CASE REPORT

Lemierre's Syndrome: A Retrospective Case Series

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Abstract

Introduction: Lemierre's syndrome is a rare and less commonly occurring clinical condition that generally relates to the septic thrombophlebitis of the Internal Jugular vein. Common bacteria causing infection are *Streptococci* followed by *Styaphylococci* and *Klebsiella*. The major proportion of cases that come to light are due to deep neck space infections that ultimately leads to thrombotic involvement of IJV, accounts for majority of the cases. The other causes include complications of chronic suppurative otitis media and thrombosis occurring in deep veins. Since the Internal Jugular Vein is involved the infection can also undergo hematogenous spread. The infection generally spreads to Spleen, Liver, Kidney, Heart and Brain. Lemierre's syndrome can be diagnosed on the basis of clinical symptoms, multitude of blood series and Imaging. Since it is an infectious condition the treatment involves systemic antibiotic therapy and early administration of broad spectrum antibiotics in high dose become necessary for prevention of complications and systemic spread of infection. We are presenting a case series of 5 cases over a period of six years from November 2015 to November 2021.

Materials and methods: This case series focused on the cases of Lemierre's syndrome that presented in the MBS Hospital, Kota over a course of 2 years from November 2019 to November 2021. The cases that were encountered were admitted, diagnosed and treated uneventfully. The investigations that the patients underwent were: Blood cultures, Chest skiagrams, Contrast enhanced CT, Skiagrams of Mastoid bone, Throat cultures and thorough clinical and ENT examination.

Results: The observation and analysis of all the 5 recorded cases reveals that 2 cases of Lemierre's syndrome were having deep neck space infection as a causative factor in development of the disease and the other 2 cases were due to complications of chronic suppurative otitis media and the last remaining case had deep venous thrombosis as a causative factor for involvement of IJV and development of Lemierre's syndrome. Early administration of broad spectrum antibiotics is absolutely necessary on suspicion of Lemierre's syndrome so as to prevent adverse clinical outcome.

Keywords: Hematogenous spread, Internal jugular vein, Internal jugular vein thrombosis, Neck infection, Neck swelling, Septicemia, Tenderness, Thrombophlebitis.

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Introduction

In 1936, André Lemierre described a syndrome with post-anginal septicemia, which was complicated with thrombosis of the IJV and the presence of distant septic emboli in the patient's bloodstream.[1,2] It has gained a special status due to few cases coming to light and as a "forgotten" and a condition which is often missed in diagnosis resulting in various complications and poor patient outcomes and sometimes may result in mortality.[3–5] André Lemierre explained that septic emboli reaching IJV could originate from many sites such as nasopharynx, oral cavity, ear having otitis media, mastoiditis, uterus having purulent endometritis, and appendicitis.[4] The involvement of IJV provides a pathway for the spread of infection through bloodstream. The symptoms are tenderness in neck region, pain, fever with chills and rigors, and erythema in pharyngeal and peritonsillar region on oropharyngeal examination. Since the IJV is involved, the infection can potentially undergo hematogenous spread. The infection generally spreads to spleen, liver, kidney, heart, and brain.[6,7] Lemierre's syndrome can be diagnosed based on clinical symptoms, multitude of blood series, and imaging. Since it is an infectious condition, the treatment involves systemic antibiotic therapy, and early administration of broad-spectrum antibiotics in high dose becomes necessary for prevention of complications and systemic spread of infection.[8,9]

Epidemiology

Lemierre's syndrome is generally seen in young adults.[10] According to a study carried out in Denmark, there was an annual incidence of 3–6 cases of Lemierre's syndrome per million people from 1998 to 2001, with a significantly higher annual incidence of 14.4 cases per million people with age-group of 14–24 years.[10,11]

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Clinical Features

In most of the cases, oropharynx is the primary site of infection and exudative tonsillitis may be present in many cases, and sometimes just mild hyperemia and ulcers in the oropharynx and peritonsillar area are noted in some cases. The general signs are tenderness and hyperemia over the angle of jaw and pain with neck movement, which is sometimes associated with

trismus. The time period between the onset of symptoms and development of septicemia and bacteremia may be a week or sometimes even less in some cases. When the bacteria enter the bloodstream, the most common sites of septic emboli are within the lungs and it often is a source of diagnostic dilemma as the presence of lung emboli moves clinician toward deep vein thrombosis (DVT).[12] After that, the presence of infiltrative exudates on skiagrams is a common finding and development of lung abscess and pleural effusion ensues.[13,14] Pneumatoceles and pneumothorax have also been reported. Hepatomegaly and splenomegaly are common, but splenic and hepatic abscess is rare.[15] Hospital stays of these patients are quite long, ranging from a few days to a few weeks.

Diagnosis

Since the symptoms lie in a common spectrum, a high degree of clinical suspicion is often required to diagnose this clinical entity. In early course of disease, high-grade fever may be the only presenting symptom. The ear, nose, and throat (ENT) exam might not reveal any significant diagnostic findings[13] and the diagnosis is not suspected until the microbiologic culture reports are available. Any suspicion of IJV thrombosis must be objectively confirmed and the clinician must try to rule out the condition once suspected. The contrast-enhanced computed tomography (CECT) scanning is a very useful investigation in this regard. It can help in diagnosis by showing intraluminal filling defects and low-grade selling of adjacent tissues.[12] Ultrasonography (USG) is less expensive and will reveal echogenic regions of thrombosis and irregular filling defects, it is advantageous in being less expensive and invasive than a contrast-enhanced radionucleotide technetium venography scan.[16,17]

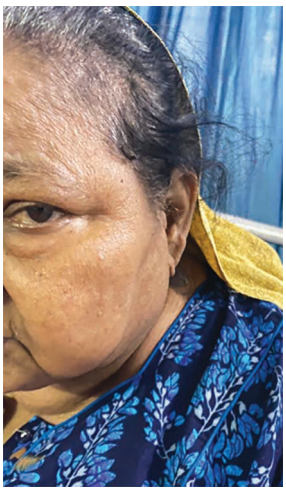
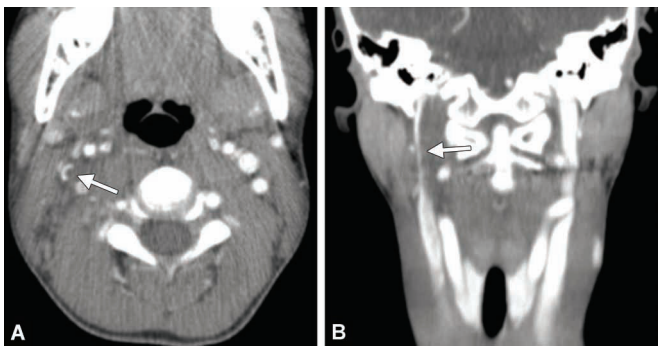


Fig. 1: Neck swelling due to deep neck space infection in a patient with Lemierre's syndrome (Source: Shivkumar, Department of Otorhinolaryngology, Kota)



Figs 2A and B: Subpart label shows IJV thrombosis visible in a patient with Lemierre's syndrome (Source: Shivkumar, Department of Otorhinolaryngology, Kota)

Materials and Methods

This case series focuses on the cases of Lemierre's syndrome that presented in the MBS Hospital, Kota over a course of 2 years from November 2019 to 2021.[18] The cases that were encountered were admitted, diagnosed, and treated uneventfully. The investigations that the patients underwent were: blood cultures, chest skiagrams, CECT, skiagrams of mastoid bone, throat cultures, and thorough clinical and ENT examination.

Inferences

A total of five cases were studied and after reviewing the investigations the inferences were drawn that two of the cases presented as a complication of deep neck space infection (Fig. 1) and the infection spreading ultimately leading to spread to the IJV and development of the Lemierre's syndrome (Fig. 2). Two cases were due to a complication of chromic suppurative otitis media (CSOM). The patients presented with chronic ear discharge and hearing loss. One case was of a 55-year-old female with a complication of DVT who had a postoperative case of prosthetic knee replacement. Four of these patients presented with chief complaints of neck rigidity and tenderness near the angle of jaw, making it the most commonly presenting complaint after fever, which was exhibited by all cases. CECT neck proved to be diagnostic as it showed peritonsillar swelling and intraluminal filling defects in the IJV in all the studied cases. USG showed echogenicity in almost all the cases but did not show any definite filling defects specifically pointing toward thrombosis. Blood cultures revealed *Streptococci*, *Klebsiella,* and *Staphylococcus epidermidis* in two cases.

Chest skiagrams of patients did not reveal any abnormalities except in that of the 55-year-old obese female having DVT due to an inactive lifestyle. Skiagrams of patients did not reveal any significant abnormalities except in those patients having a history of CSOM. The skiagrams revealed sclerosed mastoid air cells and fluid levels suggesting mastoiditis and presence of purulent material. One patient who had a deep neck space infection also had a sclerosed mastoid air cell cavity on the contralateral side of IJV involvement but had a normal ear exam making the infectious etiologies of neck space infections and oropharynx the major culprits of the IJV thrombosis and development of Lemierre's syndrome and complications of an unsafe ear and venous thrombosis other rare causes of IJV thrombosis.

Conclusion

The observation and analysis of all five recorded cases reveal that two cases of Lemierre's syndrome had deep neck space infection as a causative factor in the development of the disease, and the other two cases were due to complications of chronic suppurative otitis. Media and the last remaining case had deep venous thrombosis as a causative factor for the involvement of IJV and the development of Lemierre's syndrome. Early administration of broad-spectrum antibiotics is absolutely necessary on suspicion of Lemierre's syndrome so as to prevent adverse clinical outcomes.

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