Actinomycosis Mastoiditis Complicated by Sigmoid Sinus Thrombosis and Labyrinthine Fistula

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INTRODUCTION
Actinomycosis is a rare infection caused by a human commensal bacterium which shares some properties with fungi. Its indolent course, non-specific symptoms, and resistance to growth in culture can make the diagnosis challenging. We describe an unusual case of temporal bone actinomycosis presenting with a neck mass and otogenic Lemierre’s syndrome.

CASE REPORT
A 10-year-old male with no otologic history presented after two months of intermittent nausea and vomiting associated with headache, fatigue, fever, and generalized malaise. He reported an 11 pound weight loss over two months. He denied otologic symptoms including hearing loss, vertigo, tinnitus, and otalgia. During several trips to his pediatrician and a community hospital emergency department, he was never found to have evidence of acute otitis media. Five days prior to presentation at our hospital, he developed left neck pain and swelling. His fevers became persistent and he was admitted to the hospital for further workup.

Initial contrast-enhanced head and neck CT revealed a left internal jugular vein thrombosis and a left mastoid effusion. Contrast-enhanced MRI of the neck supported the diagnosis of left internal jugular vein thrombosis and also demonstrated enhancement of the left mastoid and middle ear. Additional imaging including MRV and temporal bone CT demonstrated left middle ear and mastoid opacification, erosion of the ossicles and lateral semicircular canal, and extension of the internal jugular vein thrombus to the sigmoid sinus to include a complete left sigmoid sinus thrombosis (Figure 1). Audiometry demonstrated a left profound sensorineural hearing loss (Figure 2). The patient underwent a left mastoidectomy with decompression of the sigmoid sinus, repair of the lateral semicircular canal fistula, and a left myringotomy with tube placement. Histological analysis revealed small fragments of bone surrounded by fibrinous debris and blood vessels associated with Actinomyces colonies (Figure 3).

Blood and tissue cultures were persistently negative. The infectious disease team recommended six weeks of intravenous meropenem followed by oral amoxicillin/clavulanic acid for a total of six months. The patient was also placed on anticoagulation with enoxaparin for six months. Postoperatively, the patient improved clinically. He was discharged home with close follow up. His vertigo, nausea, fevers, headache, and neck mass have resolved. He has had a persistent left profound sensorineural hearing loss, likely secondary to the labyrinthine fistula.

DISCUSSION
This pediatric patient had an erosive process of the temporal bone caused by actinomycosis, a rare but significant pathogen that can cause serious complications of chronic mastoiditis. Actinomyces species are Gram positive, anaerobic, filamentous bacteria. They are present as normal flora in the oral cavity and pharynx. Disease with this organism is frequently an indolent, supplicative process, most commonly in the cervicofacial region. Transit through the Eustachian tube into the middle ear has been proposed as the mechanism for otomastoid actinomycosis.

Our patient had a complex and confusing presentation. The initial symptoms of nausea, vomiting, headache, malaise, and fever are non-specific and generate a broad differential diagnosis. This case was complicated by sigmoid sinus thrombosis, labyrinthine fistula, and otogenic Lemierre's syndrome. Already exceedingly rare, with no more than 27 cases reported in the English literature, this particular example of temporal bone actinomycosis differs from previously described cases since our patient did not present with most of the classic symptoms of otomastoiditis including hearing loss, vertigo, otalgia, and ototrauma, although audiology did demonstrate a left profound sensorineural hearing loss. This case also represents a particularly destructive clinical course. While invasion of the bony labyrinth, Fallopian canal, and petrous apex have been previously reported, this pathogen typically progresses in an indolent and chronic manner.

Actinomycosis typically responds to long-term high-dose penicillins. Surgical debridement is often necessary, as this anaerobic species can survive for long periods within poorly vascularized tissues where antibiotics may not reach therapeutic concentrations.

CONCLUSION
• Actinomyces is a rare pathogen that can be responsible for complications of chronic mastoiditis.
• Histological analysis of evacuated mastoiditis debris is an effective method for detection of this difficult to culture microbe.
• Intracranial and intratemporal complications of mastoiditis should be considered in pediatric patients presenting with nausea, vomiting, and headache.

REFERENCES