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Case Report

Mastoiditis with concomitant Lemierre's syndrome

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ABSTRACT

Otalgia with mastoiditis is an infrequently encountered complication of acute otitis media (Pfaff and Moore, 2018). Even more rare is the development of infected jugular venous thrombosis, Lemierre's disease. We present a case of a six year-old girl with otalgia for over two months who presented to our Emergency Department (ED) with clinical mastoiditis, confirmed on CT scan, as well as an incidental diagnosis of complete thrombosis of the internal jugular (IJ) vein, Lemierre's syndrome. The true prevalence of Lemierre's from mastoiditis is difficult to discern. This clinical case highlights the importance of the consideration of these pathologies by the emergency physician.

1. Introduction

Mastoiditis is the most frequent suppurative complication of acute otitis media (AOM) [1] but is still very uncommon. Lemierre's syndrome, concomitant thrombosis of the internal jugular vein, is even less frequently encountered. This article addresses the ambiguous definition of Lemierre's syndrome, as well as an approach to its diagnosis and management.

2. Case report

A previously healthy six-year-old girl presented to the emergency department after having ten outpatient visits for ear complaints over the preceding two months. She had completed several outpatient courses of Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) and oral antibiotics, in line with prescribing guidelines for the treatment of acute AOM by both her primary care physician and an Otolaryngologist [2].

The patient's vital signs on presentation to the ED included a blood pressure of 93/47 mm Hg, heart rate of 83 beats/min, respiratory rate of 18 breaths per minute and tympanic temperature of 97.7 F. It was noted that the patient had a 2.3 kg weight loss since her symptoms had started two months earlier. On physical exam, the patient had an unremarkable right ear. The left ear was prominent with a large and erythematous soft mass with post-auricular fluctuance over the mastoid. The mass was well demarcated, tender to the touch, and warm. The left ear was also noted to have a bulging, fluid filled tympanic membrane (TM) with no visible landmarks.

Her physical exam findings on presentation to the ED led to a clinical diagnosis of mastoiditis which was not a finding noted on previous visits. A computed tomography (CT) scan of the patient's temporal bones confirmed mastoiditis and demonstrated (Image 1) complete opacification of the middle ear and mastoid air cells with bony erosion into the mastoid and sinus plate. There was an associated multiloculated, rim enhancing, subperiosteal abscess centered over the left mastoid air cells (measuring 4.3 × 3.5 × 1.2 cm). A large filling defect was noted in the mid-left transverse sinus extending into the left sigmoid sinus and IJ vein, with extension to the cavernous sinus and was concerning for Lemierre's syndrome (Image 2). The patient diagnosed with left acute mastoiditis, post-auricular abscess, and Lemierre's syndrome and was given vancomycin and cefepime. An Otolaryngologist was consulted and the patient was taken to the operating room (OR) where a left mastoidectomy and bilateral myringotomies with tympanostomies were performed.

From the OR the patient was admitted to the pediatric intensive care unit where her care was overseen by specialists from infectious disease, neurology and vascular surgery. She was treated with cefepime, vancomycin and metronidazole, which were later narrowed to cefepime and metronidazole. The patient had a tunneled central catheter placed for antibiotic administration. The thrombosed IJ vein was treated with anticoagulation with intravenous heparin, which was then switched to enoxaparin for three months. The Gram stain from the patient's abscess drainage in the operating room and mastoid cultures showed no organisms and no aerobic or anaerobic growth, respectively. The white blood cell count, as well as inflammatory markers (sedimentation rate and C-reactive protein) were trended and decreased to normal by the patient's time of discharge. During the course of admission magnetic resonance imaging was performed and the findings were consistent with the initial CT scan showing left mastoiditis and Lemierre's syndrome. The patient was discharged to home on hospital day six and

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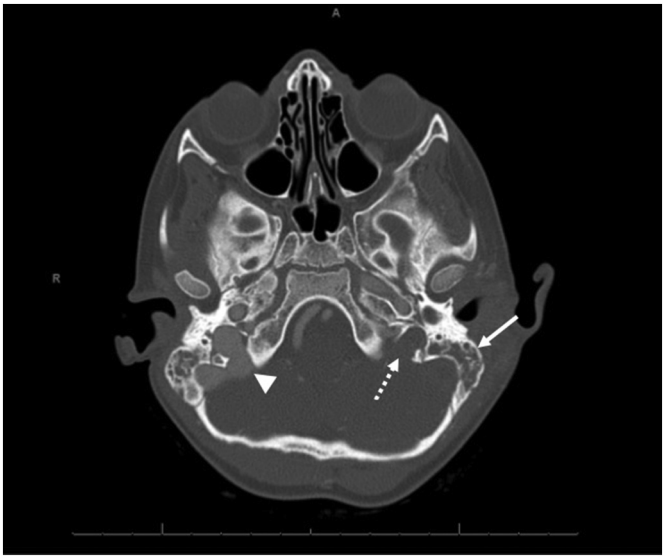

Image 1. Axial view of computed tomographic head imaging demonstrating left mastoid air cell destruction from left otitis media with associated thrombosis of the left internal venous system. (Legend: The white solid-line arrow demonstrates an area of mastoid air cell destruction. The white dashed line arrow represents the absence of contrast media in the left internal jugular vein system. The white arrow-head demonstrates contrast media filling the right internal jugular vein system.)

completed a 42 day courses of metronidazole and cefepime. The patient also completed a three-month course of enoxaparin anticoagulation.

3. Discussion

Mastoiditis is the most frequent suppurative complication of AOM [1]. The Incidence of surgical intervention for mastoiditis from acute AOM is reported as 0.004% in the United States and is typically seen in those less than two years of age. Symptoms include fever, headache, otalgia (universally present), erythema, obliteration of the postauricular crease, and otorrhea [1,3,4]. Signs may include postauricular erythema and tenderness, protrusion of the auricle, and TM abnormalities similar to AOM (bulging, decreased mobility and erythema). With typical findings the diagnosis may be made clinically, however it is suggested that a CT scan is obtained if there are neurological symptoms, when intracranial complications are suspected or there is failure to improve with conservative treatment [1]. The leading cause of acute mastoiditis is from *Streptococcus pneumonia*, however other organisms include *Moraxella catarrhalis*, Group A *streptococci*, *Staphylococcus aureus*, *Haemophilus influenzae* and *Pseudomonas aeruginosa*. Antibiotics for mild mastoiditis include penicillinase-resistant penicillins, amoxicillin-clavulanic acid, third-generation cephalosporins and newer macrolides [5]. For severe cases, vancomycin and third generation cephalosporins are recommended [6]. If surgical procedures are needed, they may range from myringotomy and tympanostomy tube placement for moderate cases, to mastoidectomy and drainage (with cultures) for severe cases. A rare complication of mastoiditis may be Lemierre's syndrome [6].

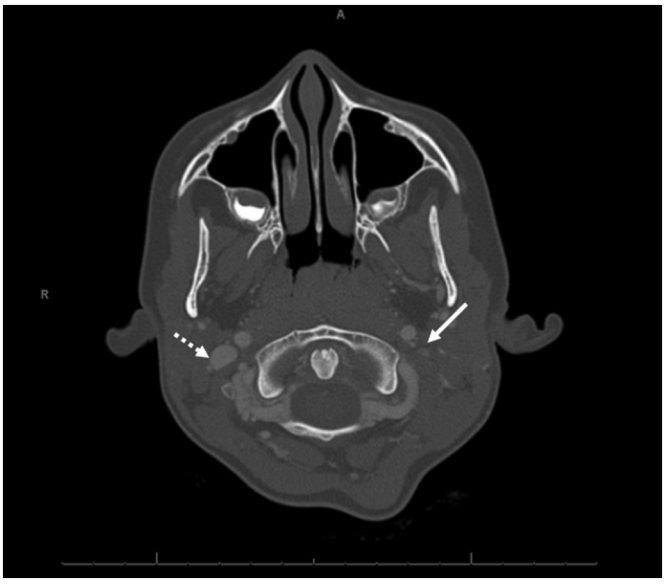

Image 2. Axial view of computed tomographic head imaging demonstrating left internal jugular vein occlusion. (Legend: The solid-line white arrow indicates occlusion of the left internal jugular vein, noted by the absence of contrast media in the vein. The dashed-line white arrow indicates the presence of contrast media in the contralateral vein.)

While mastoiditis is a rare occurrence, the complication of Lemierre's syndrome is even more infrequently encountered. André Lemierre was not the first to describe post-anginal septicemia [7], however his detailed description in 1936 earned him eponymous fame. The 1936 case definition of the disease has morphed into a more broad and variable one, dependent on the individual author or researcher, and only serves to confound a calculation of true incidence [8]. The original definition includes septicemia from IJ thrombosis secondary to pharyngotonsillitis or peritonsillar abscesses, specifically secondary to infection with *Fusobacterium necrophorum* [8], however authors of subsequent case series have expanded the definition to include infections from additional sources outside of the oropharynx, and to additional bacterial etiologies. The broader, yet poorly demarcated, definition includes all infectious etiologies associated with or resulting in IJ thrombosis. We endorse the broader definition of the disease, which would include all infectious thrombotic events concerning the IJ vein and its proximal tributaries, as this definition serves to include the potentially devastating consequences of infectious thrombotic emboli, which is the common terminal process if the disease state is left untreated.

Given the variation in definitions, the incidence of Lemierre's syndrome ranges from 0.6 to 2.3 per million people [9]. The classic Lemierre's syndrome is typically seen in previously healthy adolescents and young adults, however the broader definition which includes otalgic infections as a cause expands this population to young previously healthy children and some older adults [8]. There is generally a history of acute oropharyngeal infection with radiological evidence of secondary septic thrombophlebitis of the IJ vein and resultant metastatic complications [10-12]. Although classical cases are attributed to *F. necrophorum* infection [13], there have been cases associated with *Streptococcus pyogenes*, Group A strep, and other organisms [9-11,14]. Our literature review found four cases of Lemierre's that were attributed to mastoiditis [9,14-16]. There has not been a reported case with negative culture to our knowledge, though this may be the result of publication bias. Empiric treatment is with intravenous antimicrobials to treat the classic anaerobic causative agent, *F.* necrophorum, as well as those bacteria common to Ear, Nose and Throat disorders, including streptococcus species. Specific agents usually include either metronidazole or clindamycin and either a third-generation cephalosporin or beta-lactam with beta-lactamase inhibitor. Given the location of infection and potential for serious sequelae, including mycotic embolic stroke and other end-organ infectious complications, therapy is typically extended on the order of 30 or more days. Anticoagulation is necessary if there is thrombosis of the cavernous sinus [17]. When left untreated, Lemierre's syndrome has a mortality rate of up to 90% [5].

The challenge to the clinician is the identification of the disease process as early as possible. This case demonstrates the need for the emergency physician to be aware of the potential for complications from the routine causes of ear pain. Both mastoiditis and Lemierre's disease are uncommonly encountered. Given the limited amount of data and case

reports, it is difficult to drawn firm conclusions regarding a standard approach to the identification of the disease process. Little is known about specific precursors to the condition. Late findings of IJ thrombosis include distention of the IJ and pain over the ipsilateral sternocleidomastoid musculature of the neck, as well as signs of embolic phenomena, including brain, lung and other end-organ abscesses. Local head and neck infectious etiologies place patients at risk, as do the presence of IJ, subclavian or other local venous catheters. Outside of these two risks, which do not make up many of the case reports, there are no clear early predictors or exam findings of the risk for or development of IJ thrombosis. Severe illness is the only consistent clinical finding indicative of the disease, by which time the process is likely advanced. In our case, we were fortunate to identify the disease process early enough that there was no severe illness and no neurologic sequelae. We suggest increased recognition of this disease entity, especially in an ill person with a head or neck infection, and to have a reasonably low threshold for obtaining a CT scan with IV contrast to help evaluate for the condition, particularly when an infection has failed to respond to appropriate antimicrobial therapy. A screening tool for Lemierre's disease is not practical due to the low incidence of the disease. There is no evidence to support early or aggressive antibiotic use in ill appearing patients with specific head and neck infections, but given the risk of sequalae, it seems reasonable. As usual, blood cultures should be obtained prior to antibiotic administration. In our case, the lack of culture growth was an interesting finding, suggesting either effects from early antibiotic administration, no infectious etiology, or a viral or bacterial source which was difficult to culture in the routine setting.

4. Conclusion

Herein we identify a rare case of Lemierre's syndrome in an otherwise healthy young girl who failed to respond to appropriate antibiotic therapy for AOM. We suggest that this failure of response to appropriate antibiotic treatment in head and neck conditions, in combination with a worsening clinical status, should raise suspicion for developing the condition. Using a broad definition of Lemierre's disease may allow future researchers to find nuanced risk factors for developing the condition.

Meetings

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Conflicts of interest

N/A.

Author contributions

Mark Olaf, D.O. produced the majority of the write up, completed all editing and significant revisions, as well as the literature review and reference list. Lindsey Duguet, D.O. produced the initial draft of this submission.

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