International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

*Case Report*

# Lemierre's Syndrome-Re-Emergence of a Forgotten Disease

Gaikwad Pravin Sharad[1], Rajeev Anand[2], Mathews Baby Modathil[3], Kausar Mirza[4]

[1]DNB Radio Diagnosis Trainee (Final Year), [2]Consultant Radiologists, [3]Head of Department, Department of Radiology, [4]DNB Radio Diagnosis Trainee (Second Year),
Medical Trust Hospital, MG road, Kochi-682016, Kerala, India.

Corresponding Author: Gaikwad Pravin Sharad

*Received: 08/08/2016* *Revised: 24/08/2016* *Accepted: 29/08/2016*

## ABSTRACT

Lemierre syndrome is an uncommon disorder usually affecting young adults caused by the anaerobic bacterium Fusobacterium necrophorum and occasionally by other Fusobacterium species (F. varium, F. mortiferum and F. Nucleatum etc). It usually follows upper respiratory tract infection/sinusitis/otitis/mastoiditis/parotitis. The evidence of septic thrombophlebitis with positive blood culture, imaging features of internal jugular vein thrombosis forms the basis of diagnosis. This is known to be a disease of pre-antibiotic era as lack of antibiotic treatment forms a ground to expand the infection beyond its usual limits. The incidence rate is currently 0.8 cases per million in the general population. We present a case report of a 43-year-old man with parotid swelling, which illustrates that subsequent re-emergence of this often forgotten disease may become more common in clinical setting.

***Key words:*** Lemierre syndrome, Lemierre's syndrome, Fusobacterium, septic embolization, septic thrombophlebitis.

## INTRODUCTION

Lemierre's syndrome is a rare clinical entity with fewer than 160 cases reported since it was first described in the early 1900's. [1] The objective of this case report is to heighten awareness amongst radiologist about this syndrome to expedite diagnosis of syndrome that often manifests as non-specific clinical and radiographic findings.

## CASE PRESENTATION

A 43-year-old Indian male, with noprior medical history, was evaluated in the emergency department (ED) for an 8-days history of swelling over left parotid region, left orbital pain, fever and difficulty in breathing. He took treatment from local general practitioner for acute parotitis with Tab. azithromycin 250mg twice daily for last three days. Then he presented to our casualty with new onset worsening dyspnea. Physical examination revealed no obvious cervical lymphadenopathy. Monospot test was negative. Routine blood tests, blood cultures, chest x- ray, ultrasonography, CT scan were performed. His respiratory rate was 30 breaths per minute, temperature was 101.2°F, heart rate was 102 beats per minute, and blood pressure was 96/66 mmHg. Arterial blood gas analysis revealed patient is in respiratory acidosis. A WBC count noted to be 19,500 cells/μL with differential count of 84% granulocytes. The initial chest radiograph and USG abdomen was inconclusive. Blood cultures were drawn and sent to laboratory. Considering the patient in sepsis, he is put on broad-spectrum antibiotics. Urine legionella antigen and legionella DFA were negative.

Ultrasonography of neck and parotid region reveals left parotitis with left IJV thrombosis.

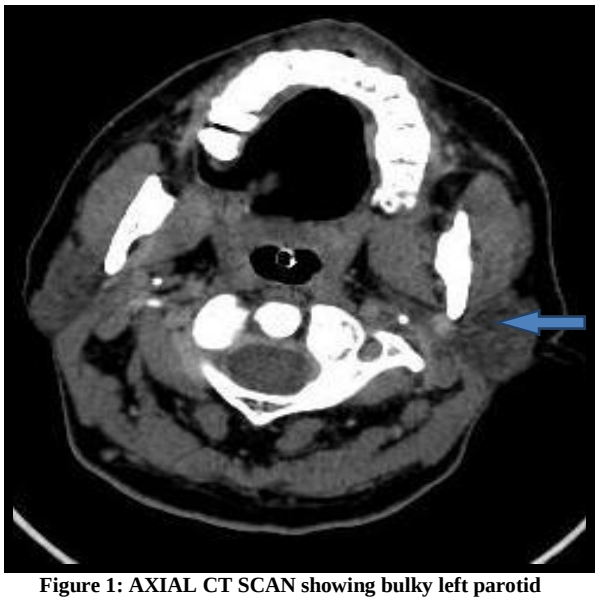

**Figure 1: AXIAL CT SCAN showing bulky left parotid**

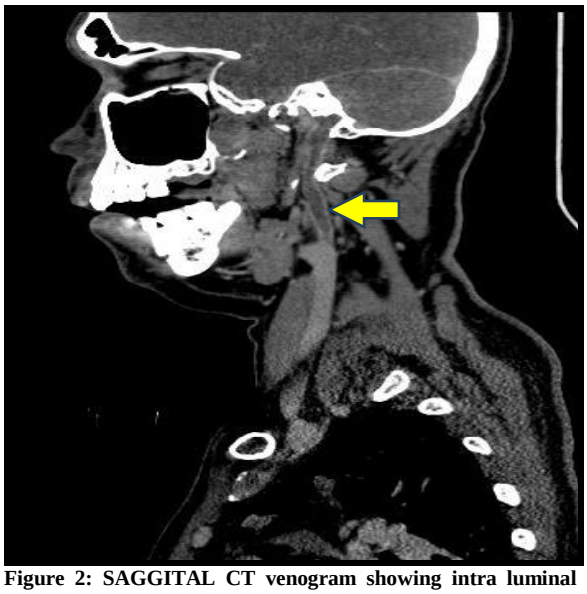

**Figure 2: SAGGITAL CT venogram showing intra luminal filling defect suggestive of left IJV thrombosis**

Forty-eight hours later, the initial gram stain of the blood indicated gram-negative rods from only the anaerobic vial. Interim blood cultures revealed Fusobacterium necroforum. Then the antibiotic regimen was changed to intravenous penicillin G and metronidazole. Based on above scenario thrombus in left IJV was supposed to be infective and search has made for shaded septic emboli. Computed tomography of the head and neck revealed left parotid inflammation, thrombosis of left IJV, left retromandibular and paravertebral vein thrombosis and bilateral cavernous sinus thrombosis (figures 1, 4, 5, 6). CT abdomen is unremarkable. Computed tomography of the chest shows bilateral septic emboli predominantly in upper lobes (figure 7, 8, 9). Computed tomography of the neck with contrast revealed lack of enhancement at the whole of the left internal jugular vein (figure 2, 3). MRI was performed to look for intra orbital extension of disease process. T2W AND STIR MR imaging of neck shows inflamed left parotid (figure 10, 11, 12) and CORONAL T2W MR imaging of posterior orbit shows left superior orbital vein thrombosis with orbital cellulitis (figure 14, 15). Evidence of bilateral cavernous sinus thrombosis also noted on coronal T2WI as heterogeneous signal intensity (figure 13).

Patient was diagnosed with Lemierre's syndrome. The patient improved in subsequent 7 days.

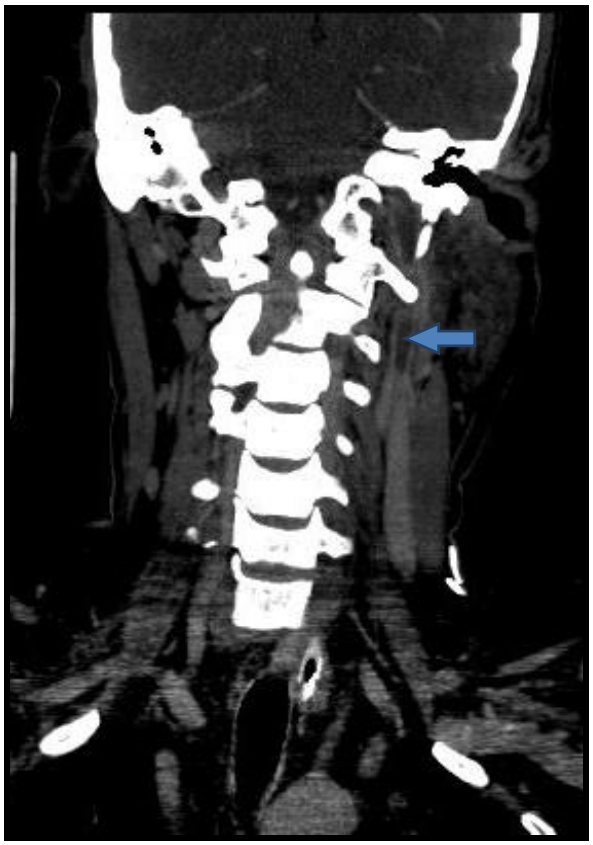

**Figure 3: CORONAL CT venogram showing left IJV thrombosis**

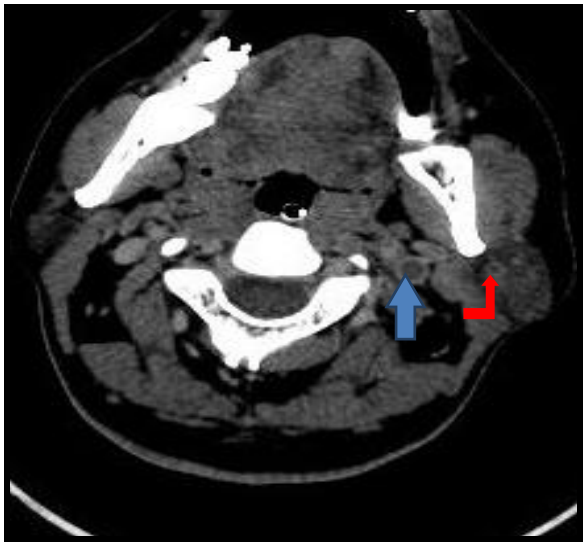

**Figure 4: AXIAL CT of neck showing left IJV and left retromandibular vein thrombosis**

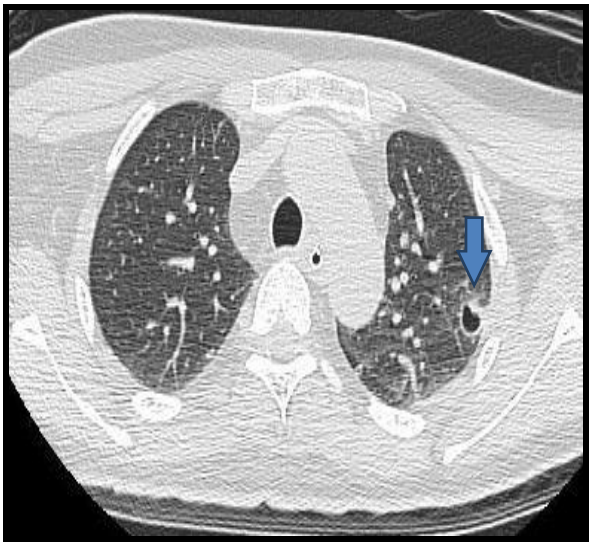

**Figure 7: AXIAL HRCT lung showing septic emboli with cavity formation in left upper lobe**

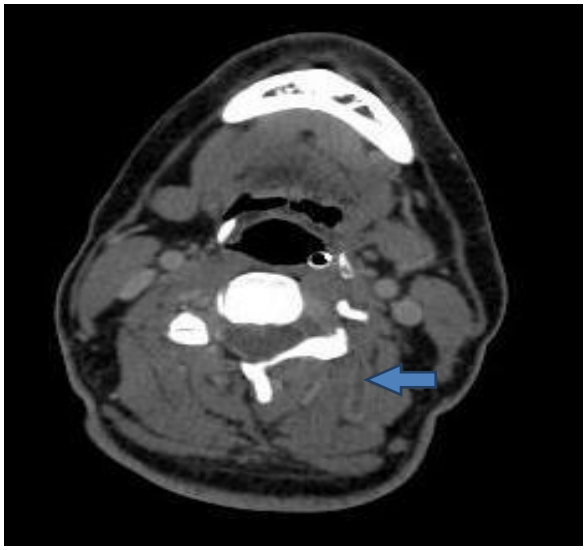

**Figure 5: axial CT SCAN SHOWING left paravertebral vein thrombosis**

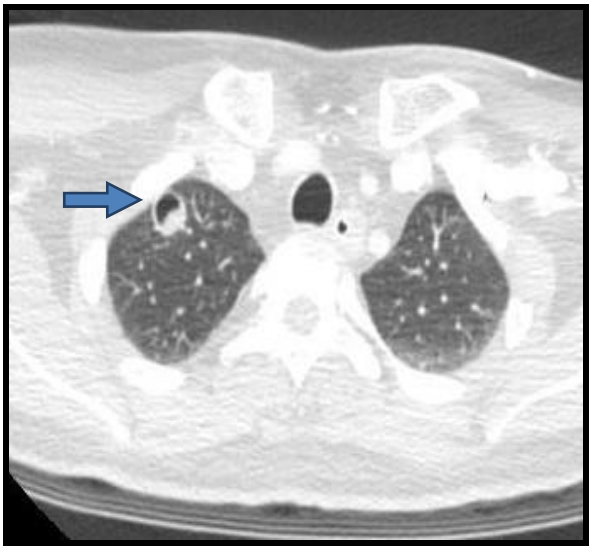

**Figure 8: AXIAL HRCT lung showing septic emboli with cavity formation in right upper lobe**

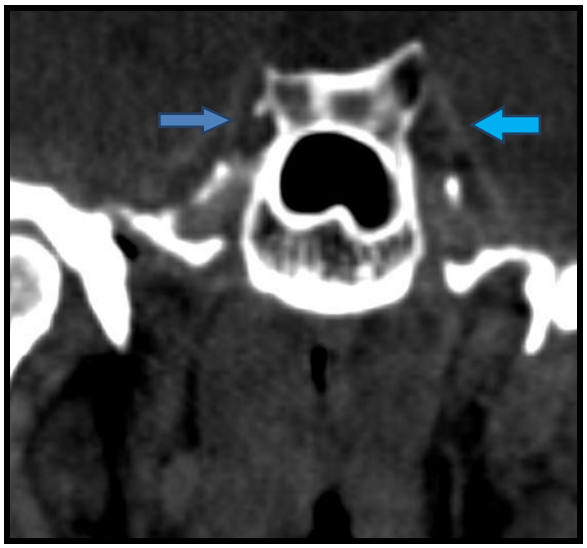

**Figure 6: CORONAL CT image showing bilateral cavernous sinus thrombosis**

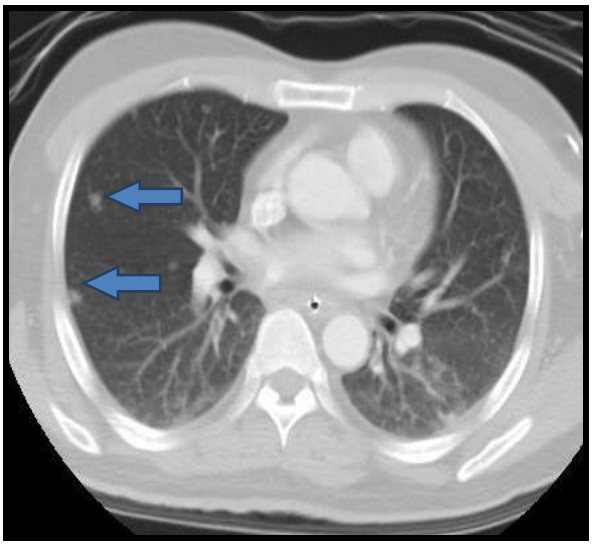

**Figure 9: CONTRAST ENHANCED AXIAL CT THORAX showing peripheral pulmonary nodule suggestive of septic emboli**

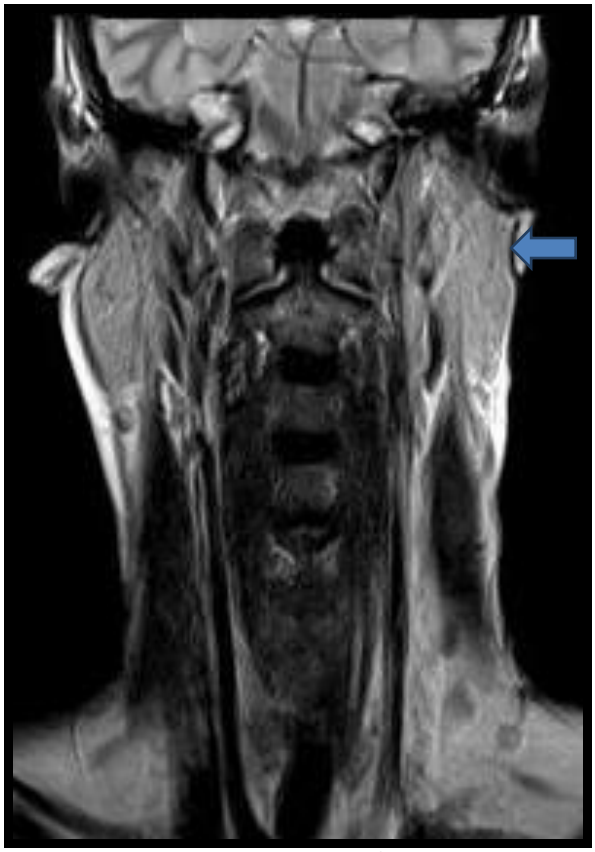

**Figure 10: CORONAL T2WI showing left bulky parotid gland with loss of surrounding fat planes**

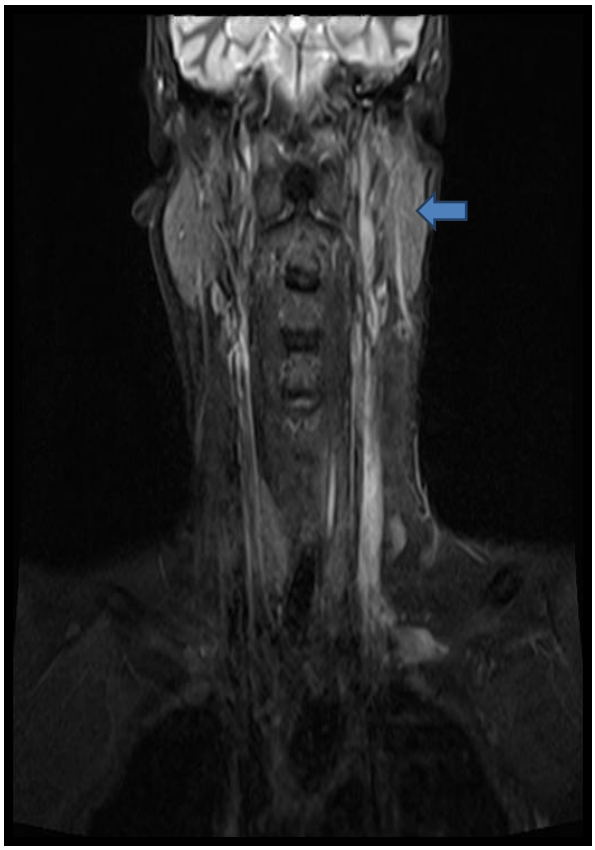

**Figure 11: CORONAL STIR image showing bulky left parotid with loss of surrounding fat planes**

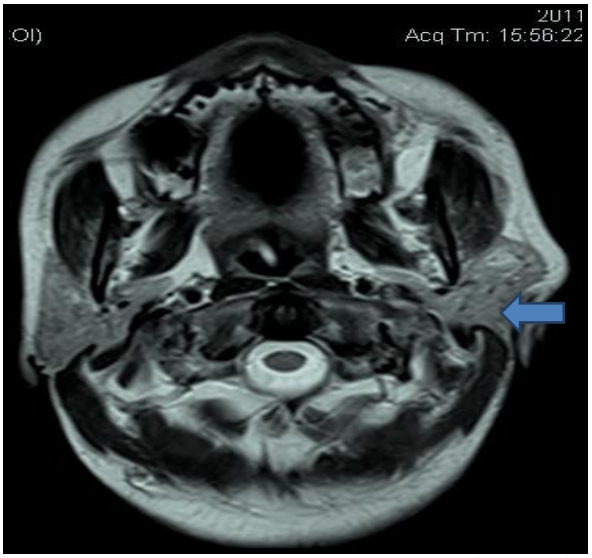


**Figure 12: AXIAL T2WI MR image showing inflamed left parotid gland**

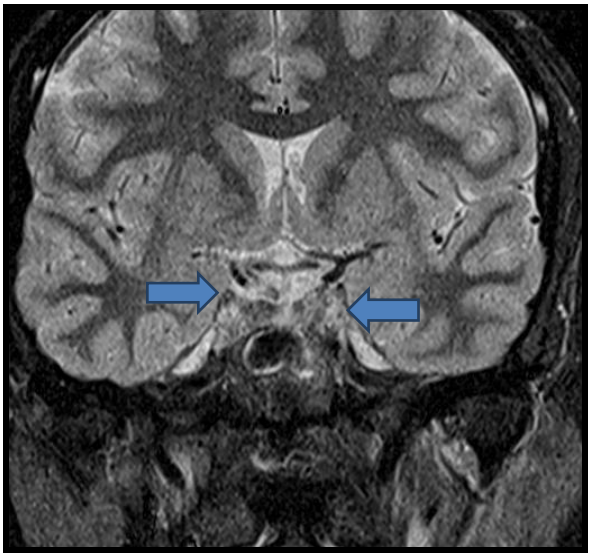

**Figure13: CORONAL T2WI MR Image showing bilateral cavernous sinus thrombosis**

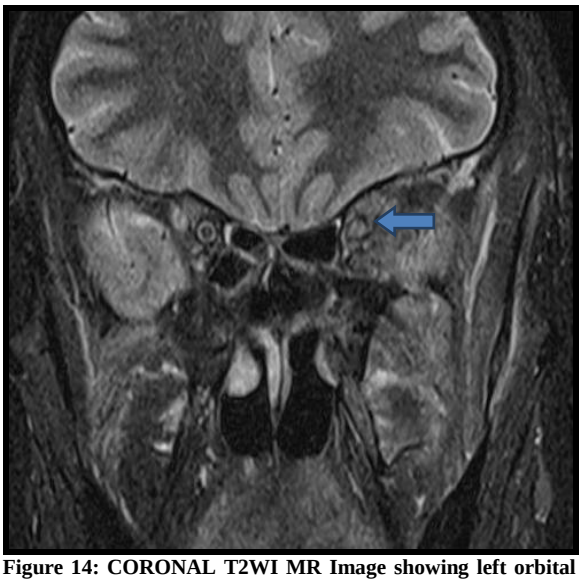

**Figure 14: CORONAL T2WI MR Image showing left orbital vein thrombosis**

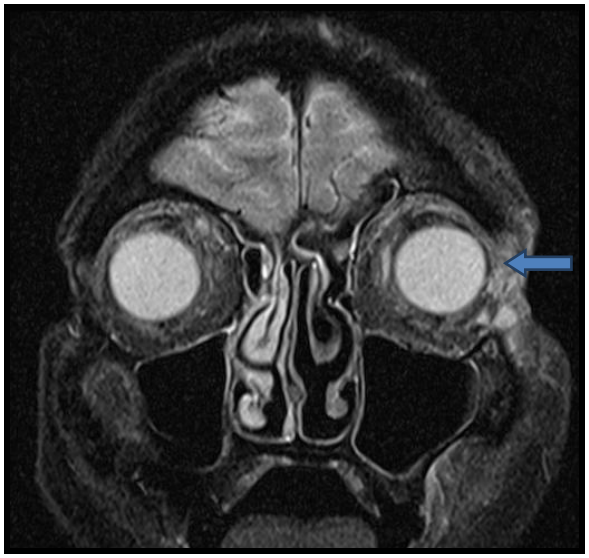

**Figure 15: CORONAL T2WI MR IMAGE showing features of left orbital cellulitis**

## DISCUSSION

Fusobacteria are obligate anaerobic, Gram-negative rods exhibits as a normal flora in the human upper respiratory tract, gastrointestinal tract, and female genital tract. Various toxins have been identified that are produced by Fusobacteria that may play a role in the pathogenicity. Hem agglutinin being one of them, its production augments the fulminant nature of the disease, causing platelet aggregation and septic thrombus formation. [2] Fusobacteriumnecrophorum and F nucleatum are the species most often the causative agents of Lemierre's syndrome, but other Fusobacterium species can occasionally be found. The exact mechanism of invasion and penetration of the pharyngeal mucosa has not been determined. Current hypothesis include the help of an underlying synergistic infectious process (bacterial or viral), with a concomitant decline in host resistance. [3] When pharyngitis due to Fusobacterium species occurs, the physical proximity of the vessels in the lateral pharyngeal space permits extension from the peritonsillar space to the internal jugular vein. This usually occurs in less than a week from the development of pharyngitis. Once invasion of the internal jugular vein is achieved, the resultant bacteraemia triggers platelet aggregation and thrombus formation. [4] Thrombusformation and rapid bacterial growth result in a nidus for metastatic septic embolization. Emboli from thrombosed neck veins metastasize to the pulmonary vasculature in up to 85% of patients, resulting in complicated pleural effusions, pulmonary abscesses, and empyema. [5] The chest radiograph typically shows multiple nodular infiltrates scattered throughout both lung fields. [6] Contrast computed tomography of the neck provides the definitive diagnosis, showing distended veins with enhancing walls, intraluminal filling defects, and swelling of adjacent soft tissues. [7] Ultrasonography can also confirm internal jugular vein thrombosis, showing localized echogenic regions within a dilated vessel. [8] Confirmation of Lemierre's syndrome is provided by demonstration of Fusobacterium species in anaerobic blood culture. [9] The recommended treatment of Fusobacterium species in Lemierre's syndrome is combination therapy with parenteral high dose penicillin and metronidazole. Intravenous clindamycin may be used in penicillin-allergic patients. [10]

## CONCLUSION

Lemierre's syndrome is a rare disorder caused by Fusobacterium species characterized by internal jugular vein thrombosis, oropharyngeal infection, septicaemia and presence of metastatic foci throughout the body.

This case has several important implications. By anticipating the poor prognosis associated with delay in diagnosis till blood culture report comes; early diagnosis with characteristic radiological findings and radiologist's familiarity with the condition provide wide scope for change in future outcome of the disease.

The differential diagnosis of Lemierre's syndrome is vast and includes viral pharyngitis, infectious mononucleosis, acute retroviral syndrome, leptospirosis, acute bacterial pneumonia, atypical pneumonia, aspiration pneumonia, infective endocarditis, and intra-abdominal abscess.

***List of abbreviations:***
°F: degrees Fahrenheit;
MmHg: millimetres of mercury, cells/ μL, cells per microliter;
DFA: direct fluorescent antibody.

## REFERENCES


1. Sara Z, Beard G, Furey W. Insight into a forgotten disease: Lemierre's syndrome. *Resid Staff Physician.* 2005; 51:37-40.
2. Singhal A, Kerstein M: Lemierre's Syndrome. South Med J, 2001, 94(9):886-887.
3. Brook I, Walker RI: The relationship between Fusobacterium species and other flora in mixed infection. J Med Microbiol 1986, 21:93-100.
4. Forrester LJ, Campbell BJ, Berg JN, Barrett JT: Aggregation of platelets by Fusobacterium necrophorum. J Clin Microbiol 1985, 22:245-249.
5. Hagelskjaer LH, Prag J, et al.: Incidence and clinical epidemiology ofnecrobacillosis in Denmark 1990-1995. Eur J Clin Microbiol Infect Dis 1998, 17:561-565.
6. Sinave CP, Hardy GJ, Fardy PW: The Lemierre Syndrome. Medicine 1989, 68:85-94.
7. Goyal M, Sharma R, Jain Y, et al.: Unusual radiological manifestations of Lemierre's syndrome: a case report. Pediatr Radiol 1995, 15(Suppl 1):S105-S106.
8. Golpe R, Marin B, Alonso M: Lemierre's syndrome (necrobacillosis). Postgrad Med J 1999, 75:141-144.
9. Stahlman GC, DeBoer DK, and Green NE: Fusobacterium osteomyelitis and pyarthrosis: a classic case of Lemierre's syndrome. J Pediatr Orthop 1996, 16:529-532.
10. Hagelskjaer KL, Prag J: Human necrobacillosis, with emphasis on Lemierre's syndrome. Clin Infect Dis 2000, 31:524-532.


How to cite this article: Sharad GP, Anand R, Modathil MB et al. Lemierre's syndrome-Re-emergence of a forgotten disease. Int J Health Sci Res. 2016; 6(9):510-515.

**********