Pediatric Middle Ear Lesion Presenting a Diagnostic Challenge

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ABSTRACT

Introduction
A three-year-old male presented with intractable headache, extremity pain, and emesis. He was transferred from another facility with concern for "cholesteatoma". After extensive workup, diagnostic mastoidectomy, and further imaging, lesion suspected as middle ear lymphangioma with concomitant bilateral transverse sinus stenosis.

Methods
Patient presented as a transfer to our tertiary center in November 2022 with new-onset progressive headache, emesis, and upper extremity pain. Per family, he had no significant past neurotologic history. Initial outside computerized tomography (CT) was concerning for right ear cholesteatoma. He was taken to the operating room for diagnostic/therapeutic mastoidectomy, where external auditory canal bony erosion was noted with cerebrospinal fluid (CSF) leak occurring immediately after the tympanomeatal flap incisions were made. No mass was discovered, though ossicular erosion and dusky mucosa were noted. Biopsies were obtained and the CSF leak repaired. His headaches improved, with no CSF leak post-operatively. After discharge, he re-presented two days later with severe headache, but normal exam. Lumbar puncture (LP) was performed with an opening pressure of 58 centimeters of water (cm H^20). He underwent magnetic resonance venogram showing bilateral transverse sinus stenosis. Initial attempts at medical management with acetazolamide failed, and the patient returned ten days later with headache. He underwent repeat LP with an opening pressure of 54 cm H^20. Neurosurgery proceeded with bilateral transverse sinus stenting and post-operative anti-coagulation.

Results
Biopsies and cultures were taken from the right middle ear. Final pathology was consistent with reactive inflammation and intraosseous vascular proliferation, favoring the diagnosis of lymphangioma.

Conclusions
No prior case report exists in the English literature demonstrating a pediatric patient with concurrent lymphangioma and bilateral transverse sinus stenosis. This case demonstrates the necessity of thorough diagnostic workup and avoiding the pitfall of anchoring to an outside diagnosis.

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INTRODUCTION

This case describes the rare presentation of a three-year-old who presented to the emergency department several times with intractable headaches. The patient was initially transferred with concern for "cholesteatoma", this misdiagnosis led to persistent confusion in the complex management of the patient.

The patient underwent extensive workup, including imaging and mastoidectomy. Final diagnosis was found to be middle ear lymphangioma with bilateral transverse sinus stenosis, consistent with Gorham-Stout Disease.

NARRATIVE

Our three-year-old male presented initially as a transfer to our tertiary center in late 2022 with intense and intractable headache, emesis and unilateral upper extremity pain, with no known medical or family history of significance.

Upon transfer, he had an outside computerized tomography (CT) scan, with concern for a right middle ear cholesteatoma. Repeat imaging was obtained at our institution, showing a soft tissue lesion of the external auditory canal, with ossicular erosion, as well as erosion of the tegmen tympani and mastoideum concerning for cholesteatoma (Figure 1). He was taken to the operating room for a mastoidectomy, where he was found to have external auditory canal bony erosion. A cerebral spinal fluid (CSF) leak occurred immediately upon raising the tympanomeatal flap. Intraoperatively there was no discrete mass identified, however lytic changes to the ossicular chain and dusky mucosa throughout the middle ear, were seen. Biopsies were obtained and the CSF leak repaired.

Post-operatively the symptoms improved, with no recurrence of the CSF leak. Two days after discharge he returned to the emergency department with severe headache with a normal exam. The emergency medicine team was unfortunately concerned about a "residual cholesteatoma," delaying care and diagnosis. Eventually, lumbar puncture (LP) was performed with an opening pressure of 58 centimeter of water (cm H^20). At this time, he underwent magnetic resonance imaging (MRI) brain (Figure 2) and venogram (Figure 3), which showed middle ear fluid signal and bilateral transverse sinus stenosis. Initial attempts at medical management with acetazolamide failed, and the patient returned ten days later with headache. He underwent a repeat LP with an opening pressure of 54 cm H^20. Neurosurgery proceeded with bilateral transverse sinus stenting with post-operative anti-coagulation. Ultimately, he was discharged without symptoms.

Final pathology from the lesion was described as a lymphatic malformation. Patient further followed at a vascular anomalies' clinic, where he was diagnosed with Gorham-Stout Disease and placed on a combination of Sirolimus and Zoledronic acid.

Figure 1. CT scan showing non-descript right middle ear soft tissue lesion with bony erosion.

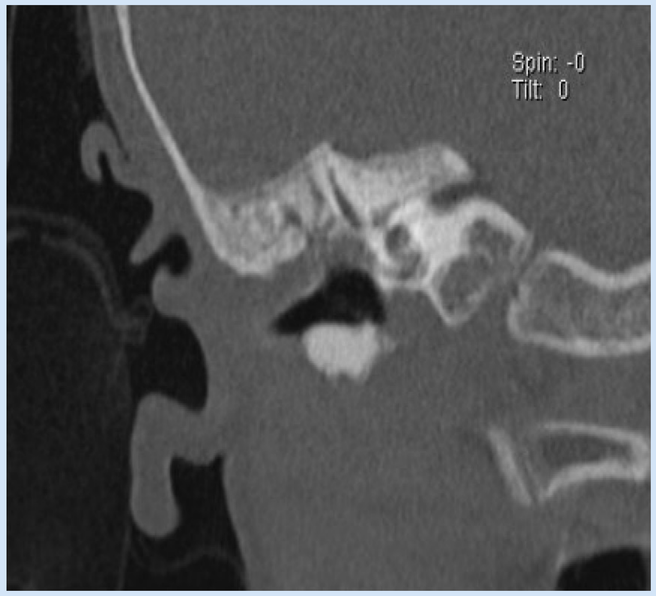


Figure 2. MRI scan showing non-descript right middle ear soft tissue lesion with bony erosion.

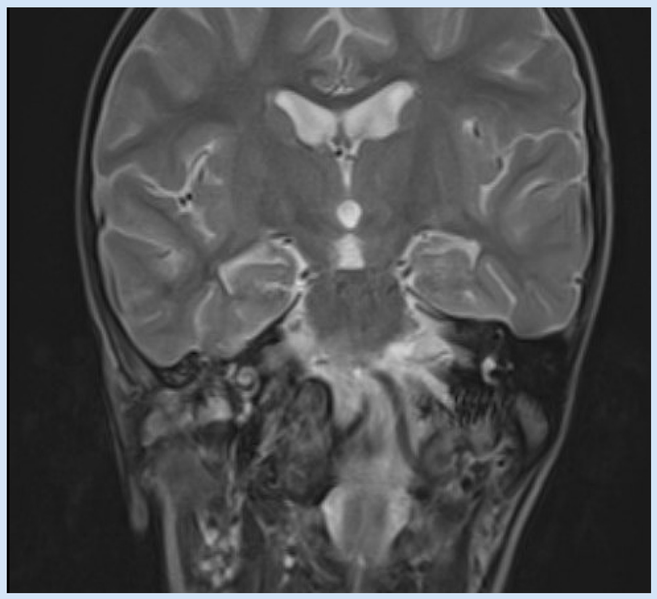
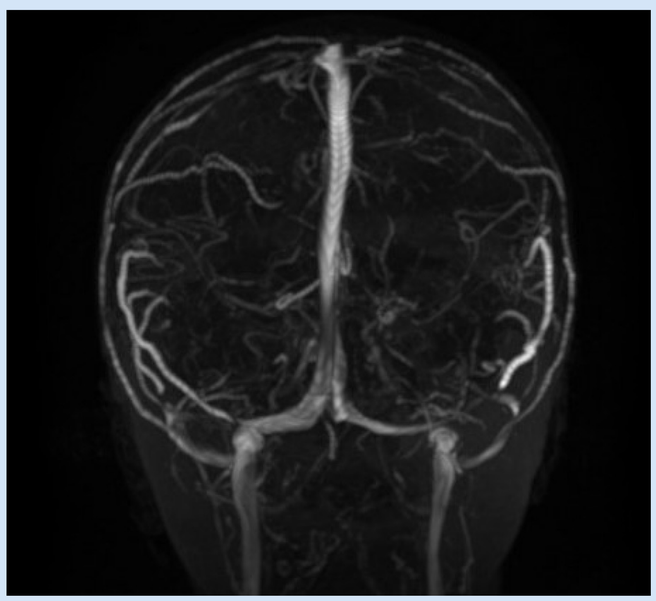


Figure 3. MR venogram scan showing bilateral transverse sinus stenosis and decreased flow.



DISCUSSION

There are few published studies exploring the etiology of pediatric middle ear masses due to their rarity. Middle ear lesions in pediatric patients are often malignant[1]. The most common malignant lesion of the middle ear in the pediatric patient is often cited as rhabdomyosarcoma[2].

Gorham-Stout Disease (GSD) is an extremely rare condition, with a recent review showing only 350 published cases[3], leading to a difficult diagnostic challenge. GSD is a disease of bone resorption, which is idiopathic in nature and is typically progressive. It may have a single or multiple bony lesions, has no sex or age predominance and is often sporadic[3].

Diagnosis of GSD requires high-suspicion and surgical pathology. Although imaging may be suggestive (non-descript osteopenia), pathology is typically required for diagnosis. Histopathology demonstrates non-malignant hyperproliferation of intraosseous vascular structures, compatible with lymphangiomatosis or hemangiomatosis, with increased bony resorption[4].

In most cases, GSD is managed pharmacologically, with the aim of decreasing bony resorption. Primary management via case reports has been bisphosphonates and immunosuppressants[5]. Treatment generally leads to decreased recurrence of symptomatic disease.

CONCLUSIONS

This case illustrated how a middle ear mass in a pediatric patient presents a diagnostic challenge, due to their rarity and unusual presentation. Additionally, this case demonstrated how communication between teams is key to ensure a proper diagnosis. Lastly, this case demonstrated how anchoring to an initial diagnosis can ultimately delay the true diagnosis and proper treatment.

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