Juvenile Xanthogranuloma of the Temporal Bone in a Young Adult

Alex D. Sweeney, MD[1]; W. Marshall Guy, MD[1]; Marc E. Nader, MD[1]; Jeffrey T. Vrabec, MD[1]

1. Bobby R. Alford Department of Otolaryngology-Head and Neck Surgery, Baylor College of Medicine, Houston, Texas.

Abstract # 130

Abstract

Educational Objective: At the conclusion of this presentation, the participants should be able to understand the diagnosis and management of juvenile xanthogranuloma and understand how the management of juvenile xanthogranuloma can change based on its location and the associated symptoms.

Objectives: We present a rare case of juvenile xanthogranuloma (JXG) of the temporal bone to illustrate how the management of this disease process can be influenced by its location and the associated symptoms.

Study Design: Case report of a juvenile xanthogranuloma of the temporal bone.

Methods: We present a patient with a juvenile xanthogranuloma of the temporal bone evaluated by the otolaryngology - head and neck surgery service of an academic, tertiary care hospital in 2012 and 2013. The literature was also reviewed for similar cases through a directed PubMed search.

Results: A 20 year old patient presented to clinic with complaints of progressive hearing loss and vertigo over six months. A previous diagnosis of JXG had been given based on an open biopsy performed at an outside institution. Physical exam revealed a fleshy tumor obliterating the left ear canal. Imaging revealed extensive bony destruction in the left, lateral skull base with extension into the inner ear and posterior fossa. Immunohistochemical analysis confirmed the diagnosis of JXG.

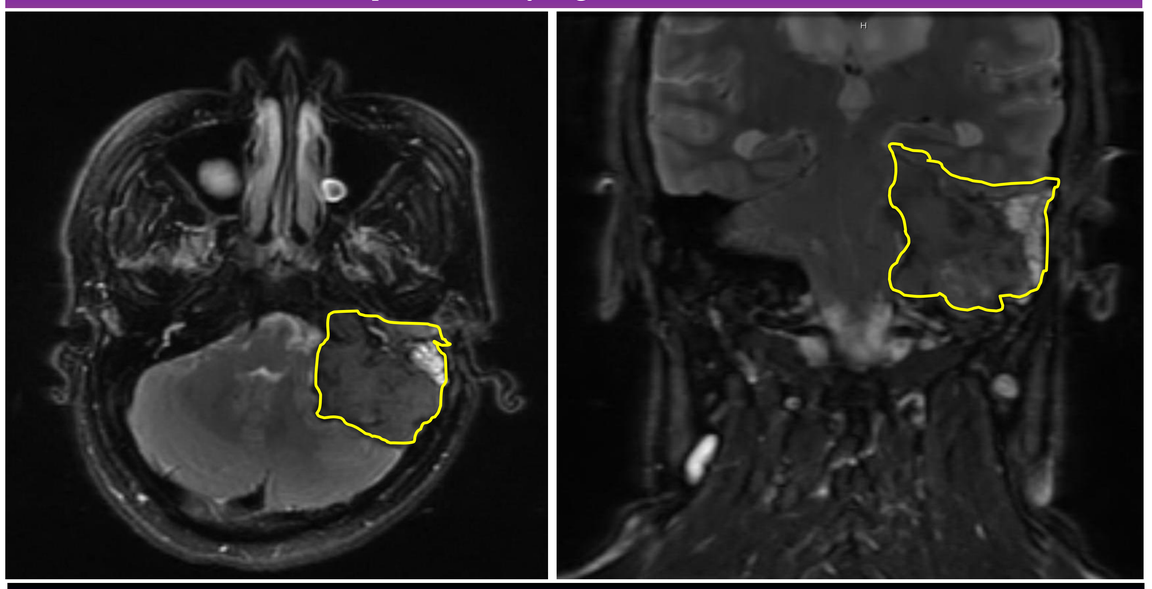
Conclusions: While JXG has been previously described in the temporal bone, this case represents the first reported presentation in an adult patient. Generally, JXG is a self-limited disease. However, temporal bone tumors can result in significant symptoms necessitating surgical management.

Background

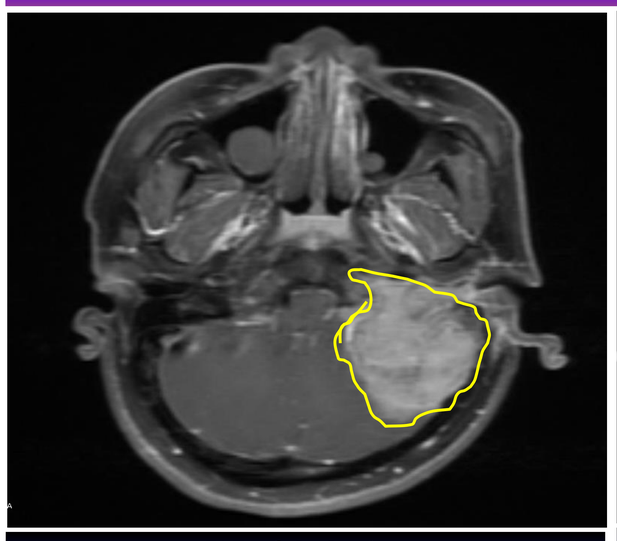
Juvenile xanthogranulomas (JXG) are a histiocytic disease of the non-Langerhans cell group.[1] They are generally benign cutaneous lesions of childhood that often involute spontaneously.[2] They rarely present in extracutaneous sites, and the eye is the most common of these locations.[3] Lesions in the liver, lungs, trigeminal nerve, nasal cavity, pancreas, genitalia, eyelid, toenail, palms, lips, groin, and soles of the feet have also been described.[2-6] The natural history of these lesions generally involves involution, even in the case of large, non-cutaneous JXG. In most cases, observation is the management strategy of choice unless functional complications arise.[2] With particular regard to symptomatic central nervous system JXG, surgical resection, chemotherapy and radiotherapy options have been reported.[2] Presentation in the temporal bone is rare and has been described in a 2 month old male, 2 year old male, and 2 year-old female. In these cases, treatment strategies have involved radical resections, stereotactic radiotherapy and surgical debulking, respectively.[7-9] We present a case of a young adult with an extensive, destructive JXG of the temporal bone and discuss our management strategy. To our knowledge, this case report represents the oldest patient presenting with an isolated, juvenile xanthogranuloma of the temporal bone.

Case Presentation

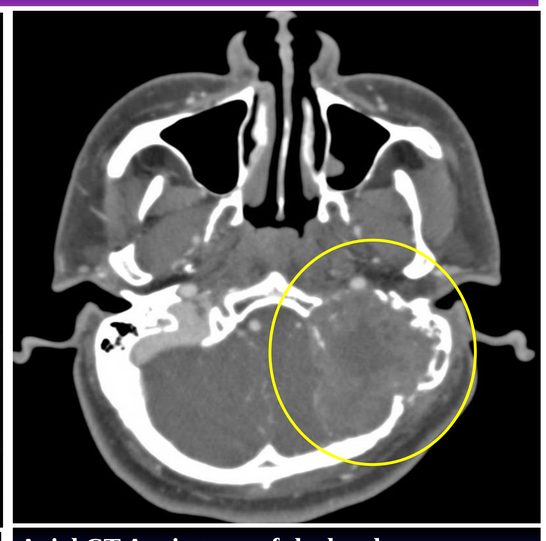
A 20 year-old male presented to our service with a 6 month history of headache, vertigo and progressive hearing loss with associated pulsatile tinnitus in his left ear, which was worse with neck flexion. The progression of these symptoms accelerated in the 3 months prior to his presentation. Initially presenting to an outside hospital, he underwent a transmastoid incisional biopsy, and pathology revealed a juvenile xanthogranuloma. No relevant family history was noted. On physical exam, a violaceous appearing tumor was noted to obliterate the external auditory canal. His post-auricular surgical site was healing well. Facial function and sensation were intact. Multidirectional spontaneous nystagmus was noted.



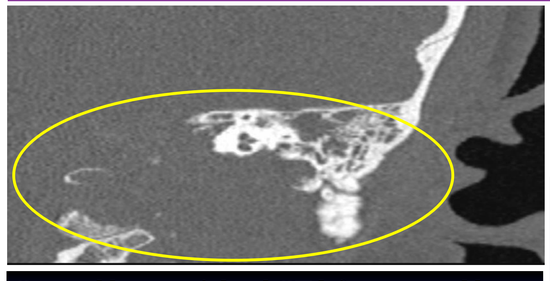
T2 post contrast axial (left) and coronal (right) magnetic resonance imaging (MRI) of the brain: Large isointense mass in the posterior cranial fossa with erosion of temporal bone and obliteration of the left transverse sinus, sigmoid sinus and jugular bulb. The yellow highlighted areas represent the soft tissue tumor component and post surgical changes in the mastoid cavity.



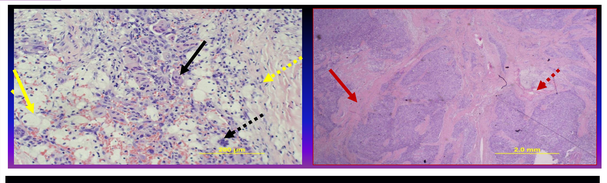
T1 post contrast MRI of the brain: Notice the hyperintense mass in the left posterior cranial fossa. The yellow highlighted areas represent the soft tissue tumor component.



Axial CT Angiogram of the head: Mass extending into the left carotid canal and obliterating the sigmoid sinus.



Coronal bone weighted CT left temporal bone: Heterogeneous density enhancing mass centered in the left posterior cranial fossa/skull base with prominent osseous and inner ear destruction.



The lesion consists of histiocytes with a minor sub-population of lipidized forms (yellow arrow), occasional multinucleated giant cells (black arrow), and pleomorphic cells (yellow dashed arrow) closely admixed with spindle shaped fibroblasts, thin ropy collagen (black dashed arrow), and larger thick collagen strands (red arrow) giving a nodular appearance with areas of bony infiltration (dashed red arrow).

Case Continued

After the pathologic diagnosis was confirmed as a juvenile xanthogranuloma, the patient was taken to the operating room for a planned, subtotal resection of the tumor, achieved through an extradural translabyrinthine, transparotid, transcervical approach including extratemporal facial nerve dissection. The subtotal approach was planned as complete removal would require sacrifice of the facial nerve and internal carotid artery. A primary closure was achieved after abdominal fat graft obliteration of the surgical defect and over closure of the ear canal.

The immediate post-operative course was relatively uncomplicated. His post-operative facial nerve function was intact. Aside from his expected post-operative deficits after a translabyrinthine surgery, he was neurologically intact. He was discharged from the hospital on post-operative day 6. In follow-up, he recovered quickly from his post-operative vestibulopathy to report marked improvement in his pre-operative vertigo. At 4 months post-operative, repeat imaging showed minimal growth of the residual tumor. However, the patient began to have persistent headaches again, and the patient is planned for a retrosigmoid debridement in conjunction with the neurosurgery service 10 months after the original surgery.

Discussion

Juvenile xanthogranulomas are generally benign, cutaneous lesions. Non-cutaneous lesions are also described, though intervention is reserved for lesions that cause functional impairment or cosmetic deformity due to the general trend for these lesions to involute spontaneously over time. [3] Our patient is the oldest reported case of a skull base JXG, and surgical intervention was pursued due to his rapidly progressing symptoms from an intracranial lesion. A subtotal surgical resection resulted in improvement of his vestibular symptoms but spontaneous regression of the remaining tumor has not been seen in this patient. Recurrence of headaches in the post-operative period has led to a decision for a second conservative resection through a retrosigmoid approach.

Conclusion

Generally, JXG is a self-limited disease with spontaneous regression described. However, destructive, intracranial temporal bone tumors can result in significant symptoms that may need to be addressed with surgical intervention

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