Case Report

DOI: 10.7860/JCDR/2022/55073.16521

Lemierre's Syndrome Presenting with Pleural Effusion

Internal Medicine Section

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ABSTRACT

Lemierre's syndrome is a condition characterised by septic thrombophlebitis of internal jugular vein following an oropharyngeal infection along with septic embolisation to other organs, mainly lungs. This report is about a 14-year-old girl who presented with complaint of high-grade fever, progressive shortness of breath and painful swelling of throat for seven days. Chest radiograph showed left hemithorax homogeneous opacity with contralateral mediastinal shift. Complete haemogram and other blood investigations revealed neutrophilic leucocytosis and raised C-reactive protein (CRP). Gram stain and aerobic culture of sputum, pleural fluid and blood were inconclusive. As there was no relief of symptoms even after empirical treatment with antibiotics, Contrast Enhanced Computed Tomography (CECT) scan of neck and thorax was done, and it showed bilateral ectatic internal jugular veins with intravascular thrombus and consolidation of left lung with ipsilateral pleural effusion. Ultrasonography (USG) of neck confirmed the presence of thrombus in internal jugular vein on both sides. She was started on intravenous clindamycin and subcutaneous anticoagulants. Gradually the symptoms of the patient resolved. To conclude, despite being called a "forgotten disease", Lemierre's syndrome requires strong clinical suspicion and prompt management to prevent mortality.

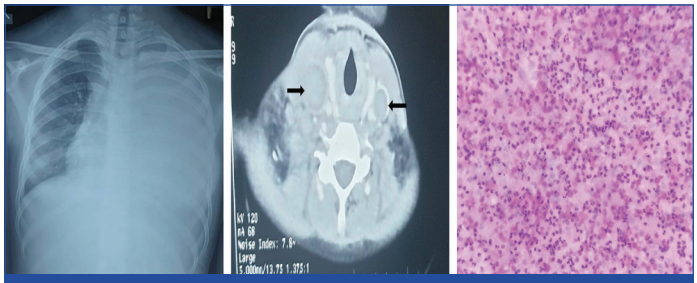
Keywords: *Fusobacterium necrophorum*, Internal jugular vein, Thrombophlebitis, Thrombus

CASE REPORT

A 14-year-old girl was admitted with complaints of high-grade fever, productive cough for seven days, followed by progressive shortness of breath, and painful swelling of face and neck for three days. The girl was treated with oral amoxicillin for pharyngitis with a palpable lymph node in the right side of the neck, in the Outpatient Department (OPD) of General Medicine of the institute. There was no change in voice and no difficulty in swallowing. There was no history of food or drug allergy, but history of recurrent upper respiratory tract infections was present.

On a general examination, there was generalised swelling of the face and neck with raised local temperature and erythema. Her temperature was 38.2°C. Other vital signs: Pulse rate 115 beats/minute, blood pressure 116/74 mmHg, respiratory rate 28 per minute, and oxygen saturation 94% at room air. On examination of the upper respiratory tract, posterior pharyngeal wall was inflamed. On chest examination, chest wall movements diminished on the left side and suprasternal suction and intercostal retraction were seen on the right side of the chest. There was diminished vocal fremitus, stony dull percussion, and absent breath sounds all over the left hemithorax. Examination of other body systems revealed no abnormalities.

Initial blood investigations disclosed neutrophilic leucocytosis and raised C-reactive protein (CRP). Chest radiography showed a left-sided homogeneous opacity with a contralateral mediastinal shift [Table/Fig-1]. Diagnostic thoracentesis and pleural fluid analysis revealed an exudative, lymphocyte predominant fluid with low adenosine deaminase (ADA) level. Gram staining and aerobic culture of sputum, blood, throat swab, and pleural fluid were inconclusive. Sputum and pleural fluid were negative for acid fast bacilli (AFB). The CECT scan of neck and thorax revealed bilateral ectatic internal jugular veins with intravascular thrombus and consolidation of the left lung with ipsilateral pleural effusion [Table/Fig-2]. Further, ultrasonography (USG) of the neck also confirmed the presence of thrombi in the bilateral internal jugular vein. Additionally, cytological examination of ultrasound-guided fine-needle aspirate from the right jugular lymph node was suggestive of acute suppurative lymphadenitis [Table/Fig-3]. From clinical history and investigations, it was diagnosed as a case of septic thrombophlebitis of internal jugular vein or Lemierre's syndrome.



[Table/Fig-1]: Chest radiography showing left sided homogeneous opacity with contralateral mediastinal shift. **[Table/Fig-2]:** Contrast enhanced computed tomographic scan of neck showing bilateral ectatic internal jugular veins (black arrows) with filling defect. **[Table/Fig-3]:** Photomicrograph (fine needle aspiration cytology of right jugular lymph node) showing plenty of neutrophil in a background of necrosis, suggestive of acute suppurative lymphadenitis. (Haematoxylin & Eosin stain: 400X). (Images from left to right)

Considering the possibility of anaerobic sepsis, intravenous clindamycin was initiated. As D-dimer level was 10.93 µg/mL (ref: 0.05-6.5 µg/mL), subcutaneous low molecular weight heparin (1 mg/kg body weight) was started. Therapeutic thoracentesis was done and 1.2 litres of pleural fluid was removed. Gradually, the girl improved clinically. Laboratory parameters also improved after seven days of antimicrobial therapy [Table/Fig-4]. Radiological improvement was observed on chest x-ray and repeated USG of neck which revealed dissolved thrombus in jugular veins. She was discharged with an advice of oral amoxicillin and clavulanic acid combination (625 mg thrice daily) and oral clindamycin (300 mg thrice daily) for six weeks. On follow-up after six weeks, the patient was doing well.

Relevant laboratory parameters	During admission	Post-therapy (day 7)
Total leucocyte count (ref: 4000-11000/cumm.)	18000	6200
Differential neutrophil percentage (%)	85	60
Erythrocyte sedimentation rate (ref: 0-20 mm/hr)	64	15
C-reactive protein (ref: less than 5 g/dL)	83	22

[Table/Fig-4]: Table showing relevant laboratory parameters during admission and post therapy (7 days).

DISCUSSION

Lemierre's syndrome, also called as necrobacillosis or post anginal sepsis is an uncommon, potentially fatal complication of oropharyngeal infection [1]. Courmont first noticed this entity in the early 1900s. Andre Lemierre established a link between anaerobic sepsis and oropharyngeal infection in 1936 by reporting 20 cases, of which 18 patients died [1]. It has been coined as the "forgotten disease" because it is rarely encountered by physicians in routine clinical practice [2].

Fusobacterium species, mainly *Fusobacterium necrophorum* have been implicated in causing this syndrome [3]. However, the cause may be polymicrobial and other microbes such as *Streptococcus, Peptostreptococcus, Bacteroides* may be isolated from some patients [3]. The bacilli have a propensity to affect young adults mostly in the 2nd decade of life [3]. In this case, the age of the girl was 14 years. The syndrome was frequently encountered before the arrival of antibiotics and at present it has an incidence of 3.6 cases per 1 million per year [4]. The infection spreads from several sites in the upper respiratory tract, such as the tonsils, peritonsillar tissue, teeth, and sinuses [3]. The index patient had symptoms such as fever, cough, dyspnoea, and face and neck swelling, which are similar to previous studies [5,6]. Metastatic emboli are typical sequelae of this syndrome and lungs are the most common site of lodgement of septic emboli, reported in 97% of cases, followed by major joints [7]. Since most of the cases present with a metastatic lung infection, a chest x-ray is usual among the first-line investigations [3]. Chest x-ray in the index patient had left-sided homogeneous opacity with ipsilateral pleural effusion, which was similar to the findings of Silva DR et al., [8].

The diagnosis of Lemierre's syndrome depends mainly on three salient features: oropharyngeal sepsis, internal jugular vein thrombophlebitis, and metastatic infection [9]. To establish the diagnosis, the growth of anaerobic bacteria on culture is essential [4]. However, the study by Johannesen and Bodtger U reported that cultures may not ascertain a specific microorganism due to various factors such as prior treatment with antibiotics [10]. So, negative blood cultures may not rule out the possibility of this syndrome, as in the index case where culture reports were inconclusive [10]. To identify the thrombophlebitis of the internal jugular vein and metastatic infection, CECT scan is the investigation of choice, although duplex USG and Magnetic Resonance Imaging (MRI) are also useful [9]. In this patient, a CECT scan of the neck and thorax was employed to detect the bilateral internal jugular vein thrombus and lung consolidation with ipsilateral pleural effusion as a consequence of metastatic infection to the lungs by septic emboli. Ultrasonography was done to confirm the thrombus and its extension.

Prolonged antibiotic treatment is the cornerstone of therapy in Lemierre's syndrome [11]. This patient was treated with intravenous piperacillin-tazobactam initially and after confirming the diagnosis, antibiotic was changed to intravenous clindamycin in order to cover anaerobes [12]. There is ambiguity regarding anticoagulation in the treatment of Lemierre's syndrome. Few published series have mentioned that 21-30% of patients received anticoagulation [3,12]. In lieu of clinical condition of the index patient, anticoagulation with low molecular weight heparin was undertaken. If thrombus doesn't dissolve despite pertinent medical management, ligation or excision of the internal jugular vein, a procedure often performed in the preantibiotic era, could be considered [12].

CONCLUSION(S)

Considerable clinical suspicion of Lemierre's syndrome is necessary in patients of internal jugular vein thrombophlebitis with the background of an oropharyngeal infection. Prompt and belligerent management is necessary to avert morbidity and mortality.

REFERENCES

[1] Baig MA, Rasheed J, Subkowitz D, Vieira J. A review of Lemierre syndrome. Internet J Infect Dis. 2005;5(2):4457.
[2] Chamseddin KH, Kirkwood ML. Lemierre's syndrome associated mycotic aneurysm of the external carotid artery with primary internal carotid artery occlusion in a previously healthy 18-year-old female. Ann Vasc Surg. 2016;36:291-e11.
[3] Karkos PD, Asrani S, Karkos CD, Leong SC, Theochari EG, Alexopoulou TD, et al. Lemierre's syndrome: A systematic review. The Laryngoscope. 2009;119(8):1552-59.
[4] Hagelskjaer Kristensen L, Prag J. Lemierre's syndrome and other disseminated *Fusobacterium necrophorum* infections in Denmark: A prospective epidemiological and clinical survey. Eur J Clin Microbiol Infect Dis. 2008;27(9):779-89.
[5] Hagelskjaer LH, Prag J, Malczynski J, Kristensen JH. Incidence and clinical epidemiology of necrobacillosis, including Lemierre's syndrome, in Denmark 1990-1995. Eur J Clin Microbiol Infect Dis. 1998;17(8):561-65.
[6] Chirinos JA, Lichtstein DM, Garcia J, Tamariz LJ. The evolution of Lemierre syndrome: Report of 2 cases and review of the literature. Medicine. 2002;81(6):458-65.
[7] Ramirez S, Hild TG, Rudolph CN, Sty JR, Kehl SC, Havens P, et al. Increased diagnosis of Lemierre syndrome and other *Fusobacterium necrophorum* infections at a Children's Hospital. Pediatrics. 2003;112(5):e380.
[8] Silva DR, Gazzana MB, Albaneze R, Dalcin PD, Vidart J, Gulcó N. Septic pulmonary embolism secondary to jugular thrombophlebitis: A case of Lemierre's syndrome. J Bras Pneumol. 2008;34:1079-83.
[9] Sinave CP, Hardy GJ, Fardy PW. The Lemierre syndrome: Suppurative thrombophlebitis of the internal jugular vein secondary to oropharyngeal infection. Medicine. 1989;68(2):85-94.
[10] Johannesen KM, Bodtger U. Lemierre's syndrome: Current perspectives on diagnosis and management. Infect Drug Resist. 2016;9:221.
[11] Eilbert W, Singla N. Lemierre's syndrome. Int J Emerg Med. 2013;6(1):01-05.
[12] Riordan T, Wilson M. Lemierre's syndrome: More than a historical curiosa. Postgrad Med J. 2004;80(944):328-34.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Mar 07, 2022
- Manual Googling: Apr 13, 2022
- iThenticate Software: May 30, 2022 (18%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Mar 01, 2022**
Date of Peer Review: **Mar 29, 2022**
Date of Acceptance: **Apr 14, 2022**
Date of Publishing: **Jun 01, 2022**