Early Clues in Diagnosing Fournier’s Gangrene

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ABSTRACT
Necrotizing Fasciitis has a high mortality if not managed early, yet early diagnosis is inherently difficult. The external tissue is initially unaffected, therefore we rely on secondary signs of toxicity, which often represent an already established systemic infection. It typically affects immunocompromised patients, who are slow to manifest these secondary signs, complicating the diagnosis further. We present a case of an immunocompromised patient, who did not manifest typical signs of systemic infection. Atrial Fibrillation was the first clue to an underlying infection, and initially the only localising infective source was a hydrocele. Cutaneous features in keeping with a Fournier’s Gangrene occurred forty-eight hours later. This case highlights the difficulty in diagnosing Fournier’s Gangrene, and the possibility that a hydrocele in a septic patient may be reactive to a more sinister underlying necrotizing infection.

Keywords: Fournier’s Gangrene, Necrotizing Fasciitis, Hydrocele

Introduction
Fournier’s Gangrene is a subset of Necrotizing Fasciitis effecting the perineum or external genitalia.

Signs and symptoms are often unreliable. Cutaneous necrosis occurs late in the disease process, and pain may be transient. If not actively examined for, it can be easily missed, and delays in diagnosis can have dire consequences.

Case
A 59-year-old man with a history of Type 2 Diabetes, paroxysmal Atrial Fibrillation and Multiple Myeloma was seen in the Haematology outpatients clinic for stem cell collection. During the procedure he went into rapid Atrial Fibrillation with intermittent hypotension. He was neutropenic and thrombocytopenic, had a CRP of 364 units mg/L, a lactate of 3.4 mmol/L, and blood sugar levels of 20 mmol/L. He had E. Coli positive blood cultures, and the only likely infective source on examination was a tender scrotal swelling. Testicular ultrasound found a small right sided hydrocele (Figure 1), with mild scrotal subcutaneous swelling, but no subcutaneous emphysema (Figure 2). Forty-eight hours later, a patch of skin necrosis at the base of the scrotum was identified, that was not seen on previous examinations. He was taken to theatre for an urgent debridement, where a 10 x 15 cm necrotic area was found, in keeping with Fournier’s Gangrene. Over the course of two weeks, he underwent further debridement and a superficial skin graft. Two weeks post discharge he was reviewed in the Haematology outpatient clinic for ongoing management of Multiple Myeloma.

Discussion
Necrotizing Fasciitis is an infection of the muscular fascia, that results in destruction of this fascial layer as well as the overlying subcutaneous tissues and dermis. The muscular layer beneath the fascia is usually spared due to its rich blood supply [1]. It is a rare condition, with estimates ranging from 0.3 to 15 cases per 100,000 [1, 2]. It is divided into polymicrobial (type 1) and...
monomicrobial (type 2) infections, with the former caused by aerobic and anaerobic bacteria, and the latter most commonly caused by Group A Streptococcus.

Fournier’s Gangrene is Necrotizing Fasciitis of the perineum and/or external genitalia. It is most commonly seen in immunosuppressed patients. Risk factors include diabetes, alcoholism and liver disease [3-6]. It is often idiopathic, but may be secondary to anorectal, urogenital or cutaneous diseases, trauma and pelvic procedures [5]. Estimations of mortality rate range from 22 – 40 per cent [7-9].

The expression of bacterial toxins allows the infection to spread rapidly along fascial planes, quickly resulting in systemic toxicity. The appearance of the overlying tissue often does not reflect the extent of infection, making early diagnosis difficult [10]. Signs and symptoms include erythema, fever, crepitus and skin necrosis. The symptom of pain is dubious, as while pain occurs initially, anaesthesia often follows due to thrombosis of small blood vessels and superficial nerves [11]. This may precede skin necrosis, making it an important early feature. Gas producing organisms cause subcutaneous emphysema, an important examination finding and radiological feature [12]. CT is the ideal imaging modality. Ultrasound can be used to identify soft tissue gas, however this was not present in the current case [13]. Diagnosis is confirmed by surgical exploration and examination of the fascial planes, and debridement of necrotic tissue is the definitive treatment [14]. Broad spectrum antibiotic therapy should be instigated to cover gram positive, gram negative and anaerobic organisms [10]. A suggested regimen includes a Carbapenem or Piperacillin-Tazobactam, Vancomycin or Daptomycin to cover MRSA, and Clindamycin for antitoxin effects [14]. The antimicrobial regimen in the current case followed this guideline, and was further tailored once sensitivities were obtained.

This case illustrates many of the difficulties in diagnosing Fournier’s Gangrene and Necrotizing Fasciitis. Physical signs are unreliable – external necrosis presents late, and pain is often transient due to small vessel thrombosis. Immunocompromised patients who are the key demographic for this disease, may mount a late or atypical immune response. At no point was our patient febrile, and the first clue to an infection was rapid Atrial Fibrillation. Once the bacteraemia was identified, accurate source identification was complicated by a hydrocele, which could be considered either a distractor or a clue to the underlying infection. Hydroceles are prone to becoming infected, however they rarely result in sepsis. It is unlikely that the hydrocele led to Fournier’s Gangrene, as a review of the literature has found only one case of this previously, in which Fournier’s Gangrene developed three weeks after the hydrocele [6]. This timeline contrasts with the current case. A hydrocele may be reactive; secondary to a local infection, and this was the most likely scenario in this case [15]. Once the hydrocele was identified, and prior to finding scrotal necrosis, the hydrocele was considered as the likely infective source, especially given the patients immunocompromised status and increased susceptibility to hematogenous bacterial seeding. This had the potential to delay the diagnosis of Fournier’s Gangrene, had the patient not been examined multiple times. This highlights the importance of considering that a hydrocele may be secondary to an underlying soft tissue infection when looking for a source of sepsis, as they are unlikely to be a primary septic source.

A timely diagnosis of Fournier’s Gangrene and more broadly, Necrotizing Fasciitis, relies upon a high level of clinical suspicion, especially as signs and symptoms can be non-specific. As external cutaneous changes often occur late, repeat examination is crucial. Hydroceles are uncommon sources of sepsis, and this case highlights the importance of considering them as potentially reactive to a more sinister disease, when looking for an infective source in a septic patient.

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References