

Original article

Necrotizing Fasciitis: Myalgia is diagnostic? A case series and literature reviews

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Abstract:

Necrotizing Fasciitis (NF), commonly known as 'flesh eating disease,' is an aggressive soft tissue infection that destroys the fascia, subcutaneous tissue and skin. Specific clinical features (crepitus or radiologic features of gas in tissues) either appear late or are of poor sensitivity. Thus, a high index of clinical suspicion is required for early diagnosis and prompt treatment which are the best methods of minimizing its high associated morbidity and mortality. We present 3 cases to demonstrate diagnostic difficulties and challenges in management and highlight the feature of pain on muscular activity.

Key Words: Necrotizing fasciitis, gangrenous fascia, soft tissue infection, Debridement

Introduction:

Necrotizing Fasciitis (NF), commonly known as 'flesh eating disease, is an aggressive soft tissue infection that destroys the fascia, subcutaneous tissue and skin.

Though NF is uncommon, it is a dangerous condition because of difficulties in diagnosis until late in the disease process when mortality can be high. Misdiagnosis ranges from 85-64%.^{1,2} The mortality is about 34%(range 6-76%).³ We present 3 cases to demonstrate diagnostic difficulties

and challenges in management and highlight the importance of looking for pain on muscular activity.

Case 1

A 67 year old male diabetic presented with fever, weakness and pain around the right knee. It started 2 days before and got progressively worse. He was unaware of any trauma or insect bite but 1 day later, noted a foul-smelling ulcerated area just below the knee. On examination, he was toxic, febrile T 38.4 C, pulse 112/min and in pain. The gangrenous ulcer was 10 x 6cm, foul-smelling, associated with five smaller ulcers and had surgical emphysema around the ulcer and up to the knee (Fig 1). Pain in the antero-lateral thigh on knee extension suggested above-knee fascial involvement.



Fig 1

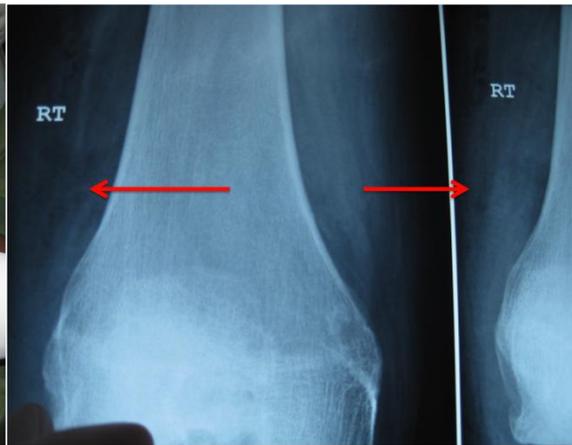


Fig 2

Although no crepitus was palpable in the thigh, x-ray showed air above knee at the level of the deep fascia laterally (Fig 2). At surgical debridement, although the skin above the knee appeared normal, there was extensive fascial and subfascial gangrene extending 30 centimeters above the knee (Fig 3). He was maintained on IV meropenem and metronidazole and improved slowly. Staph. aureus and E.Coli were found on culture. After 5 days of daily dressings he was taken back to the operating room for further minor debridement. He recovered well and was able to go home after 18 days.



Fig 3



Fig 4

Case 2

A 37 year old non-diabetic male presented with fever and worsening pain in the right forearm of two days duration. He had no history of trauma. He was febrile on admission, had a pulse of 100/min and a Wbc of 18×10^3 . On examination, the forearm was warm, erythematous and mildly tender (Fig 4). X-ray showed no gas in the tissue. He was diagnosed as cellulitis, admitted to hospital and started on amoxicillin/ clavulanate and Flagyl. Within 24 hours, his pain worsened and he could not perform a firm hand grip as it produced marked pain, signalling fascia/ muscular involvement. Gentamicin was added and surgery performed. Incision over the erythematous area revealed pus and gangrenous fascia with normal underlying muscle (Fig 5).

The gangrenous fascia was excised, the pus drained and the wound left open. Culture produced no bacterial growth. He was discharged after 5 days and secondary closure done on day 12 postop.

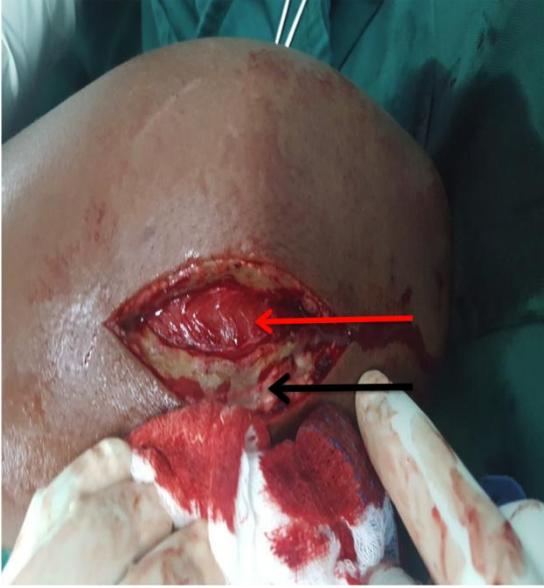


Fig 5



Fig 6

Case 3

A 57 year old male presented with a gangrenous, foul smelling area in the right groin. Six years before, he had biopsy confirmed CA prostate. He had brachytherapy with no complications except mild erectile dysfunction. One year before this presentation, he started experiencing pelvic pain and had two episodes of mild PR bleeding. In the last month, he noticed passage of faeces and wind in the urine.

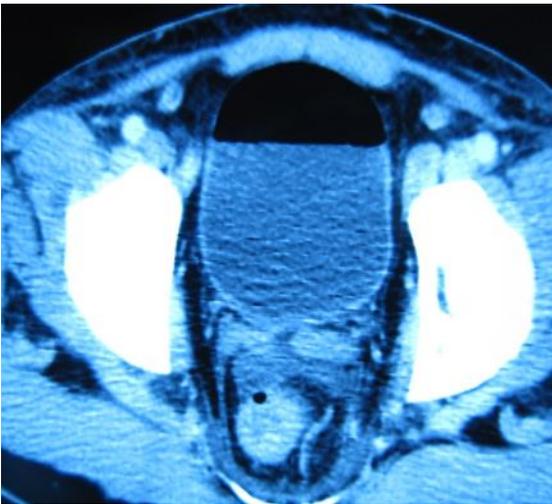


Fig 7



Fig 8

He was febrile T 38.8°C, tachycardic 120/min, and tachypnoeic 24/min. He had gangrenous skin in the right groin 8x6cm with extensive surrounding erythema, swelling and oedema (Fig 6). He held the hip in mild flexion and had pain in flexion against resistance and on passive extension. This suggested that the gangrenous skin and subcutaneous tissues involved the fascia or muscle,

indicating possible NF. On examination, he had a hard mass in the anterior rectal wall, fixed to the mucosa and a defect was palpable at the tip of the finger. His Hb was 9.2g/dl and Wbc 19.6×10^3 dl. CT confirmed recto-vesical fistula (Fig 7). He was started on IV amoxicillin/clavulanate, gentamicin and metronidazole. Urgent debridement revealed extensive gangrene of the fascia and subcutaneous tissue in the proximal third of the thigh, medially and anteriorly (Fig 8). Much pus was drained and foul-smelling gangrenous tissue excised. Post operatively, he remained toxic, developed acute renal and respiratory failure and died 30 hours later. His culture grew *Streptococcus pyogenes* and *Klebsiella* species.

Discussion:

Early in the disease signs may be minimal, and diagnosis difficult, as in our second case and the above-knee extension of disease in Case 1. There may be no skin change, erythema, swelling, induration, blebs/bullae or frank gangrene.⁴ In 89 cases, Wong et al made the diagnosis in only 13 (14%) at the time of admission¹. They emphasize the paucity of skin changes in the early stages of NF. Also, Goh recorded the correct admitting diagnosis in only about 25% of 1463 cases⁵. The misdiagnosis rate ranged from 41-96% in 6 studies reviewed by Goh et al.⁵ Thus, initially misdiagnosis is often more common than an accurate diagnosis. The more obvious clinical signs are late – large bullae, gangrenous skin, ulceration, swelling, fluctuation, crepitus with widespread oedema and inflammation.¹ The paucity of skin changes can be explained by the pathogenesis of NF. Sepsis in the fascia or deep subcutaneous tissue, especially if spreading horizontally by angiothrombotic microbial invasion, will produce widespread diffuse deep tissue gangrene with only later skin changes.^{4,6,7} In our first case, there were no clinical signs in the thigh, yet there was extensive fascial gangrene for 30cms above the knee (Fig.3). Our second case was initially treated as simple cellulitis, the NF diagnosis only being made when muscular movement became painful. Even in our third case where skin changes were obvious, holding the hip in flexion, with pain on passive extension, suggested muscular/fascial involvement. In an

attempt to diagnose and treat early, prompt surgery may be performed. However, this can result in over treatment and unnecessary surgery as occurred in about 20% of cases reported by Heitbrink et al.⁸ Radiology is often non-specific, especially in early stages of NF.^{9,10} Swain et al indicated that ‘radiology studies...were of no benefit’.¹¹ Gas in the soft tissues is found in relatively few patients 16.9 – 37%.^{4,12,13} Thus, on average, plain X-ray shows subcutaneous gas in less than 25% cases.¹⁴ CT and MRI may be of value but are often non-specific in early NF.¹⁵ In more advanced cases they may reveal fascial thickening, gas in tissue planes and fat stranding.¹⁶ Schmid et al describe MRI as quite reliable with a sensitivity of 100% and specificity of 86%.¹⁷ However, other authors have found radiology of limited use. Krieg et al found CT to be accurate in only 50% of cases.¹⁸

The predisposing factors enumerated by Vijaya Kumar et al, age greater than 50 years, atherosclerosis, burns, cancer, immune-compromised state, chronic alcoholism, corticosteroid use, diabetes mellitus, hypo-albuminaemia, intravenous drug abuse, malnutrition, obesity, occult diverticulitis, peripheral vascular disease, postoperative infection, trauma are often absent⁴. However, while our first case was diabetic and the third a cancer patient, the second case was young, had no predisposing factors and no classical signs; these may have contributed to his initial misdiagnosis. The combination of erythema, pain and fever, even in the absence of the obvious late signs, should arouse suspicion of NF.¹⁶ Goh et al further emphasized that ‘swelling and pain from cutaneous infection that is out of all proportion to the physical findings ‘ should lead to a consideration of NF; this was true of our Case 2.⁵ Although initially misdiagnosed, worsening signs and symptoms while under adequate antibiotics and analgesic coverage should also trigger suspicion of NF. When these features are present, even in the absence of radiologic findings, surgical exploration should be considered especially if there is pain on muscular activity (as we did in Case 2). The likely causes of this pain are (i) in some cases, muscular slips actually originate from the deep fascia and (ii) where not attached, the sliding muscular movement on the necrotic fascia can produce pain.

NF remains, largely a clinical diagnosis, requiring a high index of suspicion as early diagnosis and prompt treatment are the best methods of minimizing morbidity and mortality; although the mortality is high (34%) the morbidity is significant with amputation rates of about 12% and hospitalization of 32 days.^{3,19} The greatest threat to early diagnosis is the paucity of clinical signs

and classical radiographic findings in early stages of the disease. It is also important to recognize, as in our first case, that even when the diagnosis is made the disease is often more extensive than appears clinically. Thus, debridement should extend up to normal tissue on all sides.

Exacerbation of pain by putting the relevant muscle into action could help distinguish cellulitis from the musculo fascial involvement of NF. This may be a very useful sign and, to our knowledge, has not been previously highlighted. All 3 of our cases had clinical evidence of muscular involvement; we regard this as a useful clue to the diagnosis of NF and will distinguish it from more superficial infections.

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Author contribution: All authors have contributed significantly in designing and organizing to write manuscript, collecting data as well help in critical analysing the manuscript. All authors have approved the final version of this manuscript

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Figure Legends:

Fig 1: A gangrenous ulcer was 10 x 6cm (red arrow) associated with smaller ulcers (black arrows).

Fig 2: An x-ray showing air (red arrows) above knee at the level of the deep fascia laterally.

Fig 3: Showing extensive fascial and sub-fascial gangrene (black arrow) with relatively normal overlying skin (red arrow)

Fig 4: Showing a very swollen erythematous right forearm.

Fig 5: An incision over the erythematous area revealed pus and gangrenous fascia (black arrow) with normal underlying muscle (red arrow)

Fig 6: Gangrenous skin in the right groin 8x6cm with extensive surrounding erythema, swelling and oedema

Fig 7: An axial CTscan showing a recto-vesical fistula.

Fig 8: Debridement revealed extensive gangrene of the fascia and subcutaneous tissue in the proximal third of the thigh.