

Neurofibroma of the Middle Ear - A Rare Case Report

Apurva Sharma, Swapnil Gosavi

Department of Otorhinolaryngology, MGM Hospital, Navi Mumbai
Corresponding Author Swapnil Gosavi

Abstract: Neurofibromatosis consists of two distinct disease that share several findings. Neurofibromatosis 1 (NF1), or peripheral neurofibromatosis, has been determined by a gene on chromosome 17. The incidence is thought to be 1 in 2500 to 3000 live birth. Neurofibromatosis 2 (NF2), or central neurofibromatosis, has been shown to be related to a gene found on chromosome 22. It is far less common than NF1, with an incidence believed to be about 1 in 50,000. A neurofibroma of the ear may occur either as an isolated lesion or as a manifestation of von Recklinghausen' disease. Solitary neurofibroma is common benign peripheral nerve sheath tumors and occurs in patients without neurofibromatosis type 1. The most common otologic findings in NF2 is acoustic neuroma, seen exclusively in patients with NF-2. Here we present the extremely rare case of middle ear neurofibroma with incidental histopathological finding in 34 years female patient who presented with the complaint of right ear pain, with subtotal perforation.

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I. Introduction

Neurofibromatosis (NF) is one of the most common genetic disorders transmitted in an autosomal dominant fashion affecting more than 1 out of 3,000¹ individuals caused due to deletions, insertion, or mutations in neurofibromatosis type 1 (NF) gene located in the pericentric region on chromosome 17.² Extracranial solitary neurofibromas, particularly, in the ear are rare. It represents 5% of all benign soft tissue neoplasm. Although the head and neck region are a common location for benign peripheral nerve sheath tumors. Macroscopically, the tumor appears as a solid, tan-gray, glistening mass that is translucent to light. Its growth pattern is either a well-bordered intraneural enlargement or infiltration into surrounding tissue. From a clinicopathological perspective neurofibromas are classified into some subtypes: localized cutaneous form or localized intraneural form, plexiform subtype, and diffuse and massive subtypes.³ Microscopic the main architecture consists of a mucopolysaccharide-rich matrix, where spindle cells with normochromatic nuclei dominate.⁴

II. Case Report

A 34 years old year's female presented with a right ear pain and hearing loss. Ear pain was insidious gradually progressive intense pain. Hearing loss was also insidious progressive, not able to hear normal conversation. There was no history of ear discharge. There was no past history of otitis externa, otitis media or trauma and tinnitus. There was no history of headache, vertigo, vomiting and fever or any complaint from nose and throat.

On examination of the right ear, there was subtotal perforation of tympanic membrane. The pinna, postauricular region, external auditory canal and the mastoid area were normal non tender. On the left ear tympanic membrane was intact. On tuning fork tests, Rinne test was negative in the right ear and positive in the left ear. Weber was lateralized to the right ear, Absolute bone conduction test was normal thus conductive hearing loss in the right ear. PTA suggestive of moderate conductive hearing loss, x-ray both mastoids (lateral oblique view) showed sclerotic mastoid air cells on the right side. There was no obvious mass seen in otomicroscopic examination.

Patient was taken for right tympanoplasty under anaesthesia. Postauricular Wildes incision was taken, temporalis fascia harvested, posterior meatotomy done. Subtotal perforation visualised, margins freshened, tympanomeatal flap elevated along with annulus. Middle ear were filled with granulation tissue bleeding on touch and sent for histopathological examination. ossicular chain was mobile and intact, , temporalis fascia placed medial to handle of malleus, flap repositioned. After getting histopathology report which was suggestive of neurofibroma of middle ear, patient was advised HRCT temporal bone showed a right large cavity in mastoid containing soft tissue attenuation which is in communication with subcutaneous tissue in the region of pinna, erosion of dural plate anteriorly.

Hence the patient was planned for Modified Radical Mastoidectomy. Intraoperatively cortical maatoidectomy was done. Polypoidal mucosa and granulation tissue was seen in aditus et antrum, attic sinodural

angle. Hence we proceeded with Canal Wall Down Mastoidectomy Polypoidal mucosa over promontory, aditus et antrum, dural plate removed meticulously. Patient has been following up since 3 months and at her last follow up she was symptom free.

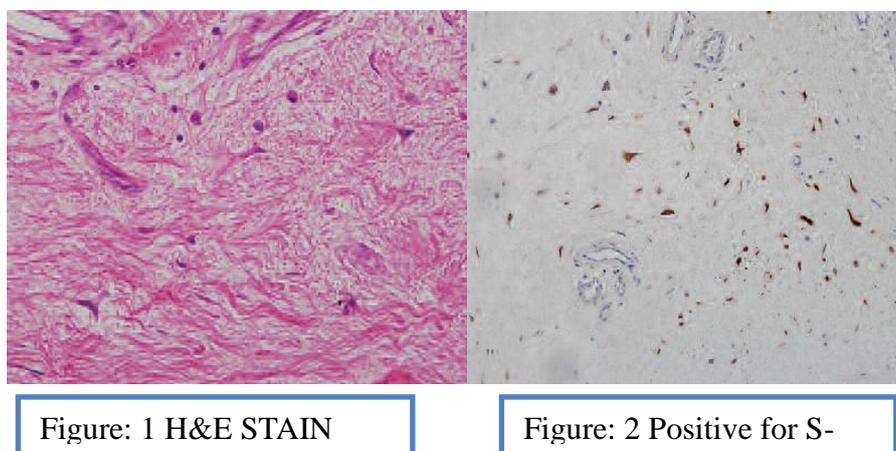


Fig. 1: Proliferation of Interlacing bundles of spindle cells arranged as haphazard manner within an extensive myxoid stroma with scattered mast cells and blood vessels with wavy dark staining nuclei in a collagenous stroma (X20 magnifications) Also showing inflammatory cell infiltrate comprising predominantly of mast cells and few neurophils, and **Fig 2:** Immunohistochemistry studies **POSITIVE FOR S- 100** (x20 magnification).

III. Discussion

Peripheral nerve sheath tumours can be divided into benign and malignant. The two major benign categories are neurofibroma and schwannoma while malignant form is malignant peripheral nerve sheath tumor.⁵ Each benign category can be a part of neurofibromatosis type 1 or von Recklinghausen's syndrome which is characterized by café au lait skin patches, lisch's nodules, axillary freckling and fibroma molluscum but localised neurofibroma usually not associated with neurofibromatosis. Neurofibroma is one of the most common inherited disorders and can affect anyone, regardless of family history, race gender, or ethnic background. These locally destructive tumors arise along the distribution of peripheral and cranial nerves. It may involve all layers of soft tissue, invade neighboring bone. Neurofibroma is a tumor derived from Schwann cells, fibroblasts and perineural cells. Neurofibroma infrequently occurs as an isolated lesion. Neurofibroma is locally destructive lesions that may undergo malignant degeneration. 2-16% of neurofibroma undergoes malignant change resulting in neurosarcomas.⁶

Differential diagnosis for neurofibroma are the following-

1. chronic suppurative otitis media with mastoiditis and cholesteatoma
2. chronic proliferative tuberculosis
3. Malignant growth of mastoid and middle ear
4. Acute otomastoiditis
5. Facial nerve schwannoma

High resolved computed tomography scanning is helpful in evaluating neurofibromas of the ear, showing the location and extent of the tumor. Infiltration of adjacent bone soft tissue can be assessed by CT scanning. The treatment is total wide excision of tumour to minimise the possibility of recurrence and attempt to restore the hearing of patient.

IV. Conclusion

Neurofibromatosis (NF) consists of two distinct neurocutaneous diseases, both of which can show otologic manifestations. The most common finding in NF-1 IS NEUROFIBROMA of middle ear and the external ear. Next most common finding in the ear is acoustic neuroma, seen exclusively in NF-2. Solitary neurofibromas particularly, in the ear is rare. Diagnosis is based on clinical symptoms, CT, magnetic resonance imaging, and surgical biopsy with histopathological findings. Hence we conclude that intraoperative any abnormal tissue in the middle ear or aditus ad antrum should be sent for histopathological examination.

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