Rare Mastoid Pathology Presenting as Otalgia

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ABSTRACT

Objectives: To raise awareness of rare causes of otalgia due to mastoid pathology and submit representative cases to the medical literature.

Methods: Retrospective review of cases of rare mastoid pathology presenting as otalgia in an academic tertiary care medical center. Etiology, presenting symptoms, medical and surgical interventions, pathology, imaging, and outcomes were recorded. The medical literature relevant to each etiology was reviewed and summarized.

Results: Three types of rare mastoid pathology presenting as otalgia were identified: 1) Gout, 2) Bone wax granuloma, and 3) Langerhans cell histiocytosis. These patients typically presented with severe otalgia and were suspected of having skull base osteitis. Thus, most patients were treated unsuccessfully for bacterial otomastoiditis prior to definitive diagnosis. The ultimate diagnosis was reached through a combination of radiologic evaluation and mastoid histopathology, which guided further curative or palliative treatments.

Conclusion: Difficult cases of otalgia or presumed otomastoiditis unresponsive to broad spectrum antibiotics require thorough radiographic evaluation. When the diagnosis remains uncertain, mastoid biopsy and histopathological analysis may provide important diagnostic information. Awareness of more rare etiologies is essential and can facilitate efficient diagnosis and treatment. To our knowledge this is the first report of acute inflammatory gout of the mastoid and second report of mastoiditis secondary to bone wax granuloma.

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INTRODUCTION

Otalgia is a very common complaint in primary care and a frequent reason for referral to an otolaryngologist or neurotologist. Otalgia can be classified as primary (otogenic) or secondary (referred). The differential diagnosis of primary otalgia is broad. Common etiologies include various forms of otitis media (acute, serous, chronic tympanomastoiditis, cholesteatoma), complications of otitis media (acute mastoiditis, subperiosteal abscess, TM perforation, petrous apicitis), malignant otitis externa, acute or chronic otitis externa, chondritis, myringitis, Bell's Palsy, Ramsay-Hunt syndrome, various skin conditions, and temporal bone neoplasms[1]. For most of these primary otalgias, the diagnosis is evident on clinical exam, imaging, or laboratory evaluation. Occasionally, a patient will present with primary otalgia of unclear origin and refractory to treatment. Here, we present three rare causes of otalgia see in a tertiary care medical center.

PATIENT #1: GOUT

A 64 year old man presented with one week of severe progressive left otalgia characterized as "stabbing" and "boring," and refractory to narcotic pain medications. He reported mild left hearing loss, but denied vertigo, otorrhea, facial weakness or numbness, fevers, nuchal rigidity, and altered mental status. The patient had a history of diabetes mellitus, distant past lower extremity gout, shingles, and myelodysplastic syndrome with leukemic transformation treated with Decitabine. On binocular microscopy, the left external auditory canal was very tender to palpation, but without edema or erythema. The eardrum position and landmarks were normal. There was no mastoid tenderness or overlying skin change. Initial laboratory evaluation was notable for leukocytosis (13K) with relative neutrophilia (82%) and an elevated erythrocyte sedimentation rate (60). CT temporal bone showed left middle ear opacification and partial mastoid opacification without coalescence or erosion (Figure 1).

At this time, the differential diagnosis included otitis externa, malignant otitis externa, and early varicella zoster infection. The patient started oral and topical fluoroquinolones and topical steroid drops. Despite aggressive treatment with narcotics, his pain persisted. The patient underwent myringotomy which demonstrated granulation tissue in the middle ear. Cultures of this tissue were unremarkable and pathology showed only acute inflammation. Meanwhile, an MRI showed heterogenous mastoid enhancement with multiple small fluid collections (Figure 2), and a three phase technetium bone scan with delayed SPECT-CT showed non-specific mastoid uptake with no focal arterial hyperemia. These findings decreased the likelihood of acute osteomyelitis.

As the patient continued to have intractable pain and no discrete diagnosis on multiple studies, he underwent mastoidectomy. There was extensive granulation tissue filling the mastoid antrum and, to a lesser degree, the mastoid cavity. No purulence was encountered. Final pathology revealed acute inflammation surrounding extensive negatively birefringent needle shaped crystals consistent with acute gout. The patient's uric acid level was 7.1. Narcotics were replaced with indomethacin, and the patient had complete resolution of otalgia.

PATIENT #2: BONE WAX GRANULOMA

A 50 year old woman with a history of right retrosigmoid resection of an acoustic neuroma at age 32 presented with two years of intermittent right otalgia. Her pain was only partially relieved by neurontin or narcotics. She had longstanding right anacusis, no otorrhea, and no recent vertigo. Previous imaging showed stable opacification of the right mastoid. Physical exam was normal except for right facial paresis (House-Brackman IV/VI). MRI showed homogenous isointense T1 and T2 signal with gadolinium enhancement in the right mastoid, surrounding bone and overlying dura (Figure 3). The CBC was normal with elevated ESR (40 mm/hr). The patient underwent mastoidectomy which revealed extensive granulation tissue surrounding a core of encapsulated bone wax. Her symptoms completely resolved.

PATIENT #2: BONE WAX GRANULOMA

A 52 year old man was evaluated for left otalgia and otorrhea. He had undergone transmastoid excision of a cerebellopontine angle tumor two years prior with left "milky, occasionally bloody" otorrhea developing shortly thereafter. There was no improvement after multiple rounds of oral and topical antibiotics. On physical exam, there was extensive granulation tissue in the external auditory canal. Biopsy demonstrated only fibroconnective tissue with acute on chronic inflammation. CT Scan showed mastoid opacification without destruction, and MRI showed a nonenhancing mass that was isointense on T1 and T2 series. During mastoidectomy, extensive granulation tissue was found encapsulating a large plug of bone wax. Pathology demonstrated chronic inflammation with extensive plasma cells consistent with foreign body reaction (Figure 4). The patient has had no further otalgia or otorrhea.

PATIENT #4: LANGERHANS CELL HISTIOCYTOSIS

A 34 year old female complained of 8 months of intermittent left otalgia and mild left hearing loss. She had been diagnosed twice with acute otitis media in emergency departments and treated both times with a wick, oral antibiotics and antibiotic drops. She was then referred to an otolaryngologist. She complained of a "water logged" sensation, and audiologic evaluation demonstrated a mild left conductive hearing loss with type B tympanogram. She underwent left myringotomy with PE tube placement. One week later, the patient's left hearing loss worsened, and she developed brief episodic vertigo with head movements, bending over, or pressure on the tragus. Eight months after symptom onset, she presented to our clinic. A non-contrast temporal bone CT scan demonstrated an erosive lesion of the left mastoid with extension into the superior semicircular canal, lateral semicircular canal, vestibule, tegmen tympani and tegmen mastoideum (figure 5). MRI of the brain and temporal bone characterized the lesion as isointense on T1, heterogeneous on T2, and strongly enhancing with gadolinium.

The patient was counseled to undergo transmastoid resection of the lesion. Routine preoperative workup revealed multiple lung nodules which abnormally enhanced on PET scan. In light of the pulmonary lesions, we performed a simple mastoidectomy with biopsies which were diagnostic of Langerhans cell histiocytosis (Figure 6). The patient completed six months of chemotherapy with vinblastine and captopurine and has had complete resolution of otalgia, vertigo, and hearing loss, and radiographic resolution of all lesions.

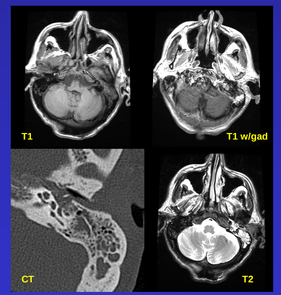

Figure 1. Patient 1 Imaging

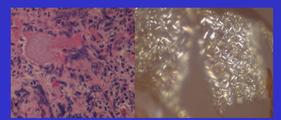

Figure 2. Patient 1 Pathology: Acute Inflammatory response (A) with negatively birefringent crystals

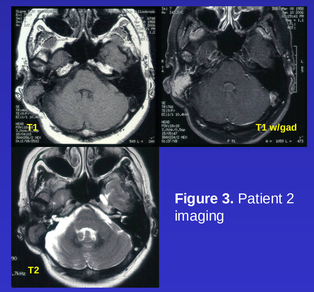

Figure 3. Patient 2 imaging

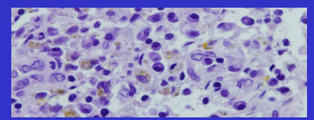

Figure 4. Patient 3 Pathology: Chronic inflammation with polyclonal plasma cell infiltrate

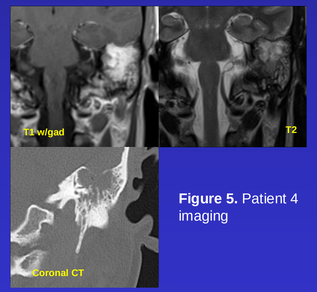

Figure 5. Patient 4 imaging

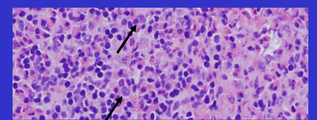

Figure 6. Patient 4 Pathology: Eosinophilia with characteristic Langerhans Cells (arrows)

DISCUSSION

These four patients represent three very rare causes of otalgia. Three of the four patients had a misdiagnosis of otitis externa and no or minimal change with topical therapy. In all patients, abnormal imaging of the temporal bone was not specific to an etiology, but did suggest an atypical process at work in the mastoid. All patients had complete resolution of otalgia and rapid response to therapy once a diagnosis was established.

Tophaceous gout of the auricle is well described and rather common. There has been multiple reports of middle ear tophaceous gout presenting with conductive hearing loss or a sensation of fullness in the ear[2-3]. To our knowledge, this is the first report of acute inflammatory gout causing otalgia and mastoid/middle ear granulation. The severity of the patient's pain and lack of response to narcotics as well as his rapid response to NSAIDs were important features that, in retrospect, are very consistent with acute arthritic gout. His serum uric acid level was also elevated above the threshold for monosodium urate crystal deposition[4].

Bone wax is commonly used in skull base and mastoid surgery to control bleeding from emissary veins or the sigmoid sinus, or to seal off areas. There have been multiple reports of foreign body reactions to bone wax throughout the body. The pathogenesis of this reaction is unclear, with some suggesting that it is an allergic phenomenon[5]. Bone wax granuloma should be considered in any patient with a history of extensive mastoid or skull base surgery, characteristic T1 and T2 isointense but enhancing mass on MRI, and otalgia refractory to first line treatments.

Langerhans cell histiocytosis (LCH) encompasses a group of neoplastic processes including histiocytosis X, eosinophilic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease. These diseases are most common in children with an annual incidence of 5.4 per million. Temporal bone involvement by LCH is rare, but well described. Temporal bone lesions almost always occur in the setting of systemic disease. Therefore, a diagnosis of temporal LCH should prompt careful multisystem evaluation for further disease.[6] Chemotherapy and radiation are the mainstays of treatment, though there is a role for surgical resection in non-systemic lesions.

CONCLUSION

Conclusion: Difficult cases of otalgia or presumed otomastoiditis unresponsive to broad spectrum antibiotics require thorough radiographic evaluation. When the diagnosis remains uncertain, mastoid biopsy and histopathological analysis may provide important diagnostic information. Awareness of more rare etiologies is essential and can facilitate efficient diagnosis and treatment. To our knowledge this is the first report of acute inflammatory gout of the mastoid and second report of mastoiditis secondary to bone wax granuloma.

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