Lemierre Syndrome: A Case of Postanginal Sepsis

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Lemierre syndrome is a rare disease that’s characterized by internal jugular vein thrombosis and septic emboli. These symptoms typically develop after acute oropharyngeal infection by Fusobacterium necrophorum. Although this syndrome is less frequently seen in modern times due to the availability of antibiotics, physicians must be aware of the syndrome in order to initiate prompt antibiotics therapy, including coverage of the anaerobic organisms. We discuss here the case of an 18-year-old female with Lemierre syndrome and we review the relevant literature on this syndrome.

Key Words : Thrombophlebitis, Fever, Sepsis

INTRODUCTION

In 1936, Lemierre described a syndrome with the characteristic elements of oropharyngeal infection, sepsis, internal jugular vein thrombosis and septic emboli, and this was all caused by Fusobacterium necrophorum. Even though this syndrome is observed less frequently in the present day, the mortality rate of Lemierre syndrome is still estimated to be 8~15%, despite of administering modern antibiotic therapy. Mortality is usually caused by septic metastasis to distant organs such as lung, as well as to the knee and shoulder joints. In this report, we describe a case of Lemierre syndrome with pharyngo-tonsilitis, and this resulted in thrombophlebitis of the internal jugular vein and a metastatic septic embolism in the lungs.

CASE REPORT

An 18-year-old female was admitted to the emergency room with a persistent, week-long fever, a sore throat, rigor and myalgia. She had been previously treated at a local clinic for three days, but she was referred to our hospital because the treatment at the clinic did not significantly improve her condition. Upon admission to our hospital, she was febrile and tachycardic. On the physical examination, her body temperature was 39.0°C and her throat was slightly inflamed. Serologic tests were performed, and the results revealed a white blood count (WBC) of 13,600/cm³, with a left shift and mild thrombocytopenia (94×10³/L). Both the blood urea nitrogen and creatinine levels were within normal limits. The lipid profiles were within normal limits, but the liver function enzymes were mildly elevated. In addition, the level of C-reactive protein (CRP) was abnormally elevated. A plain chest film showed multifocal nodular infiltration and a right pleural effusion. Based on the radiological view, we first suspected that the diagnosis was either pneumonia or pulmonary tuberculosis. Upon being admitted to the hospital, cefmetazole (2 gram daily) and amikacin (500 mg daily) were administered intravenously. The patient complained of a severe headache and neck stiffness. Therefore, we performed a lumbar puncture, but we could not find any evidence of meningitis. On day two of her hospitalization, the patient complained of swelling...
Figure 1. (A) Chest PA revealed multifocal nodular infiltrates that are mainly in the right lung field and there is the suspicion of this in the left, mid lung field. (B) Follow up chest PA after 4 days showed more aggravated and discrete nodular lesions in the right and left lower lung fields.

Figure 2. Neck ultrasonogram showed inner echoic thrombosis in the right internal jugular vein.

Figure 3. Chest computerized tomography revealed pleural based multiple cavitary nodules with combined feeding vessels, and this is compatible with pulmonary septic emboli.

and pain on the right side of her neck. Upon physical examination, the right external jugular vein has become a palpable strand with severe tenderness. A neck sonogram showed that the lower portion of the right internal jugular vein was severely dilated, with inner non-compressible thrombi and without vascular flow through the right internal jugular vein. On day five day of her hospitalization, the patient had persistent fever and worsening pain on the right side of the neck. Furthermore, she became dyspneic and developed pleuritic chest pain. Chest computerized tomography (CT) revealed multiple subpleural-based lung nodules with some cavities in both lungs, and this was combined with perinodular, linear and patchy infiltrations. The CT also showed a 10cm segment of thrombosis in the right internal jugular vein. Plain chest film showed a more aggravated nodular infiltration in the upper and lower right lung fields. Therefore, we modified the patient’s antibiotic treatment to Tazocin (piperacillin + tazobactam, 13.5 gram daily). A throat swab, trans-echocardiogram, abdominal ultrasonoram and several sets of blood and sputum cultures were taken, but we did not find any focus of infection. After changing the antibiotics, the patient became afebrile and the pleuritic chest pain was alleviated. The patient was discharged...
Figure 4. Computerized tomography of the neck showed right internal jugular vein thrombosis (arrow) and right para-pharyngeal venous thrombosis.

from the hospital without complications on the 14th day after admission.

DISCUSSION

Despite being an uncommon disease, it is important to accurately diagnose Lemierre syndrome because of its high mortality rate. Lemierre syndrome is characterized by distinct clinical features. The initial cause of the syndrome is a tonsillar or peritonsillar abscess, followed by thrombophlebitis of the tonsillar and peritonsillar veins, which can then spread to the internal jugular vein: this in turn leads to septicemia and the formation of distant metastatic abscesses. These secondary metastatic abscesses are most frequently localized in the lungs, and 95% of patients with Lemierre syndrome have demonstrated pleural-pulmonary involvement\(^2, 10\). Lemierre syndrome is usually caused by \textit{Fusobacterium necrophorum}, an anaerobic, nonpathogenic gram-negative rod that is present in the normal mucosal flora, and this organism is sensitive to penicillin, clindamycin, metronidazole and chloramphenicol\(^6\). A study by Alston et al. reported a series of 280 episodes; these typically occurred in young, otherwise healthy patients\(^7\). In that study, the most frequent site of primary infection was the oropharynx and the severity of infection ranged from mild to fulminant. The extrapharyngeal primary sites of infection include the middle ear, the female urogenital tract and the gastrointestinal tract. Making the correct diagnosis is essential for selecting an appropriate treatment, and especially for treating pharyngitis, otitis media or thromboembolic disease. In our case, we presumed that the patient had a peritonsilar abscess that had spread to create ipsilateral internal jugular vein thrombosis and this was complicated by septic emboli to the lungs. Although \textit{F. necrophorum} is the most common Lemierre syndrome pathogen, other causative organisms have been isolated in this syndrome, including the \textit{Streptococcus species}, \textit{Bacteroides species} and \textit{Peptostreptococcus species}, as well as \textit{Eikenella corrodens}\(^8, 9\). No pathognomic organism was isolated in our case, and this was probably due to the patient’s prior treatment with antibiotics. When a diagnosis of Lemierre syndrome is suspected, antibiotics designed to target anaerobic bacteria should be included in the initial therapy. The definitive antibiotic regimen is dependent upon the results of susceptibility testing. Although no pathogen was cultured in our case, broad spectrum antibiotic therapy with potent activity against anaerobic bacteria is necessary for managing the disease. Examples may include either clindamycin or a combination of a beta-lactam and metronidazole. The preferred duration of antibiotic therapy is 2-6 weeks, with conversion from intravenous to oral administration after there is marked improvement, as evidenced by the resolution of leukocytosis and the decreasing inflammatory indices. We first chose to treat our patient with cefazolin and amikacin because of our experience with these antibiotics. However, in this case, we had to change antibiotics because of the patient’s febrile condition. The patient’s condition rapidly improved after replacing the antibiotics with Tazocin.

Furthermore, the role of anticoagulation for treating this condition remains unclear since no controlled, randomized trials have been performed to test the hypothesis of whether or not this treatment is beneficial. More intensive therapies such as internal jugular vein ligation or surgical drainage of abscess are occasionally necessary\(^10\). The critical radiologic features of Lemierre syndrome are the presence of septic pulmonary emboli with the appearance of multiple peripheral, round and wedge-shaped opacities that rapidly progress to cavitation\(^11\). In our case, an overt feeding vessel leading into some of the nodules was noted, and this is compatible with pulmonary septic emboli. Ultrasonography is a noninvasive, readily available imaging tool that can demonstrate thrombi\(^12\). In our case, the ultrasound scan showed inner hyperechoic thrombosis in the right internal jugular vein, and computed tomography of the neck showed both right internal jugular venous thrombosis and a right parapharyngeal venous thrombosis.

Lemierre syndrome is an uncommon disease. However, pharyngitis or tonsillitis can progress to this rare, but lethal syndrome. Therefore, early diagnosis and the prompt
administration of broad spectrum antibiotics treatment are essential for preventing metastatic dissemination of septic embolus and to reduction of the mortality and morbidity of this disease.

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