The Forgotten Disease: A Case of Lemierre's Syndrome

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Abstract

Lemierre's syndrome (LS) is a rare but life-threatening condition that presents as an oropharyngeal infection. There is no screening test for it, and its diagnosis is clinical. We report the case of a previously healthy young man who presented with LS. His diagnosis was made with bedside ultrasound; hence, we recommend it as a screening tool in the emergency department. His delayed treatment resulted in his demise. In a developed country with easy access to antibiotics, we need to urgently raise physician's awareness toward LS and highlight the importance of early diagnosis and antibiotic use for its patients.

Keywords: Lemierre syndrome, postanginal sepsis, thrombophlebitis

Introduction

Lemierre's syndrome (LS) is a rare but life-threatening condition commonly caused by Fusobacterium necrophorum. Patients present with a more severe sore throat than that in streptococcus infection. There is no screening test for Fusobacterium, which makes its diagnosis challenging. Its diagnosis is largely clinical, and if recognized and treated early, the prognosis is good. LS is characterized by an oropharyngeal infection leading to thrombophlebitis of the internal jugular vein with secondary metastatic abscesses to the lungs and joints. If treatment is delayed, mortality can be as high as 25% compared with 0%-18%, if treated early (1).

We report the case of a young immunocompetent adult who presented with LS, but whose treatment was delayed, resulting in dire consequences. In a developed country with easy access to antibiotics, we need to urgently raise primary care physicians' awareness toward LS.

Case Presentation

A 36-year-old immunocompetent man, who is a front-desk officer at a hotel, sought treatment at his general practitioner's (GP) clinic with a four-day history of fever, right-sided throat pain, and loss of appetite. He was diagnosed as having tonsillitis, but no antibiotic was administered. Three days later, he returned to the GP's clinic with complaints of persisting fever, sore throat, cough, orthopnea, and vomiting. The patient was also hypotensive (blood pressure, 80/60 mmHg). The patient was then referred to the emergency department.

In the emergency department, his blood pressure was 95/69 mmHg after fluid boluses, respiratory rate was 24 per minute, heart rate was 120 beats per minute, temperature was 38.4°C, and oxygen saturation in room air was 93%. He was very lethargic, tachypneic, and dehydrated. Physical examination revealed erythematous tonsils, mild right neck swelling, and crepitations over the lung bases.

His blood test showed leukocytosis, thrombocytopenia, hyperlactatemia, coagulopathy, and raised C-reactive protein and creatinine levels. Chest X-ray revealed prominent pulmonary vessels and increased shadowing in both lungs. Neck X-ray was unremarkable. The patient was then treated for severe community-acquired pneumonia with type 1 respiratory failure.

He was admitted to the intensive care unit (ICU) for inotropes and was started on co-amoxiclav, ceftazidime, and azithromycin. He remained hypotensive and hypoxemic, eventually requiring intubation and mechanical ventilatory support.

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Bedside ultrasound of the right internal jugular vein (IJV) showed a thrombus. Despite his thrombocytopenia, the patient was treated with enoxaparin. Clindamycin and metronidazole were also initiated.

Lemierre’s syndrome was diagnosed based on the presence of septic emboli on chest X-ray and right IJV thrombus on the Doppler scan. Computed tomography (CT) of the neck and chest confirmed the presence of thrombus in the right IJV. There was abscess in the right tonsillar region with extension to the right parapharyngeal region. Extensive areas of consolidation with septic emboli were found in both lungs. Serology test for HIV was negative. Sputum cultures for tuberculosis tested negative. Subsequent blood culture results were found positive for Dialister pneumosintes. The patient’s health condition progressively deteriorated despite aggressive medical management, and he succumbed after 2 weeks of ICU stay.

Telephonic verbal consent by the patient’s mother was obtained to publish this case report. All details that would enable any reader (including the individual or anyone else) to identify the person are omitted.

Discussion

Lemierre’s syndrome is also known as postanginal septicaemia. It usually presents as an acute oropharyngeal infection. It causes thrombophlebitis of the IJV that may result in bacteremia, with hematogenous spread to other sites such as the lungs and joints. The infection spreads via the peritonsillar tissue to the adjacent pharyngeal space (1). Our patient’s presentation of tonsillitis and his primary physician’s failure to diagnose LS led to a delay in his referral. Although the diagnosis of LS is difficult, it should be a clinical one. Dr Andre Lemierre, the microbiologist who first described this disease, stated that the classical findings were so characteristic that it was possible to make a diagnosis without the results of any type of bacterial testing (2). There should be a high index of suspicion when patients present with respiratory symptoms, neck swelling, or signs of toxicity that occur within a week after an oropharyngeal infection (3). A scoring criterion such as the Centor criterion should be utilized to assess the likelihood of a bacterial infection and assist in the decision on whether antibiotics should be started in patients complaining of a sore throat (4).

Treatment for LS should be started based on suspicion, before positive blood culture results, which would otherwise require an incubation period of 6-8 days. Clinical suspicion should be followed up with CT scans of the neck and lungs to identify thrombus in the jugular veins and cavitating lung lesions.

Although the most common causative microorganism implicated in LS is F. necrophorum, the etiology of LS can also be polymicrobial (1). In our patient, the organism implicated was D. pneumosintes, otherwise known as Bacteroides pneumosintes. To the best of our knowledge, this is the first reported case of D. pneumosintes as the causative organism of LS. This gram-negative anaerobe is a periodontal pathogen implicated in periodontitis, bacteriaemia, and abscesses (5). Our patient was otherwise healthy with no known risk factors such as poor dental hygiene, rendering his diagnosis bizarre.

Our patient’s thrombus on the IJV was realized by ultrasound while attempting to insert a central venous catheter. As such, the authors feel that bedside Doppler ultrasound should be performed as an initial point of care investigation in patients with sepsis presenting to the emergency department with sore throat and neck pain. The diagnosis should then be confirmed by high-resolution CT of the thorax and neck (6).

Treatment for jugular thrombophlebitis and LS has not been yet standardized, but high doses of penicillin, metronidazole, clindamycin, and chloramphenicol are recommended (6). Our patient was treated with co-amoxiclav, metronidazole, clindamycin, azithromycin, and ceftazidime, although his blood culture later showed resistance to metronidazole and penicillin. Although controversial, other treatment modalities include anticoagulation and ligation and resection of thrombosed veins (1,7).

Conclusion

Lemierre’s syndrome is a rare but life-threatening condition that presents with a common sore throat. To achieve a good outcome, it requires early recognition and treatment. Bedside ultrasound can be utilized as a screening tool. Despite cutting-edge investigations and treatments, once the diagnosis is delayed, as in the case of our patient, death is the outcome. Aggressive and extended duration of intravenous antimicrobial therapy remains the predominant appropriate therapy.

Informed Consent: Verbal informed consent to report the case was obtained from patients’ parents as patient had demised.

Conflict of Interest: The authors have no conflict of interest to declare.

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References

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