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An Unusual Encounter with Lemierre's Syndrome in an Eleven-Year Pediatric Case: A Case Report

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Abstract

Lemierre's syndrome is a rare but life-threatening condition characterized by septic thrombophlebitis of the internal jugular vein, often following oropharyngeal infections such as pharyngitis. Prompt diagnosis is crucial because early intervention with appropriate antibiotic therapy can significantly reduce morbidity and mortality. Diagnosis is typically confirmed through contrast-enhanced CT imaging and blood cultures, with Fusobacterium necrophorum being the most commonly isolated pathogen. Treatment involves prolonged antibiotic therapy targeting anaerobes, typically for 4-6 weeks, and sometimes surgical drainage of abscesses. Although the role of anticoagulation is still debated, it may be indicated in cases with thrombus progression or persistent infection. Early recognition and aggressive management are key to improving outcomes and preventing fatal complications.

Keywords: Lemierre's Syndrome; IJV Thrombosis; Fusobacterium Necrophorum

Abbreviations

TLC: Total Leukocyte Count; IJV: Internal Jugular Vein; CT: Computed Tomography.

Introduction

Lemierre's syndrome, also known as 'post anginal septicemia', a relatively rare condition, is septic thrombophlebitis of the internal jugular vein (IJV). It usually occurs after an oropharyngeal infection [1]. The most common bacteria that causes Lemierre syndrome is Fusobacterium necrophorum. The time interval between initial infection and septicemia

onset is very short [2]. Delays in diagnosis are common due to the nonspecific initial symptoms, which can easily be mistaken for other more common conditions. Failure to recognize the syndrome early can lead to the progression of thrombosis and septic embolism, particularly to the lungs and other distant organs, increasing the risk of severe complications and death. Early diagnosis with the start of antibiotics can be curative as delay in diagnosis and treatment can be potentially fatal. Here we are presenting a case of left-sided neck swelling and fever, in whom there was a history of sore throat with tonsillar hypertrophy which led to IJV thrombosis.

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Case Report

An 11-year-old male presented to the ENT outpatient department with a 3-day history of fever and swelling on the left side of the neck. The patient reported a sore throat 3 weeks earlier but did not receive any treatment for it. His medical history included Guillain-Barre syndrome, for which he had been treated with IV immunoglobulin (IVIG).On examination, the patient's general condition was poor, with a temperature of 101°F, a pulse rate of 110/min, and a blood pressure of 116/76 mmHg. Local examination revealed a

diffuse, firm, tender swelling over the left side of the neck in the sternocleidomastoid (SCM) region, extending from the mastoid tip to the sternoclavicular junction. The SCM appeared bulky and shortened, and torticollis was noted, with the head tilted toward the left side Figure 1. Left-sided level 1b lymphadenopathy was also observed. Oral examination showed a congested tonsillo-pharyngeal wall with grade II tonsillar hypertrophy. Chest auscultation was normal, and the rest of the systemic examination was unremarkable.



Figure 1: Picture showing tilting of head of the patient towards left side (Torticollis).

Laboratory results were largely normal, except for an elevated total leukocyte count (TLC) of 18,000 and CRP- 140 mg/L. Ultrasonography of the neck showed an edematous soft tissue adjacent to the left lobe of the thyroid, with a 1.4×0.7 cm lymph node at level 2b. The left internal jugular vein (IJV) appeared thrombosed and dilated. A contrast-

enhanced CT (CECT) of the neck revealed a 4x2.5x2 cm heterogeneous mass in the inter/intramuscular plane of the left supraclavicular region, likely an abscess Figure 2, along with thrombosis of the left IJV and subcentimeter lymphadenopathy in levels II and III on the left side.

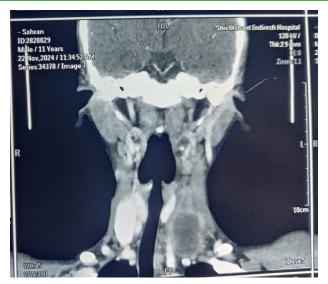


Figure 2a: Showing left sided supraclavicular neck abscess.



Figure 2b: Showing left sided Internal jugular vein (IJV) thrombosis.



Figure 3a: Showing intraoperative picture after incision and drainage



Figure 3b: Picture of secondary suturing which was done 5 days after incision and drainage.

The patient was started on intravenous antibiotics, including piperacillin-tazobactam, metronidazole, and gentamicin. Therapeutic anticoagulation therapy was initiated for 2 weeks, after which it was discontinued. Incision and drainage of the abscess was performed Figure 3, and surprisingly the pus culture showed no growth. The patient completed a 4-week course of antibiotics and is clinically better.

Discussion

Lemierre's syndrome is an uncommon yet serious condition characterized by septic thrombophlebitis of the internal jugular vein, often originating from bacterial infections in the head and neck region. Historically, in the pre-antibiotic era, Lemierre's syndrome had a mortality rate as high as 90%. Despite advances in antimicrobial therapies, recent studies indicate that mortality rates remain significant, reaching up to 20% even with appropriate treatment, highlighting the critical need for prompt diagnosis and comprehensive management strategies [3].

It was first described by André Lemierre in 1936 and is characterized by septic thrombophlebitis of the internal jugular vein usually as a sequelae of oropharyngeal infections especially pharyngitis. Although Fusobacterium necrophorum, a gram-negative obligate anaerobe, is the most common cause of the syndrome, there are also reports of other pathogens, including anaerobic streptococci, Bacteroides species, and Peptostreptococcus species, being associated with cases of Lemierre's syndrome, highlighting its polymicrobial potential [4].

Fusobacteria spp. comprise a part of the normal flora of the upper respiratory tract of human beings and are found in great numbers in the gingival crevice and subgingival plaque of all healthy adults. Pathogenic transformation of the oral commensal may occur, with invasion of the tonsillar and peritonsillar veins resulting in septicemia. It is thus not surprising that oropharyngeal sepsis is a frequent source of fusobacterial septicaemia [1].

The pathophysiological hallmark of Lemierre's syndrome is the extension of an infection from the oropharynx to the parapharyngeal space and then to the internal jugular vein with resultant thrombophlebitis. This progression is based on the anatomical location of pharyngeal veins to internal jugular vein, which are valveless and responsible for hematogenous spread. Endothelial damage from F. necrophorum leukotoxins or other bacterial virulence factors will cause thrombus formation and septic embolization to distal organs (especially lungs).

This syndrome must be diagnosed clinically based on a high index of suspicion especially, in cases presenting with lowgrade fever, sore throat, neck pain, and clinical features of septicemia/metastatic infective focus [5]. Historically, three criteria have been used to make the diagnosis of Lemierre's syndrome:

- Primary site of infection in the head or neck (commonly, a history of sore throat or anginal illness).
- Thrombosis or thrombophlebitis of internal jugular vein or other vein of head/neck district, or metastatic lesions.
- Isolation of F. necrophorum from blood culture or a site that is normally sterile [6].
- Our case fulfilled two of three defined criteria.

Contrast-enhanced computed tomography (CT) is critical in revealing internal jugular vein thrombosis and septic emboli, the classic findings of Lemierre's syndrome. Blood cultures help confirm the microbiological diagnosis, Fusobacterium necrophorum has been commonly isolated under anaerobic conditions. However, the fastidious nature of this organism necessitates prolonged incubation and the use of anaerobic blood culture bottles, which can result in false negatives and missed diagnoses, highlighting the importance of meticulous microbiological methods [5].

The primary treatment for Lemierre's syndrome involves prolonged antibiotic therapy, typically lasting 4-6 weeks, with particular attention to anaerobic bacteria. Broadspectrum antibiotics, such as beta-lactams (e.g., piperacillintazobactam, carbapenems), combined with metronidazole, are commonly used to ensure comprehensive coverage. Additionally, surgical drainage of any abscesses formed is often required to manage complications. The role of anticoagulation in Lemierre's syndrome remains unclear, as current evidence is primarily based on case reports. While it is suggested that anticoagulation might help prevent the propagation of thrombus or reduce septic embolic events associated with internal jugular vein (IJV) thrombosis, there is no definitive consensus on its routine use. Anticoagulation therapy is generally recommended if there is evidence of thrombus progression or persistent fever and bacteremia after one week of appropriate antibiotic treatment [7].

Conclusion

Lemierre's syndrome is a rare but serious condition that requires early diagnosis and aggressive management due to its potential for severe complications, including septic embolism and organ failure. Timely identification, especially in patients with head and neck infections, is crucial to initiate appropriate antibiotic therapy and prevent mortality. Although the role of anticoagulation is still debated, it may be indicated in cases with thrombus progression or persistent infection. With proper treatment, outcomes can improve, but the high mortality rate underscores the need for heightened clinical awareness.

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