In patients with fever, suspicion may be aroused by something being "not quite right" for a diagnosis of cellulitis. The classic cyanotic and bullous skin changes may only appear late in the process; however, the site of infection may appear unusual. The pain may seem too severe for cellulitis, despite relatively mild skin signs, or there may be overlying sensory loss. The patient may seem disproportionately unwell for the degree of skin involvement. The progression of the illness also suggests the diagnosis. The patient may seem relatively well initially, but will deteriorate despite treatment with antibiotics. Close observation is important for identifying those patients whose disease is not progressing as expected.

In patients presenting with pain alone, the severity of the pain and absence of trauma may suggest the diagnosis of necrotising fasciitis. In patients with unexplained pain, especially severe or rapidly progressing pain, a search for covert sepsis and an investigation of inflammatory markers is advisable.

What investigations are useful?

No investigations are diagnostic, but blood test abnormalities such as a raised C reactive protein concentration occur relatively early, reflecting the systemic inflammatory response. The most accurate diagnostic scoring system to date is the Laboratory Risk Indicator for Necrotizing Fasciitis (table $2 \Downarrow$). ¹⁹ A score of ≥ 6 was 93% sensitive and 92% specific for necrotising fasciitis in a Singaporean population but achieved only 74% sensitivity and 81% specificity in a UK validation study (H Y Sultan et al, unpublished UK data, 2011). Blood cultures take too long to influence immediate management but have a role in guiding further antibiotic treatment.

Although the validity of this score for a UK population may be in doubt, it demonstrates the relative importance of certain laboratory tests. Hyponatraemia in the presence of sepsis and clinical signs of soft tissue infection should be considered highly suspicious for a necrotising soft tissue infection.

Computed tomography can show fascial swelling, inflammation, and sometimes soft tissue gas and is sensitive (100% in one small series) but less specific²⁰; magnetic resonance imaging is also sensitive²¹ but often not feasible or available.

Ultrasonography can be diagnostic but requires a highly skilled operator. ²² Although plain radiography is not the imaging of choice when the diagnosis of necrotising fasciitis is suspected because sensitivity and specificity are low, it may show subcutaneous gas (fig $2 \downarrow$).

The mainstay for investigation and treatment remains surgical exploration. The decision to explore the soft tissues should be made early. An incision over the site of maximal skin change is needed to assess the underlying tissues. Healthy subcutaneous fat and fascia indicates that further resection is not needed, and the morbidity to the patient is limited to a short scar.

However, if exploration shows necrotic fascia, fat, or the "dishwater" appearance of liquefied necrotic tissue, resection can be done until healthy tissues are reached. This can be facilitated by the "finger sweep test" (necrotic fascia loses its adherence to surrounding tissues and the plane opens abnormally easily until the limit of the disease is reached). Where doubt over the appearances persists, send specimens for histology to look for evidence of necrosis and microbiology for urgent Gram staining.

Necrotising soft tissue infections used to be considered streptococcal, but they are now understood to be often caused by mixed pathogens, including gas forming bacteria such as *Clostridium* species.

How is necrotising fasciitis managed?

Early referral to a surgeon and an early decision to explore and debride is the cornerstone of treatment. In established sepsis, debridement does not bring about a rapid change in the condition of patients. However, over the following hours they tend to stabilise but then often spend several days needing invasive support in intensive care and have an overall hospital stay averaging 33 days.²³ Subsequently, the patient will be referred to the nearest plastic surgery or burn centre for ongoing wound care and reconstruction.

Adjuvant measures include systemic support in an intensive care setting and antimicrobial treatment. Broad spectrum antibiotics such as a benzylpenicillin and flucloxacillin are used. Clindamycin has an additional role owing to its bacteriostatic mechanism. It inhibits the production of the streptococcal superantigen, which greatly contributes to septic shock.²⁴

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Tables

Table 1 | Percentages of patients who show signs and symptoms of necrotising fasciitis at presentation

Study and country	Severe pain	Fever	Tachycardia (with or without hypotension)	Skin erythema	Skin oedema	Skin tenderness	Blistering or bullae	Ecchymosis or skin discoloration	Crepitus
Wong et al ¹² (n=89), Singapore	98	53	74 (18)	100	92	98	45	No data	14
Childers et al ¹¹ (n=162), United States	100	70	No data	95	82	No data	16	49	25
Frazee et al ¹⁶ (n=122), United States	No data	44	59 (21)	80	66	54	12	No data	7
Angoules et al ¹⁷ (n=451), United Kingdom	63	15	(12)	73	49	No data	15	No data	7

Table 2| Diagnostic scoring system of the Laboratory Risk Indicator for Necrotizing Fasciitis

Variable	Score
C reactive protein (mg/L):	
>150	4
White blood cell count (per mm³):	
<15	0
15-25	1
>25	2
Haemoglobin (g/dL)*:	
>13.5	0
11-13.5	1
<11	2
Sodium (mmol/L):	
<135	2
Creatinine (µmol/L):	
>141	2
Glucose (mmol/L):	
>10	1

^{*}The corresponding SI values for haemoglobin are >135 g/L, 110-135 g/L, and <110 g/L.

Figures

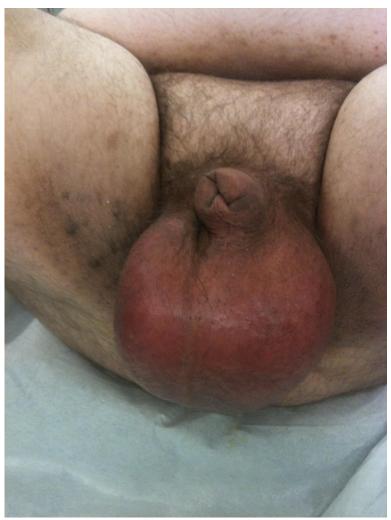


Fig 1 Necrotising fasciitis of the scrotum, with erythema of the scrotum. Subtle necrosis can also be seen in the thigh area. Further images are also available at http://dermnetnz.org/doctors/bacterial-infections/necrotising-fasciitis.html



Fig 2 Extensive subcutaneous gas in the thigh of a man with necrotising fasciitis of the buttock (arrows)