The “Atypical Sore Throat” – a Pediatric Case of Lemierre’s Syndrome

INTRODUCTION

Suppurative jugular thrombophlebitis otherwise known as Lemierre’s syndrome is a life-threatening diagnosis. Previously referred to as “the forgotten disease”, the syndrome manifests with such common symptoms as fever, sore throat, and lymphadenopathy. For this reason, it is often misdiagnosed.

Lemierre’s syndrome was first characterized by Dr. Andre Lemierre in 1936 with his report of 20 cases published in the Lancet. The disease typically begins with an oropharyngeal infection and subsequently develops into septic thrombophlebitis within the internal jugular vein (IJV), with seeding to distant organs (1). While pharyngitis is commonly the first sign of infection, other primary sources have been documented including odontogenic infection, mastoiditis, otitis media, sinusitis, and parotitis (2). While the pathophysiology remains somewhat obscure, this infection is believed to cause microbial seeding of surrounding tissues via hematogenous, lymphatic, or even contiguous spread from a local abscess, resulting in thrombophlebitis of the IJV within 1-3 weeks (2). The most commonly isolated pathogen is Fusobacterium necrophorum, which has been found in as many as 81.7% of cases (3). Cases involving other pathogens such as streptococcus, bacteroides, eikenella, or MRSA as well as sterile cultures, have also been documented (2).

Without appropriate antibiotic therapy, Lemierre’s syndrome can rapidly progress to sepsis and mortality. Other complications arise from the seeding of distant organs such as the lungs, liver, bones and joints, kidneys, cardiovascular or central nervous system (3-4). If diagnosed and treated early, Lemierre’s syndrome is amenable to treatment. The issue is early diagnosis, which is made complicated by the diversity of clinical presentation and relative rarity of the disease (5-7). Lemierre’s is often diagnosed when blood cultures reveal anaerobic Gram-negative rods. This leads to investigation for evidence of septic thrombophlebitis (2). Despite an increasing number of case reports in recent years, Lemierre’s syndrome remains a mysterious disease. It is not commonly thought of when patients present, and as such, diagnosis is frequently delayed.

This report presents a case of Lemierre’s syndrome in a 7-year-old female that was diagnosed after a full course of antibiotic therapy for otitis media and query mastoiditis.

CASE REPORT:

First Presentation:

A 7 year-old previously healthy female presented with a two-week history of low-grade fever in late March 2014. She was seen by her family physician and diagnosed with right-sided otitis media. She was treated with oral amoxicillin that was discontinued after 3 days due to vomiting.

Second Presentation:

She subsequently presented to the Emergency Department (ED) on April 5th, this time with right-sided neck pain suspicious for meningismus. Again, she had a fever of 37.4°C, rising to 38.4°C. A full septic work up revealed a white blood cell count (WBC) of 16.1, hemoglobin (Hb) 124, and platelets 383. Lumbar puncture was negative. With ongoing neck pain and suspected mastoiditis, treatment with IV ceftriaxone and vancomycin was started. She was discharged home on oral cefprozil, and completed a 10-day course. Her blood culture was positive for Streptococcus intermedius after 81 hours of incubation, which was considered likely to be a contaminant. Although the diagnosis of mastoiditis was considered, an ENT consult was not obtained.

Third Presentation:

She improved with the 10-day course of antibiotics and returned to school. On April 18th, she again developed fever, vomiting, and bilateral calf and neck pain. Her vital signs were: Temp 38.7, HR 144, RR 32, O2 92%. She had a small right-sided cervical lymph node. Complete blood count showed WBC of 24 with a left shift, neutrophils of 21, bands 1.7 and platelets of 437. A nasopharyngeal swab was negative for RSV, Influenza A and B, and monospot was negative. Repeat blood cultures were positive for Streptococcus viridans after 51 hours incubation. The patient was again discharged as cell counts normalized and her pain and subjective signs had improved.

This report presents a case of Lemierre’s syndrome in a 7-year-old female that was diagnosed after a full course of antibiotic therapy for otitis media and query mastoiditis.
CONCLUSION

Several case reports published over the past decade emphasize the importance of maintaining a high clinical suspicion for Lemierre's Syndrome (5-7). This is noted in particular when patients present with pharyngitis and several days of fever. Our case occurred in a pediatric patient in whom sore throat was not a feature and blood cultures did not isolate *Fusobacterium*. As such it does not follow the more typical features of this syndrome, and clinical suspicion was not raised until several weeks after symptoms began.

Recent publications have suggested that the prevalence of Lemierre's syndrome has increased due to the restricted use of antibiotics for pharyngitis (4). In our case, it is possible that a full course of antibiotics administered early prevented more serious and systemic disease. Further research is needed to better elucidate the pathogenesis of Lemierre’s syndrome and the mechanism(s) of infectious spread. This may help us to better understand this mysterious and “forgotten” disease and provide clinicians with more concrete signs to raise clinical suspicion and support early diagnosis.

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References: