



LEMIERRE'S SYNDROME: A REVIEW

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ABSTRACT Lemierre's syndrome is a rare and less commonly occurring clinical condition that generally relates to the septic thrombophlebitis of the Internal Jugular vein. Mostly the clinicians have forgotten about this due to less incidence, the most common bacteria causing infection are and Streptococci followed by staphylococci and Klebsiella [1]. 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 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. [3][4] 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. [3][4] We are presenting a case series of 5 cases over a period of six years from November 2015 to November 2021.

KEYWORDS :**INTRODUCTION**

In 1936, Andre' Lemierre described a syndrome with post-anginal septicemia which was complicated with thrombosis of the Internal Jugular Vein and presence of distant septic emboli in the patient's bloodstream. [5][6] 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. [7][8][9] Andre' 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. [8]. The involvement of Internal Jugular Vein provides a pathway for spread of infection through bloodstream. The symptoms are tenderness in neck region, pain, fever with chills and rigors, erythema in pharyngeal and peritonsillar region on oropharyngeal examination. Since the Internal Jugular Vein is involved, the infection can potentially undergo hematogenous spread. The infection generally spreads to Spleen, Liver, Kidney, Heart and Brain. [3][4] Lemierre's syndrome can be diagnosed based on clinical symptoms, multitude of blood series and Imaging. Since it is an infectious condition. 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.

Epidemiology

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

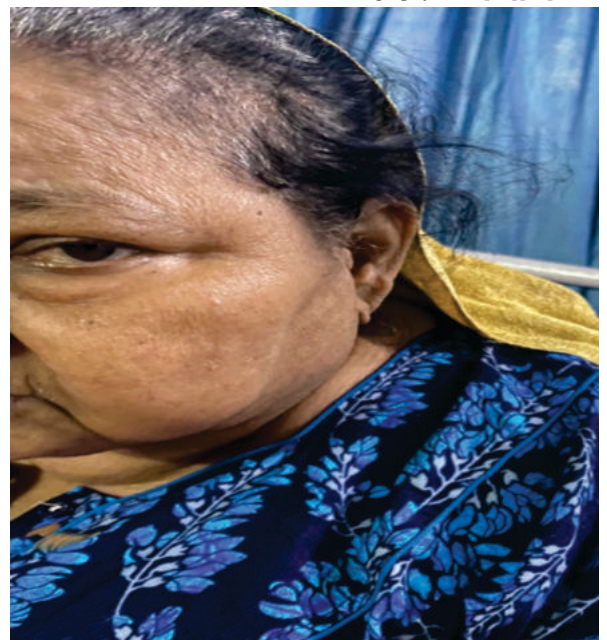
Clinical Features

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 hyperaemia and ulcers in the oropharynx and peritonsillar area are noted in some cases. [12] The general signs are the tenderness and hyperaemia over the angle of jaw, pain with neck movement which is sometimes associated with trismus. The time period between the onset of symptoms and development of septicemia and bacteraemia may be a week or sometimes even less in some cases. [12] When the bacteria enter bloodstream the most common sites of septic emboli is within the lungs and it often is a source of diagnostic dilemma as the presence of lung emboli moves clinician towards the DVT. [13] After that the presence of infiltrative exudates on skiagrams is a common finding and development of lung abscess and pleural effusion ensues. [14][15][16] Pneumatoceles and Pneumothorax has also been reported. Hepatomegaly and Splenomegaly is common but Splenic and Hepatic

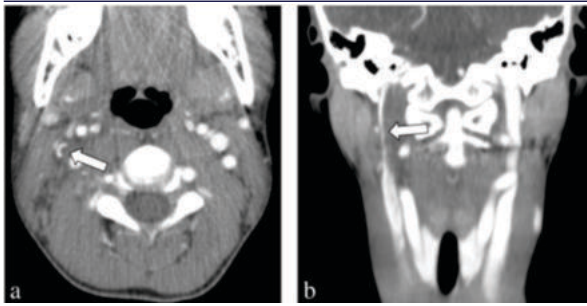
abscess is rare. [17] Hospital stays of these patients are quite long, ranging from a few days to few weeks.

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 ENT exam might not reveal any significant diagnostic findings [18] 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 scanning is a very useful investigation in this regard. It can help in diagnosis by showing intraluminal filling defects and low grade swelling of adjacent tissues. [19] 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 radionuclide Technetium venography scan. [21][22]



Neck swelling due to deep neck space infection in a patient with Lemierre syndrome Image courtesy: Shivkumar, Department of Otorhinolaryngology, Kota



IJV thrombus visible in a patient with Lemierre Syndrome Image courtesy: 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 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.

Inferences

Total of 5 cases were studied and after reviewing the investigations the inferences were drawn that 2 of the cases presented as a complication of deep neck space infection and the infection spreading ultimately leading to spread to the IJV and development of the Lemierre's syndrome. 2 cases were due to a complication of CSOM (Chronic suppurative otitis media). The patients presented with chronic ear discharge and hearing loss. 1 case was of a 55-year female as a complication of DVT who a post operative 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 common 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 studies cases. USG showed echogenicity in almost all the cases but did not show any definite filling defects specifically pointing towards the thrombosis. Blood cultures revealed Streptococci, Klebsiella and Staphylococcus Epidermidis in 2 cases.

Chest Skiagrams of patients did not reveal any abnormalities except in that of the patient 55 years obese female having deep vein thrombosis due to 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 level suggesting mastoiditis and presence of purulent material. One patient who had deep neck space infection also had a sclerosed mastoid air cell cavity on the contralateral side of IJV involvement but had 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 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.

REFERENCES:

1. Lee Wen-Sen, Jean Shio-Shin, Chen Lu-Fun, Hsieh Szu-Min, Hsueh Po-Ren. Lemierre's Syndrome: A forgotten and re-emerging infection. *Journal of Microbiology, Immunology and Infection*. 2020 August; 53(4):513-517.
2. Brook Itzhak. Fusobacterial head and neck infections in children. *International Journal of Pediatric Otorhinolaryngology*. 2015 July; 79(7):953-958
3. Srivali Narat, Ungprasert Patompong, Kittanamongkolchai Wongnarm, Ammannagari Nischala. Lemierre's syndrome : an often missed life threatening infection. *Indian Journal of Critical Care Medicine*. 2014 March; 18(3): 170-172
4. Gupta Nishant, Kralovic Stephen M., McGraw Dennis. Lemierre syndrome: not so forgotten!. *American Journal of Critical Care*. 2014 March; 23(2):176-179
5. Lemierre A. On certain septicaemias due to anaerobic organisms. *The Lancet*. 1936 Mar 28; 227(5874):701-3.
6. Osowicki J, Kapur S, Phuong LK, Dobson S. The long shadow of Lemierre's syndrome.

7. Journal of Infection. 2017 Jun 1; 74:S47-53.
7. Chen FL, Jean SS, Ou TY, Yu FL, Lee WS. Pulmonary empyema caused by co-infections of *Mycoplasma pneumoniae* and *Fusobacterium necrophorum*: A rare case of Lemierre syndrome. *Journal of Microbiology, Immunology, and Infection-Wei mian yu gan ran za zhi*. 2017 Aug; 50(4):552-4.
8. Lee WS, Wang FD, Shieh YH, Teng SO, Ou TY. Lemierre syndrome complicating multiple brain abscesses caused by extended-spectrum β -lactamase-producing *Klebsiella pneumoniae* cured by fosfomycin and meropenem combination therapy. *Journal of Microbiology, Immunology and Infection*. 2012 Feb 1; 45(1):72-4.
9. Lin HY, Liao KH, Jean SS, Ou TY, Chen FL, Lee WS. Lemierre syndrome with cervical spondylodiscitis and epidural abscess associated with direct injection of heroin into the jugular vein. *Journal of Microbiology, Immunology, and Infection-Wei mian yu gan ran za zhi*. 2015 Apr; 48(2):238-9.
10. Kristensen LH, Prag J. Lemierre's syndrome and other disseminated *Fusobacterium necrophorum* infections in Denmark: a prospective epidemiological and clinical survey. *European Journal of Clinical Microbiology & Infectious Diseases*. 2008 Sep; 27(9):779-89.
11. Ramirez S, Hild TG, Rudolph CN, Sty JR, Kehl SC, Havens P, Henrickson K, Chusid MJ. Increased diagnosis of Lemierre syndrome and other *Fusobacterium necrophorum* infections at a Children's Hospital. *Pediatrics*. 2003 Nov 1; 112(5):e380-3.
12. Sinave CP, Hardy GJ, Fardy PW. The Lemierre syndrome: suppurative thrombophlebitis of the internal jugular vein secondary to oropharyngeal infection. *Medicine*. 1989 Mar 1; 68(2):85-94.
13. Lustig LR, Cusick BC, Cheung SW, Lee KC. Lemierre's syndrome: two cases of postanginal sepsis. *Otolaryngology—Head and Neck Surgery*. 1995 Jun; 112(6):767-72.
14. Weesner CL, Cisek JE. Lemierre syndrome: the forgotten disease. *Annals of emergency medicine*. 1993 Feb 1; 22(2):256-8.
15. Seidenfeld SM, Sutker WL, Luby JP. *Fusobacterium necrophorum* septicemia following oropharyngeal infection. *Jama*. 1982 Sep 17; 248(11):1348-50.
16. Gato A, García C, Mateo O, Pontes MJ, Baez B, Pérez V. Diagnóstico por imagen en el síndrome de Lemierre. *Enferm Infecc Microbiol Clin*. 1993 May; 1:263-6.
17. Kern W, Dolderer M, Krieger D, Büchler M, Kern P. Lemierre's syndrome with splenic abscesses. *Deutsche medizinische Wochenschrift* (1946). 1992 Oct 1; 117(40):1513-7.
18. Weesner CL, Cisek JE. Lemierre syndrome: the forgotten disease. *Annals of emergency medicine*. 1993 Feb 1; 22(2):256-8.
19. Lustig LR, Cusick BC, Cheung SW, Lee KC. Lemierre's syndrome: two cases of postanginal sepsis. *Otolaryngology—Head and Neck Surgery*. 1995 Jun; 112(6):767-72.
20. Carlson ER, Bergamo DF, Coccia CT. Lemierre's syndrome: two cases of a forgotten disease. *Journal of Oral and Maxillofacial Surgery*. 1994 Jan 1; 52(1):74-8.
21. Celikel TH, Muthuswamy PP. Septic pulmonary emboli secondary to internal jugular vein phlebitis (postanginal sepsis) caused by *Eikenella corrodens*. *American Review of Respiratory Disease*. 1984 Sep; 130(3):510-3.
22. Mañé S, Torres M, Bugés J, Rivas A, Bruno C, Rodriguez E, Martinez JA, Nubiola A. Scintigraphic demonstration of jugular obstruction in a case of Lemierre syndrome. *Clinical nuclear medicine*. 1992 Mar 1; 17(3):233-5