Lemierre’s syndrome: A persistent unusual neck pain and swelling

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Abstract

Lemierre’s syndrome is a systemic complication commonly caused by oropharyngeal infection by Fusobacterium species, which manifests itself as an internal jugular vein thrombosis formation. It is a rare occurrence nowadays with the availability of broad spectrum antibiotics for treatment. Most cases in the literature presented with a life-threatening condition. We are reporting a case of Lemierre’s syndrome that presented with persistent neck pain and swelling, initially diagnosed as cervical lymphadenitis.

Introduction

Lemierre’s syndrome is a life-threatening condition, first reported by Courment and Cade in 1890, and later described by Andre Lemierre in 1936.1 It is a condition that affects mostly young, otherwise-healthy immunocompetent adults and is characterized by a sequence of symptoms commonly following a recent history of oropharyngeal infection.2 The patient typically presents with an acute oropharyngeal infection subsequently complicated by thrombophlebitis of the internal jugular vein and distant septic thromboemboli, which can eventually lead to multi-organ failure.2 The incidence of Lemierre’s syndrome decreased to a great extent with the introduction of antibiotics in the 1940s.1,2,5 We are reporting a case of a patient with a provisional diagnosis of cervical lymphadenitis whose ultimate diagnosis was Lemierre’s syndrome.

Case Report

A 15-year-old boy presented to our centre with a one-week history of right-sided neck pain associated with odynophagia, dysphagia, and intermittent fever. This was preceded by a sore throat and fever, which was treated as acute tonsillitis two weeks prior to this presentation. Examination revealed tenderness over the right side of the neck with palpable swelling over the right levels III and IV cervical regions, which was firm in consistency with an ill-defined margin (Figure 1). The ear, nose, and throat examination showed bilateral non-erythematous grade II tonsils. Blood investigations showed leukocytosis (27600/μL) and a raised erythrocyte sedimentation rate of 92mm/hr. The renal, liver, and coagulation profiles were normal.

He was admitted and treatment was started with intravenous amoxicillin-clavulanic acid. However, the patient remained feverish in the ward. In view of the lack of improvement in the patient’s condition despite intravenous antibiotics, further investigation with ultrasound and computed tomography (CT) scan of the neck was carried out. These revealed an extensive right internal jugular thrombosis, beginning at the level of the thyroid isthmus and extending inferiorly to the retrosternal region (Figures 2 and 3). Blood culture and sensitivity (C&S) grew Fusobacterium necrophorum. Collectively, these findings confirmed the diagnosis of Lemierre’s syndrome.

He was continued on intravenous amoxicillin-clavulanic acid with the addition of metronidazole for a total duration of 2 weeks, as well as anticoagulant therapy.

Figure 1. Lateral and anterior view of the neck shows fullness of the right neck region.
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Figure 2. Ultrasonography of neck shows a right internal jugular thrombus

Figure 3. Contrast-enhanced computed tomography of neck shows a filling defect in the right internal jugular vein at the root of neck.

Discussion

Lemierre’s syndrome is a systemic complication of an oropharyngeal infection which manifests itself as an internal jugular vein thrombosis formation. Even though the majority of the cases of Lemierre’s syndrome originate from an oropharyngeal infection, it can also begin in the gastrointestinal, genitourinary, and lower respiratory tracts. Without proper diagnosis and prompt treatment, Lemierre’s syndrome can have an aggressive course. It carries a mortality rate of 5 – 18%. The common presentations of this syndrome are sore throat, fever with chills and rigors, cervical lymphadenopathy, neck pain, and swelling. Septic embolism to the lungs is extremely common (>80%). Septic arthritis (13-27%), osteomyelitis (0-9%), skin and soft tissue lesions (0-16%), and abnormal liver function with jaundice (11-49%) are possible complications. In up to 18% of cases, the patient might develop a potentially fatal complication, such as septic shock. Rarely will a patient present with central nervous system manifestations (<3%). Cavernous sinus septic thrombosis complication have been reported; these may be due to retrograde propagation from the internal jugular thrombosis.

To date, the pathogenesis of Lemierre’s syndrome has yet to be elucidated. However, several theories have been proposed. First of all, it is possible that the causative microorganism invades into the lateral pharyngeal wall via lymphatic drainage. This would then cause perivenous inflammation, subsequently leading to luminal thrombosis. Another possible mechanism that might play a role in the occurrence of Lemierre’s syndrome is that the bacteria penetrates through the pharyngeal mucosa, which has had its structure altered by the preceding viral or bacterial pharyngitis.

Before antibiotics were introduced in the 1940s, Lemierre’s syndrome carried a mortality rate of approximately 90%. With the widespread use of antibiotics in the treatment of pharyngitis, the incidence of Lemierre’s syndrome decreased drastically. Nonetheless, since the 1990s, the number of cases of Lemierre’s syndrome has been documented to be on the rise worldwide. A few recent reports postulated that this phenomenon could be attributed to the more judicious use of antibiotics in the treatment of pharyngitis, as advised by most clinical guidelines. However, more studies are warranted to investigate whether or not the rising cases of Lemierre’s syndrome in these past decades were related to less use of antibiotics in pharyngitis. Nevertheless, there is still no recommendation as to the exact duration and types of antibiotics to be used in the treatment of bacterial pharyngitis to prevent progression into Lemierre’s syndrome.

In our case, the patient had been treated for acute tonsillitis in primary care and prescribed oral antibiotics. A clinically-well patient with only mild neck pain was consistent with the diagnosis of cervical lymphadenitis, a common
reason for a palpable neck swelling. Without the use of ultrasound and the CT scan of the neck, we would have missed the internal jugular vein thrombosis in our patient. Therefore, in young adults with a unilateral neck swelling that failed treatment for pharyngitis, close monitoring should be the rule.10,11

While the causative pathogen in the majority of the Lemierre’s syndrome cases is *Fusobacterium necrophorum*, *Peptostreptococcus*, Group B and C Streptococcus, Staphylococcus, Enterococcus species, and Proteus have also been reported to be etiological agents.3

*Fusobacterium necrophorum* is still sensitive to the penicillin group, which makes it the drug of choice in treatment.10,12 Other antibiotics, such as cephalosporin, metronidazole or clindamycin, can still be used as first-line treatments for Lemierre’s syndrome.11 A 3–6 weeks course of intravenous antibiotic therapy is advocated and given until the patient is afebrile, and anticoagulation may be necessary to prevent propagation of the internal jugular vein thrombosis towards the cavernous sinus.11 Ligation and excision of the internal jugular vein is only reserved for intraluminal collection.5

In our case, treatment began with amoxicillin-clavulanic acid, and metronidazole was then added after the diagnosis of Lemierre’s syndrome was confirmed. Both agents were given for a total duration of two weeks. In addition, anticoagulant therapy was started for the treatment of the internal jugular vein thrombosis.

**Conclusion**

Patients’ presentation of neck swelling and fever may not only be consistent with diagnoses of cervical lymphadenitis, tumour, or neck abscess. Lemierre’s syndrome should be entertained as a differential if the appropriate constellation of symptoms occurs after a history of bacterial pharyngitis. The diagnosis of Lemierre’s syndrome is clinched with radiographical evidence of an internal jugular vein thrombosis and positive culture, mainly of *Fusobacterium* species.

**Take-home message:**

1. Lemierre’s syndrome is a rare, but serious, complication which may occur after an oropharyngeal infection.
2. Penicillin is still the drug of choice for Lemierre’s syndrome.
3. Persistent neck swelling and tenderness with high grade fever, following an oropharyngeal infection, should heighten one’s suspicion of Lemierre’s syndrome, and early referral for further investigation is the rule.

**References**

