Lemierre Syndrome, The Forgotten Disease

Abstract:

Lemierre syndrome is a rare and potentially fatal entity affecting otherwise healthy and young adults. The infection originates in the throat and spreads via a septic thrombophlebitis of the internal jugular vein, with development of distant septic emboli. This clinical picture is characteristic, but many clinicians are unaware of the condition and diagnosis is often delayed with potentially fatal consequences.

Introduction

Lemierre's syndrome (or postanginal septicemia, or suppurative thrombophlebitis) is a rare but severe life-threatening complication of acute tonsillitis. It is characterized as thrombophlebitis of the internal jugular vein that is associated with spread of septic emboli to the lungs and other sites. Before the availability of antimicrobial agents, death was the common result (90%), unless patients were treated with surgical ligation of the vein. The incidence of Lemierre syndrome is about one per million per year. It appears to have been relatively common in the preantibiotic era, in 1955 Alston identified 280 cases in the world literature. It was rarely reported in the 1960s and 1970s possibly because of widespread use of penicillin for throat infection. Several papers in the 1980s highlighted that this "forgotten disease" had not gone away. There has been an increase in reporting of Lemierre syndrome over the last 10 years due to increased antibiotic resistance or changes in antibiotic prescription patterns.

Case Report

A 28-year-old male, with no previous medical history, presented in the emergency room with a 3-day history of left neck swelling, pain, fever, odynophagia, dysphagia and 2-week history of tonsillitis and general weakness. He was treated with clarithromycin orally for 1 week by his GP. His temperature was 38.3°C, regular pulse of 98/minute, respiratory rate of 19/minute and blood pressure of 124/75 mmHg. Oral examination demonstrated erythema and mild swelling of the tonsils without exudates, left latero-cervical region was tender to palpation, with a mass of 4 cm diameter at level 2, firm, immobile and non fluctuant. Flexible nasofibroscopy was normal. The lungs were clear to auscultation. Laboratory data demonstrated WBC=12, CRP=70. A CT scan of the neck revealed a hypodense filling defect in the left internal jugular vein, with extensive surrounding inflammatory changes. Blood culture demonstrated Streptococcus intermedius. The patient was treated with a combination of cefuroxime, metronidazole and fractionated heparin for 24 hours, after that fractionated heparin for 6 weeks. The patient improved significantly, with complete resolution of symptoms. At follow-up, the patient was completely asymptomatic and appeared to have recovered without any residual effects.

Discussion

Lemierre syndrome mainly affects young healthy patients. It is an anaerobic suppurative thrombophlebitis involving the internal jugular vein, usually as a complication of pharyngeal, dental or mastoid infection. Fusobacterium necrophorum is the causative agent in 70% of cases. Other, Fusobacterium species were responsible for infection in 10% and other gram-negative organisms were responsible in 8%. 12% of cases grew negative cultures. The initial symptoms are usually nonspecific and include sore throat, fever, rigor and lateral neck tenderness. The disease usually begins as pharyngitis or tonsillitis. Spread of the infection to the deep pharyngeal tissue allows anaerobic organisms to drain into the lateral pharyngeal space, leading to internal jugular vein thrombophlebitis. Septic clots dislodge from internal jugular vein thrombus, causing pulmonary infarcts. Hæmagotóasis seeding can also occur, resulting in septic arthritis, meningitis, endocarditis or soft tissue infection. Diagnosis is made with positive blood culture combined with appropriate imaging findings, including retrograde venography (the gold standard), gallium scan, ultrasonography, computed tomography and magnetic resonance venography. The therapy of Lemierre Syndrome is a 4-5 weeks course of intravenous antibiotic with activity against Fusobacterium necrophorum, such as penicillin G, clindamycin or metronidazole. The role of anticoagulant therapy is controversial, but several clinical studies have shown that heparin may be beneficial, especially for thrombosis retrograde to the cavernous sinus. The mortality rate in untreated patients is as high as 30 - 90% with an embolic event rate of 25% and endocarditis rate of 12.5%.

As in this patient, an early clinical diagnosis of Lemierre's syndrome was made and confirmed by imaging studies. Our patient presented and was treated before septic metastases spread. This early recognition and intervention were probably the key contributing factors to our patients good outcome and survival.

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References


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