CASE REPORT

Lemierre's syndrome; a rare cause of septic arthritis

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SUMMARY

Lemierre's syndrome is a rare condition characterised by pharyngitis leading to septic thrombophlebitis of the internal jugular vein. Complications include pulmonary septic emboli, septic arthritis and disseminated intravascular coagulation. The authors present a case of a healthy woman aged 25 years with septic arthritis of the shoulder due to this unusual cause. This diagnosis was made via a combination of clinical, radiological and microbiological findings. It was successfully treated via surgical and antimicrobial interventions. The patient made a good recovery with minimal associated morbidity or loss of function. This case highlights the importance for awareness and high index of suspicion for rarer causes of septic arthritis in young healthy adults as early appropriate intervention maximises prognosis.

BACKGROUND

Lemierre's syndrome sometimes known as postanginal septicaemia was first described by Andre Lemierre in 1936 based on a case series of 20 patients.¹ It is rare, however, if left inadequately managed or untreated, can be debilitating or more seriously potentially fatal.

It is characterised by internal jugular vein thrombosis with associated sepsis. Lemierre's syndrome develops subsequent to an episode of acute pharyngitis with a typical interval of 4–5 days. It typically affects children, adolescents and young adults.

The bacterium *Fusobacterium necrophorum* is often considered pathognomonic for Lemierre's syndrome.² It is a Gram-negative anaerobic rod found normally in the flora of the pharynx and gastrointestinal tract. The proposed aetiology of this condition is direct spread of infection from pharyngitis in the peritonsillar space to the nearby internal jugular or facial veins. This then allows the dissemination of metastatic septic emboli and fusobacterial septicaemia.²

The predominant symptom in Lemierre's syndrome is a significant sore throat and fever, which typically precedes further symptoms and sequelae by 4–5 days (but up to 12).³ The most common source of dissemination is the lungs and therefore chest pain and shortness of breath are typical complaints. Septic emboli also have a predisposition to affect large joints such as the hip, knee or shoulder.

The authors report a case with an unusual presentation of this rare condition which if not diagnosed and acted on could have led to significant disability or even proven fatal. Increased awareness and high clinical index of suspicion

should allow vigilant clinicians to diagnose and intervene early on.

CASE PRESENTATION

A previously generally fit and well right hand dominant woman aged 25 years presented to the emergency department with a 10-day history of progressive left shoulder pain and swelling. This was associated with general malaise and lethargy preceded by a severe sore throat around 2 weeks prior to presentation. Her past medical history included mild asthma and hay fever only for which she occasionally used a salbutamol inhaler as and when needed. One month prior to admission, she received a contraceptive implant in her left arm. She did not have any allergies. There was no relevant family history, she was a smoker of approximately five cigarettes a day and worked as a support worker in a children's community team.

On admission, she was afebrile, tachycardic at 110 bpm but normotensive. On examination, she had a swollen, erythematous left arm and shoulder which was tender and hot to touch. Tenderness was present in the supraclavicular region and lateral aspect of the neck. She was unable to move the shoulder due to pain. Passive range of motion of the elbow was full and pain free. There was no evidence of compartment syndrome and she was distally neurovascularly intact.

INVESTIGATIONS**Blood tests**

Leucocytosis (white cell count $22.77 \times 10^9/L$), neutrophilia ($20.93 \times 10^9/L$) and a raised C reactive protein (CRP) of 270 mg/L.

Imaging

A plain film of the left shoulder on admission did not show any periosteal or bony changes or fractures. A contraceptive implant was visible (see figure 1). An initial urgent ultrasound scan demonstrated no evidence of collection or free gas.

An MRI of the left shoulder was undertaken which demonstrated diffuse soft tissue signal change centred on the left acromioclavicular joint, posterior chest wall and neck with a large focal collection within the left trapezius (figure 2).

Microbiology

Tissue cultures showed heavy growth of multisensitive *F. necrophorum*. Blood and urine cultures were all negative. HIV test was undertaken due to the atypical nature of infection which was negative.

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Figure 1 Plain film of left shoulder on admission.

DIFFERENTIAL DIAGNOSIS

The initial differential on presentation included severe cellulitis or septic arthritis of the affected shoulder. Clinically this was felt not to represent necrotising fasciitis. The difficulty in differentiating clinically between cellulitis and septic arthritis prompted the early imaging and further investigations described above.

TREATMENT

The patient was treated for sepsis from unknown origin and transferred to the intensive care unit for resuscitation and fluid balance monitoring. The patient deteriorated over the following 12 hours despite appropriate antibiotic therapy advised by microbiology. She was taken to theatre for an emergency exploration of the left shoulder. Findings included thick pus in deltopectoral groove extending down to the antecubital fossa and more proximally around the glenohumeral joint with a small breach of the capsule. Full debridement of all necrotic and infected tissue followed by washout of the glenohumeral joint and soft tissues was performed (see figure 3). The wound was left open and gently packed.

Figure 2 Sagittal and coronal Short T1 Inversion Recovery (STIR) MRI showing soft tissue changes and focal collection.

Figure 3 Deltopectoral approach to shoulder post washout.

A relook exploration was undertaken the following day which revealed frank pus exuding from under the conjoint tendon and coursing under the clavicle towards the cervical region. The surgical incision was extended proximally to decompress the more proximal collection. Thorough irrigation with copious amounts of saline was used and the wound left open with packs in situ.

Three days later, a further relook demonstrated a self-draining abscess over the posterolateral aspect of the shoulder, with no intracapsular involvement. The glenohumeral joint showed no further evidence of infection and was closed after further irrigation. The remaining wounds were irrigated and a negative pressure dressing applied.

Five days later at further exploration the tissues appeared healthy with no signs of superficial or deep infection. The wound was closed in layers.

She was treated with intravenous antibiotics (clindamycin and metronidazole) on microbiology advice initially and continued until her wound was dry and healed and her CRP had fallen to <50, at which point she was converted to oral metronidazole (at 2 weeks) followed by discharge.

OUTCOME AND FOLLOW-UP

On admission, the patient was unable to move or use their left arm due to pain and swelling. This gradually improved during the admission and with each operative intervention. A further plain film of her left shoulder was taken prior to discharge which did not show any abnormalities (figure 4). She was with a plan for follow-up at 1 week in fracture clinic.

The patient presented to clinic 8 days later and was well in herself. The wound was healthy and blood tests were reassuring with a CRP of 4. X-ray of the left shoulder at this point was unremarkable (see figure 4). The main problem at this point was stiffness and weakness in the left shoulder and so she was referred for intensive physiotherapy.

Her next review was 3 weeks later following physiotherapy and hydrotherapy demonstrated an increased range of movement and improved power in her rotator cuff musculature. She did not complain of any further pharyngodynia or other symptoms related to Lemierre's syndrome.

The most recent clinic review at the time of writing was 10 weeks posthospital discharge. Her shoulder ROM, strength and overall function continued to improve. She had no impairment to performing all activities of daily living. X-rays of the shoulder showed no evidence of chondrolysis or secondary degenerative change.

Figure 4 Plain film of left shoulder at follow-up.

DISCUSSION

Lemierre's syndrome is a rare condition with fewer than 160 cases reported to date.⁴ Postulated incidence from a study by Hagelskjær *et al*⁵ in Denmark has been reported as 3.6 cases per million per year.

The literature shows that this is primarily a disease affecting younger patients who are generally otherwise fit and well. A review of the literature has previously shown that the majority of cases have presented in the second decade of life for those affected. A review by Karkos *et al*⁶ found that the majority of cases (as with our case) present in the second decade of life (51%) and then third (20%) and first decade (8%). There does not appear to be a predominance of gender.

To date, there are no agreed evidence-based criteria for diagnosis; however, hallmarks of this condition include initial oropharynx infection, positive blood culture for *F. necrophorum*, internal jugular thrombosis and one or more metastatic foci of infection.⁷ Classically, the literature states that *F. necrophorum* is a *sine qua non* for Lemierre's syndrome⁷; however, a microbiological review by Riordan has suggested otherwise.⁸ A case review highlighted that in 86% of cases *F. necrophorum* was isolated, 8% of cases cultured other organisms and in 6% of cases no organisms were cultured.⁸ It has been commented on that some of these other organisms (such as *Proteus* sp) would likely not be capable of causing this condition. Also, there are many factors such as timing, equipment and confounding factors that would preclude the identification of *F. necrophorum* for some cases. Therefore, the evidence is inconclusive as to whether *F. necrophorum* is truly the sole pathogen for Lemierre's syndrome. The culture of *F. necrophorum* in our case however was key to making the diagnosis and initiating a suitable management plan in response.

There is no current gold standard in terms of diagnostic imaging; however, CT forms the mainstay currently. Treatment consists of prolonged intensive intravenous antibiotic regimen, although as of yet there is no agreed optimum agent(s).⁸ Many studies have suggested the early instigation of surgical management in conjunction with medical management, particular if there are distant septic emboli or collections.^{2 3 8} The few case reports that describe septic arthritis highlight the importance

Patient's perspective

- ▶ I do not remember the first few days of being hospital but do remember before and afterwards. I had a sore throat due to tonsillitis and started getting shoulder pain a few days later. This was getting worse and painkillers were not helping. When I went into hospital I felt really unwell, was in severe pain and my family were very worried about me. I spent several days on the intensive care unit but eventually got better.
- ▶ Leaving hospital has only been the start of my recovery. It has taken about 4 months to get back to my usual self with a normal working shoulder. I was off work and driving for about 2 or 3 months. The physiotherapy team and the hydrotherapy were very useful in helping my recovery particularly.
- ▶ I hope that by sharing my story it will lead to better awareness of this condition among doctors to help patients get diagnosed and treated quickly.

Learning points

- ▶ Lemierre's syndrome is a rare but potentially fatal condition if left untreated.
- ▶ Starts with pharyngitis-like symptoms (pharyngodynia and fever).
- ▶ Can lead to septic emboli in lungs or joints presenting as chest or joint pain.
- ▶ Early diagnosis is key to preventing systemic complications and improved prognosis.

of early surgical intervention regardless of affected joint.^{9–11} It is clear that in the case presented here that the patient improved significantly with surgical intervention and we would therefore advocate early aggressive surgical input where there is evidence of a collection of infection.

Mortality rates have varied in the literature as high as up to 18%⁵; however, a recent systematic review reports a rate of 5%.⁶ This has improved dramatically since the preantibiotic era where Lemierre described a 90% mortality rate.¹ Complications include disseminated intravascular coagulation, subclavian vein thrombosis, cerebral venous sinus thrombosis and visceral metastatic infective emboli.⁹

In conclusion, we highlight the presentation of a rare infective pathology in an otherwise fit and well young patient. The patient quickly became very unwell but fortunately the use of early surgical intervention in combination with intensive care support and medical treatment resulted in a good functional outcome. We feel that successful management relies on increased awareness, a high index of suspicion and early intensive treatment.

Contributors JBO created initial manuscript. KA-H and BB revised manuscript, made adjustments and additions.

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