Lemierre's syndrome: the link between a simple sore throat, sore neck and pleuritic chest pain

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ABSTRACT

An unusual case of tonsillitis which showed progression to this rare syndrome despite treatment with intravenous antibiotics. Lemierre's syndrome is a rare condition characterised by a triad of: sepsis, thrombophlebitis of the internal jugular vein along with pleuropulmonary and/or distant metastatic abscesses. Diagnosis rests on a high index of suspicion and is confirmed by culture of *Fusobacterium* spp. from blood or infected sites. Radiological investigations are established aids for confirmation of diagnosis. Treatment is with appropriate antibiotics for at least 6 weeks and surgical drainage of abscesses if required. This case has been presented to highlight the mode of presentation, diagnostic tools employed and the management of the complications that featured in this condition.

KEYWORDS

Oropharynx – Sepsis – Jugular vein – Thrombosis

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Lemierre's syndrome is characterised by a triad of oropharyngeal sepsis, thrombophlebitis of the internal jugular vein and distant metastatic abscesses. It has an incidence of 1 per million per year and classically is seen in young healthy adults, with a mean presenting age of 20 years. Since its apt description by Lemierre in 1936, its presentation has changed with the advent of antibiotics. Difficulty may be encountered in reaching a diagnosis. Early recognition and prompt diagnosis is essential to facilitate treatment and prevent progression to numerous catastrophic complications as described in the literature. Lemierre's syndrome has a high morbidity and a considerable potential for fatality.

Treatment is with antibiotics for at least 6 weeks. The main causative organism, *Fusobacterium necrophorum*, is extremely sensitive to penicillin although cases of resistance have been reported.¹ Excision of the internal jugular vein may be necessary in protracted cases where this may otherwise continue to act as a septic focus. Anticoagulation may be necessary, but there have been no randomised controlled trials to confirm its role or efficacy.

Case report

A 17-year-old Caucasian male presented with an 11-day history of sore throat and general malaise. He had no significant risk factors for sexually transmitted diseases and was not known to suffer from recurrent sore throats.

On admission, his baseline observations were normal. Oropharyngeal examination showed only a mild, generalised erythema of the pharynx. The tonsils were not enlarged although the patient's mother mentioned that his tonsils were 'virtually meeting in the midline' at the onset of his symptoms. There was no neck swelling, tenderness or any significant lymphadenopathy. An initial diagnosis of acute tonsillitis was made and he was commenced on intravenous benzylpenicillin and metronidazole after a throat swab and blood specimens had been taken. Blood results showed a significantly raised white cell count of 36.3 with a marked neutrophilia of 55.4. Liver function tests were mildly deranged but Paul Bunnell test was negative. Throat swabs and serial blood cultures all returned negative.

A day after his admission, he complained of left-sided chest pain. A chest X-ray showed the appearance of a cavitating lesion in the left upper lobe (Fig. 1). At initial review, a provisional diagnosis of an atypical left lobar pneumonia was made and clarithromycin was added to his antibiotic regimen. His chest pain gradually worsened and on about the third day he developed cellulitis on the anterior aspect of the neck, worse on the left side. Contrast CT scan showed thrombosis of the left internal jugular vein, a small abscess collection adjacent to the left submandibular gland, generalised soft tissue thickening (Fig. 2) and multiple bilateral pulmonary abscesses (Fig. 3), confirming the diagnosis of Lemierre's syndrome. There was no evidence of intra-abdominal suppuration.

Figure 1 Chest X-ray showing a cavitating lesion in the upper lobe of the left lung.

on the abdominal CT scan. The patient was started on full anticoagulation with warfarin. Echocardiogram performed on the eighth day showed no evidence of vegetations on the heart valves. On the eleventh day following admission, he spiked a temperature of 38.5°C with a swinging pattern. Clinical examination of the chest showed decreased air entry at the left lung base, with stony, dull percussion notes. Chest ultrasonography confirmed a loculated empyema at this site. This was drained by video-assisted thoracoscopy, with two residual chest drains to facilitate lung re-expansion.

Figure 3 Contrast-enhanced CT scan of the chest showing bilateral multiple lung abscesses.

Figure 2 Contrast-enhanced CT scan of the neck showing a thrombosed left internal jugular vein.

Microscopy and culture of pus drained was negative for organisms. On day nineteen, he was noticed to have haemoglobin of 7.0 and was transfused with three units of blood. He made good progress subsequently. HIV screening was negative and he was found to be immunocompetent.

A repeat chest CT scan after 2 weeks confirmed reduction in the size of pulmonary abscesses. The patient was discharged on oral antibiotics for 6 weeks and anticoagulation for 3 months. Chest radiograph after 10 weeks showed a complete resolution and full lung expansion. The previously thrombosed left internal jugular vein was completely re-canalised on ultrasonography.

Discussion

Lemierre's syndrome is a rare medical condition caused by an acute oropharyngeal infection progressing to thrombophlebitis of the internal jugular vein with frequent complication by metastatic infections. Other primary sites of infection that have been implicated include the mastoid bone, middle ear, appendix, endometrium and the urinary tract. Pharyngotonsillitis accounts for at least 85% of cases but oropharyngeal primary infection may also be odontogenic in origin.² In the pre-antibiotic era, numerous cases were described of an acute medical condition typically featuring an anaerobic oropharyngeal infection leading to septic thrombophlebitis of the

internal jugular vein. This became known as Lemierre's syndrome in 1936 after André Lemierre⁵ wrote a succinct article in *The Lancet* on this subject which he described as 'postanginal septicaemia', a term attributed first to Shottmuller in Germany in 1918. It usually occurs in healthy young adults, with a mean age of 20 years. The most common causative organism is *Fusobacterium necrophorum*, a Gram-negative obligate anaerobe present normally in the nasopharynx, GI tract and female genital tract. It can usually be cultured from blood and sites of metastatic infection.

F. necrophorum (necrobacillus) has a predisposition to abscess formation. Its production of endotoxins accounts for its pathogenicity: (i) leukocidin (white cells lysis); (ii) haemolysin (red cells lysis); and (iii) haemagglutinin (platelet aggregation and septic thrombi formation).

Mortality from this condition in the pre-antibiotic era approached 90%, but from about 1950 onwards this has reduced although the current literature would suggest a figure as high as 20%. Of the 20 patients originally described by Lemierre, only two survived. A high index of suspicion is necessary to make a prompt diagnosis, as there is often a delay of one week or more between the initial infection and progression to Lemierre's syndrome. Lemierre's syndrome has an incidence of one per million per year.

There are three classical stages in its development: (i) primary infection – infection of deep pharyngeal tissue; (ii) lateral pharyngeal space tissue invasion, resulting in thrombophlebitis of the internal jugular vein; and (iii) metastatic spread of bacteria/septic emboli.

Infection of the deep pharyngeal tissue allows anaerobic organisms to drain into the lateral pharyngeal space. This space is the shape of an inverted cone and extends from the hyoid to the base of the skull. Once there is invasion of the internal jugular vein, septic embolisation to the lungs and distant sites results. Pleuropulmonary manifestations are commonest (79.8%)⁴ and present usually with multiple abscesses in the lung parenchyma or empyema, but pneumothorax is also well described. Distant embolisation probably results from haematogenous seedlings. Spread of infection can also result in external jugular vein thrombosis,⁵ carotid artery thrombosis,⁶ thrombophlebitis of abdominopelvic veins, meningitis, osteomyelitis, septic arthritis, paravertebral abscess, or soft tissue abscesses. Intracranial spread may result in meningitis, intracranial venous sinus thrombosis or cranial nerve palsy.⁷ In the pre-antibiotic era, up to 12% developed endocarditis.

Lemierre's syndrome has become rare since the advent of antibiotics and when it occurs diagnosis is, therefore, now more likely to be missed. Early recognition and prompt treatment by the clinician is essential.

Treatment is with prolonged antibiotics (at least 6 weeks)⁸ and drainage of metastatic collections where necessary. If signs of sepsis persist despite treatment, ligation and excision of the internal jugular vein may be required to remove this septic focus. Several authors have suggested anticoagulation for this rare condition. Evidence would suggest that it is done at the discretion of the clinician as there is insufficient data to establish its role. It is believed by some authors to speed up clot resolution and overall improvement in patients with Lemierre's syndrome.^{2,9,10}

Conclusions

Lemierre's syndrome is a rare condition with a high morbidity and potential for fatality. The unwary clinician may easily overlook its diagnosis. Treatment also cuts across several specialties and would easily involve the general practitioner, ENT surgeon, haematologist, chest physician and thoracic surgeon as this case has shown.

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