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Case Report

Atypical Lemierre's Syndrome Caused by *Proteus mirabilis*: A Case Report

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Abstract

Lemierre's syndrome is defined by septic thrombophlebitis of the internal jugular vein following pharyngitis. It is commonly preceded by oropharyngeal bacterial infections. It is rare and typically affects previously healthy adolescents and young adults. Pathogens associated with Lemierre's syndrome include organisms of the normal oropharyngeal flora. The reported most common causative organism is the anaerobic *Fusobacterium necrophorum*. In this case report, we present a 3-year-old boy with an atypical presentation of Lemierre's syndrome complicating mastoiditis caused by *Proteus mirabilis.*

Keywords: *Lemierre Syndrome; Atypical Lemierre Syndrome; Lemierre Disease; Lemierre; Mastoiditis; Thrombosis; Proteus spp; Case Report*

Introduction

Lemierre's Syndrome is named after Dr. André Lemierre who described 20 cases of septicemia following tonsillar infections in 1936. Lemierre's Syndrome is described by septic thrombophlebitis of the internal jugular vein preceded by acute bacterial oro-pharyngitis, otitis media, mastoiditis, pharyngeal abscesses, infectious mononucleosis, or dental infections. The common causative organism associated with Lemierre's syndrome is *Fusobacterium necrophorum*. However, other organisms have also been described in the literature. Other implicated organisms include *Enterobacteriaceae*, *Eikenella corrodens*, *Porphyromonas asaccharolytica*, *Bacteroides*, and *streptococci*. The manifestations of Lemierre's Syndrome include septic embolization to the lungs, commonly, or to other organs e.g. joints, bones, muscles, spleen, liver, or central nervous system [1]. The features of this disease entity vary depending on the organ system involved. Signs of internal jugular vein thrombosis include swelling, tenderness, or induration over the neck [2]. Atypical presentation of Lemierre's syndrome includes an original focus of infection to be outside the head and neck, implicated organism other than *Fusobacterium* spp, and thrombophlebitis of a vessel other than the internal jugular vein [3]. It is rare, affecting previously healthy young individuals with a global incidence of 1/1,000,000 [4]. An increase in incidence due to antimicrobial resistance and restrictions in antibiotic prescription has been described [5-7]. This disease entity is potentially fatal with a mortality rate of 4 - 12% [5].

Case Report

In this case report, we describe an atypical presentation of Lemierre's syndrome of a 3-year-old boy presenting with mastoiditis secondary to *Proteus mirabilis*. This is complicated by Lemierre's syndrome with thrombosis of the distal right transverse sinus.

Patient information

A 3-year-old boy with a past medical history of chronic suppurative otitis media and bilateral cholesteatoma presented to the Emergency Department with a chief complaint of a two-week history of fever, and non-bilious, non-projectile vomiting associated with right-sided purulent ear discharge with occasional blood. A week later, he developed a protrusion of the right ear associated with erythema and tenderness. The fever was high grade, measuring 39.5-40 via axillary route with only partial response to antipyretics. A review of systems was remarkable for a significant decrease in activity and oral intake. There was no history of altered level of consciousness, neck stiffness, photophobia, or phonophobia. Of note, multiple visits to ambulatory care were sought by parents receiving symptomatic treatment and amoxicillin-clavulanic acid for 6 days before presentation, without improvement. The child's immunization status was up to date. Moreover, there was no significant family history reported by parents.

Clinical findings

Upon presentation, he was conscious, irritable, ill-looking, and afebrile. He was hemodynamically stable with systolic blood pressure of 101mmHg, diastolic blood pressure of 48 mmHg, and heart rate of 130 bpm. Respiratory rate was within normal value for age. The temperature was documented to be 37.4 with multiple spikes of fever reaching 39c. He maintained > 94% SpO_2 on room air. Physical examination was remarkable for right post-auricular erythema, swelling, protrusion, and tenderness. Right-sided otorrhea consisting of purulent fluid was noted. Other systemic examinations, including neurological assessment, were unremarkable.

Diagnostic assessment

Pertinent laboratory findings included within normal leukocyte count of 8.6, neutrophil count of 5.20, and thrombocytosis of 627. Inflammatory markers were elevated with CRP of 222.5 procalcitonin of 1.17, and ESR of 106. Furthermore, a blood culture and a culture from the right ear discharge were withdrawn. Blood culture showed no growth. Ear culture later revealed *Proteus mirabilis* which was sensitive to amoxicillin/clavulanic acid and fosfomycin. Our differential diagnosis included mastoiditis with intracranial extension. Due to the child's ill appearance and findings on physical examination, a CT brain scan was promptly performed, and piperacillin/tazobactam was initiated initially. Later, a CT brain scan revealed a right coalescent mastoiditis with epidural abscess and thrombosis of the distal right transverse sinus (Figure 1).

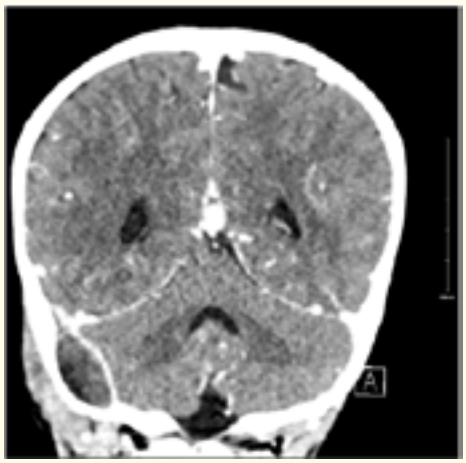

Figure 1

Therapeutic intervention

He underwent a right cortical mastoidectomy with middle ear exploration five days after admission. During the operation, culture was taken from the fluid of the right mastoid, which subsequently revealed coagulase-negative *Staphylococcus*. The procedure was tolerated well without events or complications. piperacillin/tazobactam was stopped after CT brain findings. He was started on vancomycin 15 mg/kg/dose q6h and meropenem 40 mg/kg/dose q8h for a total of 6-week duration. Enoxaparin was initiated for anticoagulation.

Follow-up and outcomes

With antimicrobial and anticoagulation therapies, his overall status returned to his baseline with complete resolution of symptoms. He remained on anticoagulation therapy pending a follow-up CT brain imaging. Repeated brain CT scan 12 weeks on anticoagulation treatment showed a regression of the size of the collection from 1.3 cm to 0.3 cm (Figure 2).

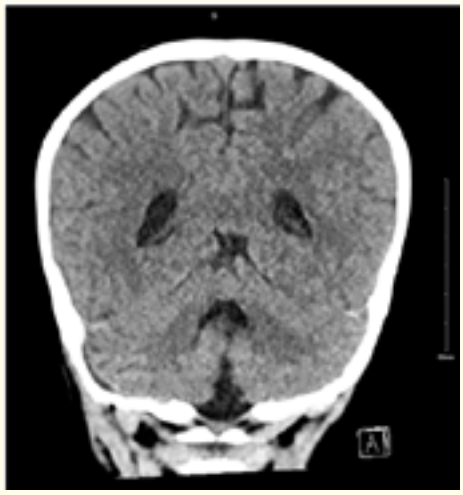

Figure 2

Discussion and Conclusion

Lemierre's syndrome is rare, and our knowledge of this disease comes mostly from case reports. Hence, recognizing this disease entity can be challenging for healthcare workers. Moreover, the classical criteria to diagnose Lemierre's syndrome include a focus of infection in the head or neck, internal jugular venous thrombosis, and isolating *Fusobacterium* spp [4]. To describe Lemierre's syndrome as atypical, it must include the following: 1. A focus of infection outside the head and neck, 2. Implicated organism other than *Fusobacterium* spp, and 3. Thrombophlebitis of a vessel other than the internal jugular vein. A new consensus challenging the classical criteria has been recognized for this disease due to a variety of reasons [3]. Internal jugular venous thrombosis is not always evident at the initial presentation in cases of Lemierre's syndrome [3], as the signs and symptoms of internal jugular vein thrombosis may occur throughout this disease, they can be missed or not be present at initial presentation. A classical presentation with Lemierre's syndrome without evidence of internal jugular venous thrombosis on ultrasound, CT, or MRI has been described in the literature [8]. *Fusobacterium* spp is widely known to be associated with Lemierre's syndrome. However, only half of the reported cases published have described this organism to be the causative agent [3]. *Fusobacterium* spp may not be isolated from cultures as many cases of Lemierre's syndrome present when empiric antimicrobial therapy has already been initiated [3], as in our case report. Moreover, in otogenic infections with *F. necrophorum*, 69% were found to

have mastoiditis. Of these, 66% developed intracerebral complications [8]. Suggesting an aggressive behavior of this organism when established as the causative agent [8].

The most commonly causative organisms for Lemierre's syndrome include *Fusobacteria* spp, other organisms associated with this disease include *Enterococcus*, *Bacteroides*, *Streptococcus pyogenes*, *Peptostreptococcus* and *Proteus mirabilis* [9] as described in this case report.

The use of anticoagulation therapy is controversial in cases of Lemierre's syndrome. There are no available guidelines or a clear consensus for the initiation of anticoagulation therapy. The decision to start anticoagulation therapy should be tailored to individual cases. In a study by Valerio L., *et al.* it was found that patients with early thromboembolic complications who did not receive anticoagulation therapy were more likely to develop intracranial involvement, underwent more surgical procedures, and were administered four or more antibiotics compared to those without early thromboembolic complications [10]. Suggesting favorable prognostic outcomes in the use of anticoagulation therapy before the rise of thromboembolic complications and absence of intracranial involvement at presentation [10].

Lemierre's syndrome is a potentially fatal disease with an estimated mortality rate of 4 - 12% [5]. However, an increase in survival rates has been described in the recently published case reports. This can be attributed to an increase in the awareness of this disease, and subsequently an increase in its identification resulting in prompt initiation of antimicrobial therapy.

Lemierre's syndrome is rare. This case report sheds light on the atypical presentation of Lemierre's syndrome, which is negligible in the literature with only a few cases reporting it. Additionally, the majority of these cases are present in the setting of ambulatory care. A high index of suspicion amongst clinicians is crucial to identify and promptly manage it to prevent the devastating morbidity and mortality associated with this disease entity if left untreated.

Informed Consent

A verbal consent was obtained from the patient's mother.

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