Lemierre's syndrome masquerading as necrotizing fasciitis

Introduction

This article presents the case of a young man who presented with shoulder pain and neck swelling on a background of recent tonsillitis. A series of investigations was performed, including blood biochemistry and full blood count, blood cultures and computed tomography. The patient was then taken to theatre for incision and drainage of multiple abscesses, the contents of which were sent for microbiology. The patient required prolonged admission to the intensive care unit and remained in hospital for 30 days. Follow up 6 months later confirmed a full recovery with no adverse sequelae.

Discussion

The simple 'sore throat' is an ailment that affects the vast majority of the population, with 90% a direct result of a self-limiting viral illness. However, it is also the primary presenting complaint of potentially life-threatening bacterial infections such as group A beta-haemolytic streptococcus or *Fusobacterium necrophorum* (Centor and Samlowski, 2011). It is important that these infections are detected in a timely manner to reduce associated morbidity and mortality.

F. necrophorum is responsible for a variety of presentations, including the simple sore throat, persistent sore throat syndrome and Lemierre's syndrome (Batty et al, 2005). Lemierre's syndrome is associated with significant morbidity and mortality and typically occurs in previously fit and well young patients following an acute oropharyngeal infection. Most clinicians will never encounter such a case but recent increases in prevalence suggest that perhaps thresholds of clinical suspicion should be lowered. Incidence (estimated conservatively at 0.9 per million) (Jones et al, 2001) is difficult to estimate, owing to challenges in establishing a firm diagnosis, but is thought to be increasing as a result of antibiotic rationalization, variable antibiotic resistance profiles and, interestingly, reduction in rates of tonsillectomies (Riordan, 2007). Equally, it is suggested that rising incidence may simply reflect increased identification of the organism, rather than a true rise in incidence of the disease.

Establishing incidence is further compounded by difficulties associated with accurately defining Lemierre's syndrome. Definitions range from 'necrobacillosis' (septicaemic illness with isolation of *F. necrophorum* in blood cultures), 'post-anginal sepsis' (thrombophlebitic complications following tonsillitis) to the more established *F. necrophorum* sepsis of

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Case Report

A 35-year-old male wrestler presented to the emergency department with a 2-week history of painful right shoulder associated with tender anterior neck swelling. He had been seen 11 days previously with right shoulder pain and discharged with a diagnosis of adhesive capsulitis. Of note on initial review was a preceding tonsillitis, treated with clarithromycin by his GP.

On arrival in the department he reported fatigue and systemic upset during the preceding week. He was tachycardic, normotensive and afebrile. His right shoulder was exquisitely tender to touch. Examination revealed right anterolateral deltoid erythema associated with fluctuance, and a large midline cervical lymphadenopathy. The only other finding of note was reduced air entry at the base of the left lung field. Investigations revealed normochromic normocytic anaemia (haemoglobin 7.6 g/dl), leucocytosis (white cell count 25.7x10^9/litre, neutrophils 23.46x10^9/litre) and markedly raised C-reactive protein (475 mg/litre). X-ray of the right shoulder revealed multiple locules of gas in the soft tissue above the right clavicle and chest X-ray confirmed a left-sided pleural effusion.

Following multidisciplinary review (orthopaedic surgery, plastic surgery, infectious diseases, ear nose and throat surgeons) a working diagnosis of necrotizing fasciitis was made. Computed tomography was performed to delineate the extent of infection before operative debridement. This revealed a large (2.2x2.7x4.0 cm) collection in the deep cervical fascia posterior to the left submandibular gland that was communicating with a midline abscess (4.7x2.8x7.8 cm) at the level of the thyroid cartilage (*Figure 1*). This collection extended into the right sternomastoid. Additional findings were a collection in the right trapezius muscle (6.8x4.2 cm), a loculated collection in the right posterior cervical spinal muscles extending from C3 to C7, osteomyelitis of the distal right clavicle, a large left pleural empyema with collapse of the left lower lobe and multiple cavitating nodules in the right lung (*Figure 2*).

The patient proceeded to urgent operative debridement, where computed tomography findings were confirmed. There was no evidence of surrounding necrotic tissue, thus calling into question the working diagnosis. More than 50 ml of purulent exudate was drained from the neck abscesses and fenestra created through the right clavicle to release sequestrum. A large bore chest drain was inserted, intravenous clindamycin, ciprofloxacin and vancomycin were commenced and the patient was admitted to intensive treatment unit. Initial microbiology revealed Gram-negative rods and vancomycin was changed to metronidazole accordingly. The patient had an eventful postoperative recovery. He continued to have recurrent empyemas which were managed with chest drains and prolonged antibiotic regimens, eventually being transferred to an infectious diseases ward after 2 weeks in intensive treatment unit. Subsequent microbiology isolation revealed *Fusobacterium necrophorum* as the responsible organism and a diagnosis of Lemierre's syndrome was confirmed. The patient was discharged after a hospital stay of 30 days. Follow up 6 months later confirmed a full recovery with no adverse sequelae.

oropharyngeal origin (with evidence of non-local metastatic lesions).

Authors prominent in the field generally agree that diagnosis of Lemierre's syndrome requires either a history of anginal illness with remote metastatic lesions (typically pulmonary) in conjunction with internal jugular vein thrombophlebitis or isolation of *Fusobacterium* spp. (from blood cultures or a normally sterile site). The requirement for internal jugular vein thrombophlebitis as a necessary feature to establish diagnosis remains contentious as large numbers of otherwise 'typical cases' would be excluded.

Initial diagnosis is essentially clinical, with a history of oropharyngitis preceding onset of fever by approximately 5 days. Tender cervical lymphadenopathy in the presence of leucocytosis and markedly raised inflammatory markers are typical but not specific. Diagnosis is established by supportive findings of central vein thrombophlebitis, multi-organ metastatic lesions (septic foci) or simply isolation of *F. necrophorum* from blood cultures. Necrotizing soft tissue infections must be considered and excluded. Early symptoms of necrotizing fasciitis include swelling, erythema, pain and tachycardia, all of which were present in this patient with Lemierre's. However, rapid progression to discolouration, bullae, necrosis and crepitus would raise the suspicion of necrotizing soft tissue infection. If the diagnosis is in doubt, operative debridement and identification of underlying necrotic soft tissue allows clarification and management can be altered accordingly (Anaya and Dellinger, 2007).

Figure 1. Abscess in the deep cervical fascia posterior to the left submandibular gland (2.2x2.7x4.0 cm) communicating with a midline abscess (4.7x2.8x7.8 cm) at the level of the thyroid cartilage and extending into the right sternomastoid.

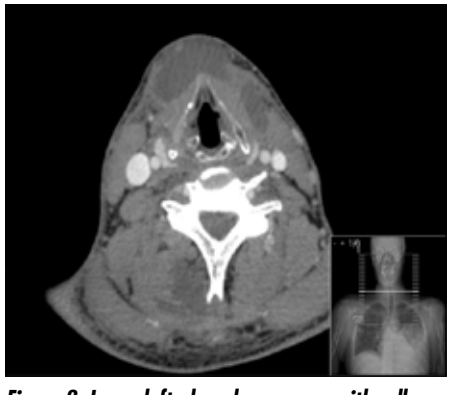
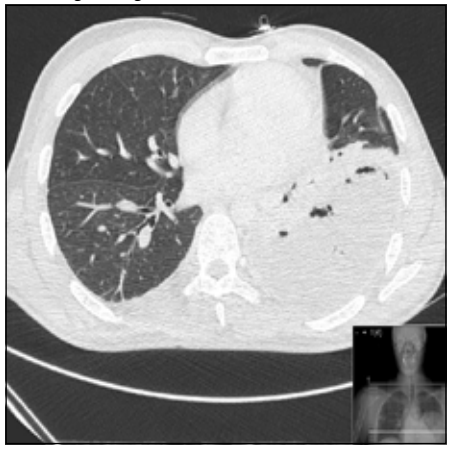


Figure 2. Large left pleural empyema with collapse of left lower lobe and multiple cavitating nodules in the right lung.



Antibiotics are the mainstay of management in Lemierre's syndrome. The prolonged incubation period of *F. necrophorum* often delays diagnosis (in this case it was 12 days before culture isolation). Documented in-vitro sensitivities include penicillin, tetracycline and clindamycin, but in-vivo analysis has shown metronidazole to be the ideal therapeutic choice. Prolonged combination therapy (metronidazole and penicillin) is advised because of the risk of mixed infection. Anticoagulation has been implemented successfully in certain instances of confirmed great vessel thrombophlebitis, but remains an unproven treatment (most reports suggest improvement without anticoagulation).

Surgical management (debridement and/or incision and drainage of affected sites) is occasionally required to expedite clinical improvement, particularly in the context of extensive disease or anticipated significant clinical deterioration. Other surgical options include great vessel surgical ligation (in cases of progressive thrombophlebitis as a source of septic emboli) (Escher et al, 2008) and site drainage (multi-site abscess or empyema) (Li et al, 2009). Increasingly, there is support for lowering thresholds for surgical intervention, particularly where patients fail to improve with medical management (Riordan, 2007).

Owing to both the 'rarity' and aggressive nature of Lemierre's syndrome, diagnostic thresholds ought to be lowered to avoid significant morbidity associated with delayed or missed diagnoses. Importantly, in cases of persistent tonsillitis (despite standard first-line treatment), this diagnosis should mandate consideration of earlier engagement of support specialities (specifically microbiology or infectious diseases), thereby prompting review of the antimicrobial regimen in such cases. Prompt diagnosis and management is associated with good prognosis but the mortality rate remains around 5% (Chirinos et al, 2002). Acutely unwell patients with severe or refractory soft tissue infections must be assessed and managed promptly to obtain the best outcome. However, while clinicians must have a low index of suspicion for conditions such as necrotizing fasciitis, thorough history and examination are important to identify other significant differentials. **BJHM**

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LEARNING POINTS

- This case highlights a rare but potentially life-threatening disease that can be successfully treated with appropriate management.
- This complication can occur despite standard first-line treatment.
- The condition presented in an acutely unwell young patient without any significant comorbidities.
- The report discusses the difficulties in diagnosing Lemierre's disease, as well as the diversity in presentation, both of which are important considerations for clinicians.