An Unusual Presentation of Lemierre Syndrome

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Abstract

Lemierre syndrome is an entity defined by septic thrombophlebitis of the internal jugular vein following an oro-pharyngeal infection, which is usually acute and complicated by metastatic infection in different organs. The usual causative organism is \textit{Fusobacterium necrophorum}. On looking back at the case reports of Lemierre syndrome, we have found different sites of primary infection and also different presentations depending on the primary site and the site of involvement resulting from metastatic septic embolization. However, chronic otitis media as the primary site of infection and bloody diarrhea as the presenting symptom were very rarely presented. The case presented here was referred to Faghihi hospital of our academic medical center with bloody diarrhea. After work ups, the patient was diagnosed as a case of Lemierre syndrome on the base of chronic otitis media.

Keywords: Lemierre syndrome; Chronic otitis media; Internal jugular vein; Thrombosis; Septic emboli

Introduction

In 1936, Dr. Andre Lemierre described a syndrome characterized by anaerobic septicaemia, internal jugular vein (IJV) thrombosis, and septic emboli that arose secondary to infections of the head and neck. However he was not the first to describe this syndrome, Courmant and Cade and Schottmuller described the same syndrome before him in 1900 and 1918 respectively.\textsuperscript{1,2} The usual etiologic agent in Lemierre syndrome is \textit{Fusobacterium necrophorum}, an anaerobic, nonmotile, nonspore-forming, gram negative rod.\textsuperscript{3-5} The first stage is the primary infection, which is usually pharyngitis. It has been sporadically reported to complicate acute otitis media and mastoiditis\textsuperscript{6} or an infected cholesteatoma.\textsuperscript{7} The lung is the most common site of metastatic infection, but additional metastatic abscesses are often found elsewhere in the body.\textsuperscript{8,9}

Nowadays, Lemierre syndrome is a rare disease because of the advent of antibiotic therapy and it is often referred to as the “forgotten disease”.\textsuperscript{4,8} However, its serious consequences justify a prompt diagnosis of the syndrome and serious considerations regarding its management.

Case Report

A 19-year-old boy referred to the emergency department with nausea, vomiting, abdominal cramp and bloody diarrhea since the two previous days. He was febrile and toxic, and physical exam revealed tympanic abdomen with moderate generalized tenderness. He denied any significant medical problem in the past. The patient was admitted and intravenous antibiotic therapy with metronidazol and ceftriaxone was started. Laboratory data at that time revealed leukocytosis (WBC: 31,900/\mu l), thrombocytosis (Plt: 562,000/\mu l) and anemia (Hb: 10.5g/dl). Stool analysis showed many RBCs but no pus cells. In abdominal X-ray, dilated small bowel loops were demonstrated. On the second day of admission, he was already febrile and crepitation was noted in the abdominal wall in physical exam. Abdominal X-ray was repeated, and surgical consultation was done, but no evidence of hallow viscous perforation was documented in the X-ray or noted by the surgeon. Only dilated fluid filled the small bowel loops which was demonstrated in abdominal sonography. Sedimentation rate in the patient was reported to be more than 140 mm/hr. The

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next day, he felt better but complained of pleuretic chest pain and trespus. In the physical exam, a tender swelling in the right side of the neck, decreased breathing sounds, and crackles in the base of right lung were noted. ENT exam revealed perforated right tympanic membrane with profound pussy discharge. Chest X-ray demonstrated prominent right pulmonary conus and infiltration in the peribronchial area in the right lower lung field. CT scan of the neck with IV contrast was done, showing thrombosis of the right internal jugular vein in the total course with surrounding soft tissue edema (Figure 1). At that time, the patient mentioned a history of trauma to the right ear and pussy discharge from ear canal thereafter for several months. Anticoagulation with heparin was started and antibiotic therapy continued. High Resolution CT scan of the temporal bone without contrast was done which showed loss of aeration in the right mastoid air cells and soft tissue density in the attic ridge of the right middle ear in favor of cholesteatoma. In the spiral CT scan of the chest, multiple cavitary lesions suggestive of abscess formation in the right lung and right pleural effusion were reported (Figure 2). The pleural fluid culture grew *Staphylococcus aureus* sensitive to vancomycin and cephalothin, so vancomycin was added to antibiotic regimen. No cerebr al venous sinuses involvement was demonstrated in the brain MRI. The patient’s condition improved clinically and paraclinically. A mild increase in the liver transaminases was detected during the hospital course which was improved in the next serial checking. Anticoagulation was continued with warfarin and he was discharged after 4 weeks with good general condition. The plan was to continue PO antibiotic therapy for the next 2 weeks and surgery for chronic otitis media after completion of the antibiotic course. Then, mastoidectomy and drainage of cholesteatoma was done at the planned time with no complication.

This was a case of Lemierre syndrome regarding the focus of infection in the ear complicated by IJV thrombosis and septic embolization. There was no positive culture for *F. necrophorum*, and the reason was that blood culture was not obtained for anaerobic organisms before antibiotic treatment. The patient had already responded to empirical antibiotic regimen and was in acceptable good general condition when the only positive pleural fluid culture was discovered. Therefore, *Staphylococcus aureus* in this case was probably responsible for the secondary infection. The cause of his unusual presentation of abdominal cramp and bloody diarrhea was most probably septic embolization to the mesenteric vasculature. This is logically concluded from physical examination points of abdominal distension, crepitation and tenderness regarding paraclinical findings of the small bowel dilatation and no fecal leukocytosis in a patient with bloody diarrhea and a potent source of septic embolization.

**Discussion**

Studies of the incidence rates for Lemierre syndrome have shown figures between 0.6 and 2.3 per million populations and more commonly in males. The syndrome is most often caused by *Fusobacterium necrophorum*; however, several other organisms were reported, alone or in combination with *F. necrophorum*. The primary infection is usually pharyngitis in more than 80% of cases, but also it might be mastoiditis, or odontogenic infections. In classic cases of LS, an episode of pharyngotonsillitis, bacterial or viral, impairs the oropharyngeal mucosal defense system. This allows *F. necrophorum* to invade the pharyngeal space and then IJV. Once thrombophlebitis of the IJV occurs, septic emboli can arise and spread to distant sites and organs.
The clinical presentation in the primary stage depends on the primary site of the infection. In a vast majority of patients, sore throat and evidence of pharyngeal inflammation are the primary findings. Fever occurs in most but not all cases. Gastrointestinal complaints such as abdominal pain, nausea and vomiting have been reported to be present in about half of the cases.

Once invasion to the parapharyngeal space and IJV occurs, symptoms may be related to the local involvement of this area or metastatic complications. The clinical findings of the invasion of this compartment result from the compromise of the vital structures in the parapharyngeal space. Ruptures of the carotid artery, Horner syndrome, and paralysis of the trapezius muscle have been reported. The most common finding at this stage is a tender, swollen neck but dysphagia and tremeus may also be present.

After involvement of IJV, complications of bacteremia and metastatic infection may be evident. Lungs as the first pass filters are the most common sites of involvement in Lemierre syndrome. Septic infarcts almost invariably lead to multiple abscess formation, which is announced by intense thoracic pain, dyspnea, and sometimes, hemoptysis. Chest radiograph typically shows scattered nodular infiltrates and small pleural effusions. Early cavitation is common and might sometimes be seen in the first chest radiograph. Empyema, pneumatoceles, and pneumothorax have also been described as well as respiratory failure and pulmonary embolism.

Metastatic abscess formation can also occur in the large joints, bone, muscle, soft tissues, brain, meninges, liver, spleen, kidney, pericardium, and endocardium. The criteria mentioned by Sinave et al. for “Classical Lemierre’s syndrome” are as follows: (i) Primary infection in the oropharynx, (ii) Septicaemia documented by at least one positive blood culture, (iii) Clinical or radiographic evidence of IJV thrombosis and (iv) At least one metastatic focus. However, nowadays because of the early use of antibiotics, blood cultures may be more negative.

Radiographic evaluation of the neck to confirm IJV thrombosis can be accomplished by ultrasonography, contrast CT scan, or MRI. CT scan is more sensitive than ultrasonography but involves radiation exposure and administration of intravenous contrast. MRI is very accurate in the diagnosis with excellent soft tissue delineation, and lack of radiation exposure. Radiologic evaluation is of value in assessing metastatic complications.

F. necrophorum has been traditionally considered to be susceptible to penicillin, clindamycin, and metronidazole. Resistance to penicillin and erythromycin has been reported but not to clindamycin and metronidazole. Treatment with anaerobic monotherapy alone is not advised. Even though the initial infecting organism is usually F. necrophorum, there is evidence that secondary infection and pus collection can occur with organisms like Staphylococcus aureus. There are no recommended figures for the duration of antibiotic therapy but treatment is usually given for a period of 3 to 6 weeks.

Ligation of the IJV was the treatment of choice in preantibiotic era, and is now probably only indicated in patients with persistent septic embolization despite antibiotics.

The use of anticoagulative drugs remains controversial. Although some authors recommend its use as an adjunct to antibiotic therapy, regarding the potential for more rapid resolution of thrombophlebitis and bacteraemia, others have questioned the benefit of anticoagulants, regarding the risks of hemorrhage and extension of infection.

Lemierre syndrome has a wide spectrum of different clinical presentations. Although rare, it has significant morbidity if it remains untreated, so early diagnosis and prompt treatment is the most important factors in predicting the patient’s prognosis. Diagnosis is not always straightforward because of variable clinical features and difficulty in obtaining microbiologic evidence. Therefore, a high index of suspicion is needed for correct diagnosis and management.

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Conflict of interest: None declared.

References

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