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Case Report

Case Report: Unusual Presentation of Neurofibroma in the External Auditory Canal

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Abstract

We present an unusual case of a neurofibroma located in the external auditory canal (EAC), which manifested with atypical symptoms and an unusual clinical course. Neurofibromas are benign tumors originating from nerve tissue, commonly associated with neurofibromatosis type 1 (NF1) [8,9]. However, when located in the EAC, they are rare and may present with symptoms that mimic other more common ear pathologies [3,8]. This case emphasizes the diagnostic challenges and highlights the need for thorough investigation in patients with atypical ear-related complaints.

Keywords: Neurofibroma; External Auditory Canal; Conductive Hearing Loss; Neurofibromatosis; Case Report; Tumor; Ear Mass; MRI; Surgical Excision

Introduction

Neurofibromas are benign, slow-growing tumors derived from Schwann cells and are most commonly seen in patients with neurofibromatosis type 1 (NF1) [9,10]. The external auditory canal (EAC) is an uncommon site for these tumors [1,3]. When neurofibromas do occur in this region, they are typically asymptomatic and present with symptoms such as hearing loss or otalgia [3,6]. However, their appearance can be misleading, and they may mimic other more common conditions like cerumen impaction or otitis media [3,8]. In this case, the patient presented with a rapidly growing mass in the ear canal and neurological symptoms, making the diagnosis challenging.

Case Presentation

A 52-year-old female presented with progressive hearing loss, intermittent ear pain, and a sensation of fullness in the right ear for two months. There was no family history of neurofibromatosis.

Examination revealed a firm, rubbery, non-tender mass in the right EAC near the tympanic membrane. Audiometry showed a moderate conductive hearing loss. Imaging (CT and MRI) revealed a well-defined mass within the EAC consistent with neurofibroma [3,8].

The swelling was excised via a postauricular approach. The mass was adherent to the canal skin and removed completely. Histopathological examination confirmed neurofibroma, showing Schwann cells with wavy nuclei and fibroblasts in a collagenous stroma [5,7]. The patient recovered uneventfully.

Clinical findings

Physical examination

Normal outer ear, with no visible deformity or signs of infection. The EAC showed a non-tender, firm, well-demarcated mass that partially obstructed the canal. No swelling or tenderness in the surrounding soft tissue. The tympanic membrane appeared normal without signs of inflammation.

Audiology

Pure tone audiometry showed a moderate conductive hearing loss in the right ear, with air-bone gaps indicative of obstruction or middle ear involvement.

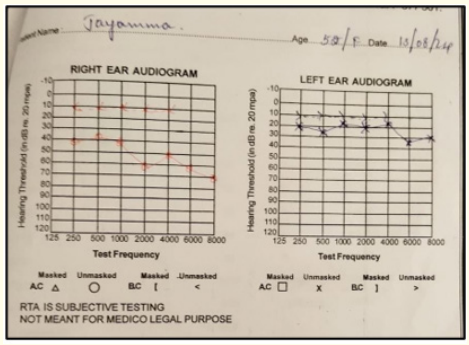

Figure 1: Pre Operative PTA.

Imaging

CT scan of the temporal bone

A CT scan revealed a well-defined, soft tissue mass within the EAC without bony involvement. The mass was causing partial obstruction of the canal.

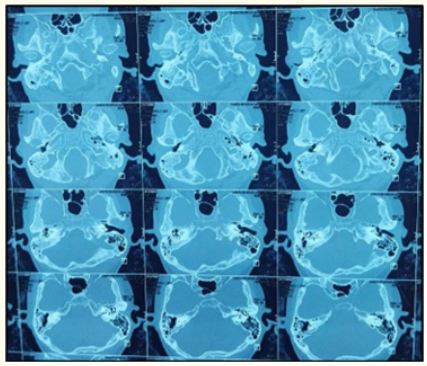

Figure 2: HRCT Temporal Bone Axial View.

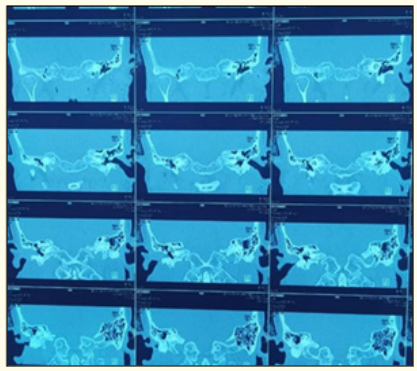

Figure 3: HRCT Temporal Bone Coronal View.

MRI

An MRI of the head showed a well-enhancing, heterogeneously hyperintense mass on T2-weighted images within the EAC, which was consistent with a neurofibroma. The mass did not show signs of malignant transformation but was encroaching close to the facial nerve, which raised concerns about possible nerve involvement.

Procedure

The swelling was excised by postauricular approach under local anesthesia. The mass was adherent to the skin of the meatus and ear canal. After removing the entire mass was removed along with the excess skin which was adherent to it. Mass was sent for Histopathological examination of the mass showed it to be neurofibroma. The patient made an uneventful recovery, and a good cosmetic result was achieved.

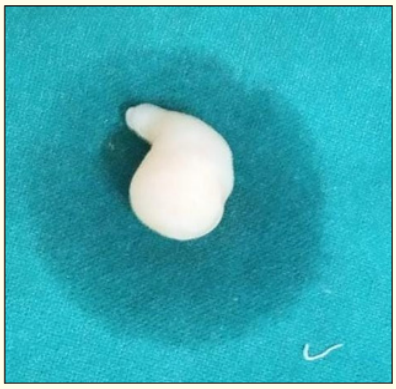

Figure 4: Excised Tumor Mass.

Histopathology

The lesion composed of well-defined hypocellular lesion composed of schwann cells with wavy nuclei, fibroblasts with interspersed collagenous stroma. No significant cellular atypia was seen. Mitotic activity is not appreciated. The patient made an uneventful recovery, and a good cosmetic result was achieved.

Diagnosis

The imaging findings and clinical presentation were suggestive of a neurofibroma. Given the location and growth pattern, it was unusual for a neurofibroma to present so aggressively in the EAC without accompanying symptoms of neurofibromatosis or other signs of systemic involvement. No other masses or signs of systemic neurofibromatosis were detected upon further workup. The histological examination following biopsy confirmed the diagnosis of a benign neurofibroma. The tumor consisted of a mixture of Schwann cells, fibroblasts, and collagen fibers, consistent with the typical appearance of neurofibromas.

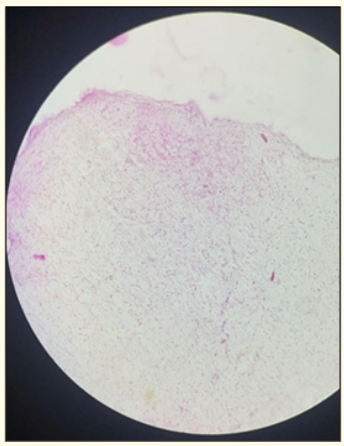

Figure 5: HPE Under 30X.

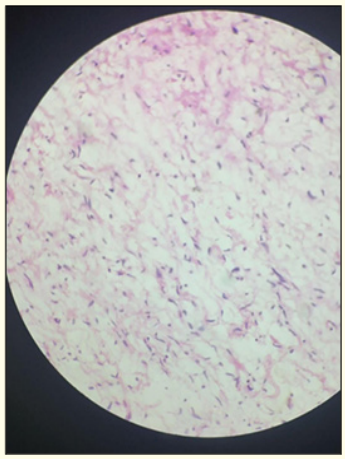

Figure 6: HPE Under 100X.

Treatment and outcome

The patient underwent surgical excision of the neurofibroma via a post aural approach. Intraoperatively, the tumor was found to be well-circumscribed and did not invade the surrounding bone or soft tissues. The facial nerve was carefully preserved during the procedure.

Postoperatively, the patient had complete resolution of her hearing loss and ear fullness, and there were no signs of recurrence at the 6-month follow-up. The histopathological diagnosis confirmed the benign nature of the tumor.

Discussion

Neurofibromas arising in the EAC are rare, with only a few cases reported in the literature [1,3,6,8]. They may present with vague symptoms such as hearing loss or otalgia, and occasionally with neurological involvement if adjacent nerves are affected [6,8].

The clinical course in this patient was unusual due to rapid tumor growth and significant symptoms, which raised suspicion for malignancy. Similar cases have been described by Stevenson et al. and Trevisani et al., emphasizing the diagnostic challenge of such lesions [1,6].

Imaging—particularly MRI—is crucial in differentiating neurofibromas from other EAC masses such as keratosis obturans or cholesteatoma [8]. In this case, MRI findings were typical of benign neurofibroma, showing a well-enhancing, T2-hyperintense lesion without bone erosion.

Neurofibromas may occur as isolated lesions or as part of NF1 or NF2 syndromes [9,10]. Isolated lesions, such as in this patient, usually have a good prognosis after complete excision [5,9].

Conclusion

Neurofibroma of the external auditory canal is a rare entity that should be considered in the differential diagnosis of patients presenting with ear masses and conductive hearing loss [3,8]. MRI aids in diagnosis, and surgical excision remains the mainstay of treatment [6,8]. Early identification and management lead to excellent outcomes with minimal recurrence risk [5,9].

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