



## Primary Ectopic Atypical Meningioma of the Middle Ear: A Case Report

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### Abstract

Meningiomas are the second most common primary tumors of the Central Nervous System. Most cases occur in intracranial sites; but in very rare cases some do occur in extracranial regions. Ectopic meningiomas have no communication with intracranial meninges and misdiagnosis of this pathology may be encountered due to its anatomical complexity. The report below is one of the few uncommon documented cases of Primary Ectopic Atypical Meningioma of the middle ear which should increase the awareness of otolaryngologists over difficulties arising to diagnose this rare phenomenon.

### Introduction

Meningiomas are tumors/neoplasms that arise from central nervous meninges- the arachnoidal (meningothelial) cap cells [1]. This common type of tumor accounts for around 18% to 20% of all intracranial lesions whereas less than 2% account for extracranial meningiomas [2,3]. These extracranial meningiomas, based on their origins, can be classified into:

- a) Primary/ ectopic – occurring from the ectopic site or
- b) Secondary- usually extensions from intracranial masses [4,5].

Reported cases of ectopic meningiomas arise from skull bone, paranasal sinus, parotid gland, middle ear, neck and skin.

Meningiomas are more common in females than males with overall incidence of (2:1) due to estrogen or progesterone levels in women [5]. Most cases of meningiomas are usually benign in nature. However, malignant transformation also occurs. According to the World Health Organization Classification, meningiomas are categorized as follows:

- Grade 1- Benign. (80% )
- Grade 2- Atypical. (15% to 20%)
- Grade 3- Anaplastic. (1% to 3%)

Benign meningiomas grow slowly and have 5% recurrence in 5 years whereas atypical ones grow quicker with 40% recurrence in 5 years. Anaplastic meningiomas are the worst type and have an 80% recurrence rate [3]. Benign lesions that are small (<2.5 cm), slow growing and asymptomatic require close observation. Surgery may not be needed in such cases but in atypical cases, surgical excision is the mainstay of management. Adjuvant therapy- radiation can also be considered in some cases. Anaplastic meningiomas require both surgery and radiation therapy.

### Case Presentation

A 46 year old female patient presented to the otology outpatient department of The First Affiliated Hospital of Zhengzhou University with chief complaint of recurrent bilateral ear discharge and hearing loss for past 30 years. A brief history revealed that she had tinnitus occasionally but denied having otorrhea or any other neurological deficits. She attended local clinics for the above symptoms whereby “ear drops” were given to her. While these improved her symptoms, she unfortunately had repeated episodes over time which led to gradual increase in hearing loss and dizziness. In our hospital, video otoscopy, Pure Tone Audiometry, Tympanometry and Temporal Bone CT scan were ordered. Subsequently an initial diagnosis of Chronic Suppurative Otitis Media in bilateral ears was made.

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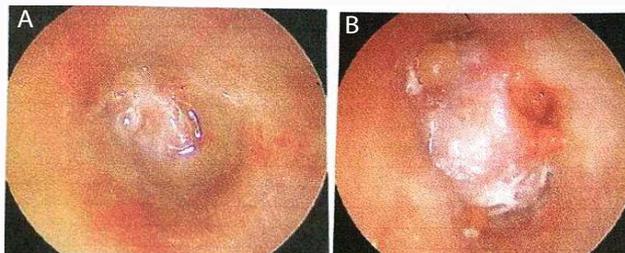
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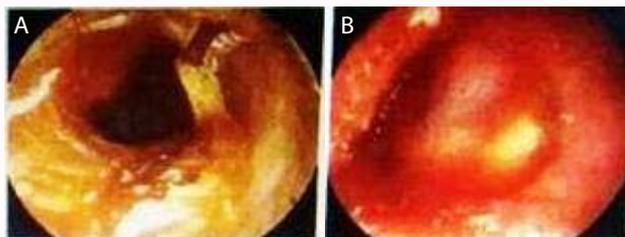
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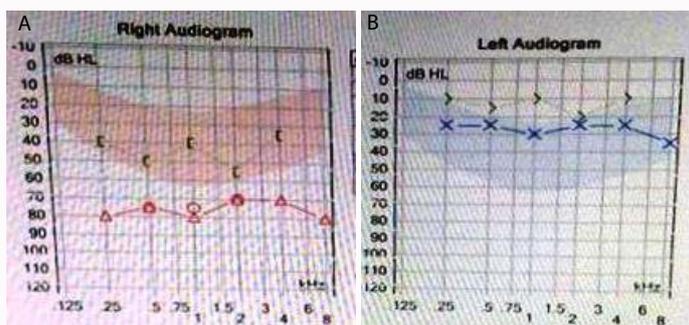
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**Figure 1:** A) Yellowish purulent effusion was observed in right ear B) Following cleaning of the ear canal, the tympanic membrane could be clearly visualized.



**Figure 2:** A) Illustrates Left ear filled with cerumen and purulent discharge B) After cleaning, an abnormal tympanic membrane and a swollen ear canal was observed.



**Figure 3:** A) Audiometry in Right Ear revealed Moderate to Severe Mixed Hearing Loss B) Left Ear was consistent with Mild Conductive Hearing Loss.

### Video otoscopy

As illustrated in (Figure 1A, 1B), yellowish purulent effusion was observed in right ear. Following cleaning of the ear canal, the tympanic membrane could be clearly visualized. Tympanic membrane was abnormal and handle of malleus could not be seen.

### Pure tone audiometry

Audiometry in right ear revealed moderate to severe mixed hearing loss whereas left ear was consistent with mild conductive hearing loss (Figure 3A, 3B).

### Tympanometry

Type B tympanogram was noted in both ears - suggesting minimum or no movement of the tympanic membranes; possibly due to presence of fluid or a mass in middle ear (Figure 4A, 4B).

### Temporal bone CT scan

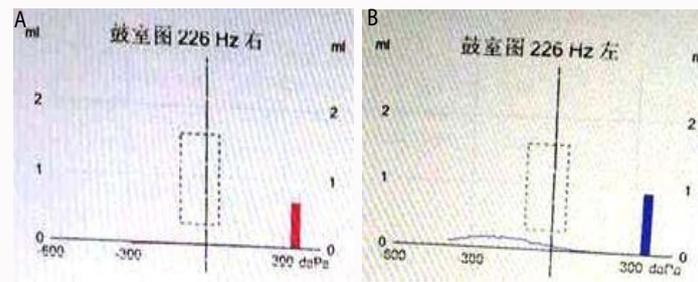
Assessing the axial view of Temporal bone CT scan, no deformity was found in both auricles and the external auditory canals were unobstructed. Mastoid air cells and the middle ear cavity in the right ear had increased density. The ossicles were normal. In left ear, the mastoid air cells were well pneumatized and left middle ear cavity as well as ossicles was normal. Cochlea and Semicircular canals of bilateral ears were normal. No dilatation of Internal Auditory Canal

on both sides (Figures 5A-5C). Consequently, Right Canal Wall down Mastoidectomy + Ossicular Chain Reconstruction + Tympanoplasty was done on the patient. During surgery, the following findings were noted:

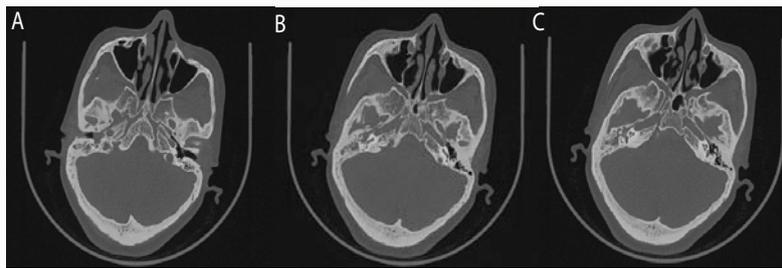
1. While drilling the mastoid bone, dark brown-like secretions accumulated in the mastoid air cells were seen and granulations in the middle ear cavity.
2. The granules had invaginated the pars-flaccida + the pars tensa was perforated
3. Incus was destroyed as well as part of the malleus.
4. Stapes was intact.
5. Eustachian tube was unobstructed.

### Discussion

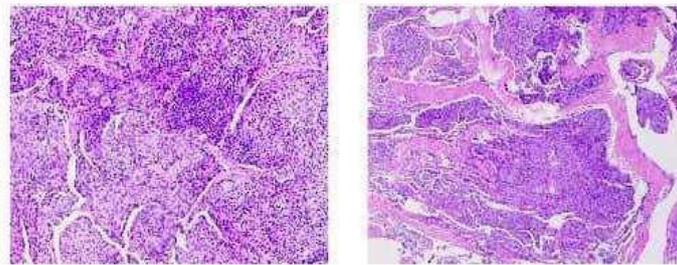
Up to now, very few cases of meningiomas extending to the temporal bone and middle ear cavity have been reported. These were mostly secondary extracranial conventional type meningiomas extending from an intracranial lesion to the temporal bone or middle ear [6]. Differential diagnosis of meningiomas in middle ear is: schwannomas, paragangliomas, middle ear adenomas, metastatic carcinomas, cholesteatomas, glomus tumors, retrotympanic



**Figure 4: (A and B)** Tybe B tympanogram was noted in both ears - suggesting minimum or no movement of the tympanic membranes; possibly due to presence of fluid or a mass in middle ear.



**Figure 5:** Temporal Bone CT SCAN.



**病理诊断:** (右中耳)梭形细胞肿瘤, 结合形态及免疫学标记符合脑膜瘤。  
 免疫组化结果: EMA (+), Vimentin (+), PR (+), GFAP (-),  
 S-100 (-), SMA (-), AE1/AE3 (CK) (-), CD34 (血管+),  
 CD56 (56C04) (±), SYN (-), Ki-67 (约2%+)。

**Figure 6:** Specimen was sent for histopathology examination which later confirmed the diagnosis of Primary Atypical Meningioma of Middle Ear.

vascular tumors, and lipoma. Other than hearing loss (conductive, sensorineural or mixed hearing loss), which is the most common symptom [7], the following symptoms are also associated with middle ear meningiomas: headaches, dizziness, vertigo, tinnitus, otalgia, bleeding, “metallic taste,” and sometimes chronic cough. The severity of symptoms usually depends on location of tumors. More severe cases can present with epilepsy, speech disturbances, cognitive impairment and visual disturbances. It has been reported that about 16% of Middle ear Meningiomas are usually accompanied by Chronic Otitis Media. One theory behind this is that there is the possibility of the tumor blocking the Eustachian Tube which prevents the drainage of Middle Ear fluid [8]. Causes of meningiomas include exposure to ionizing radiation, neurofibromatosis type 2- a genetic disorder and some risk factors associated are trauma to head, use of cell phones and also increased level of estrogen and/or progesterone, which can explain the high incidence of the disease in women compared to men [9]. Radiological Imaging like CT and MRI are very helpful in

differentiating middle ear meningiomas to other middle ear diseases. They give an idea of the extent of tumor, whether there was absence of ossicles or ossicular erosion and details of internal auditory canal and associated structures [6]. As observed in this case, the patient was initially diagnosed as bilateral Chronic Suppurative Otitis Media but it is only after surgery, when histopathological report was obtained that the definite diagnosis of Meningioma was confirmed. Since conventional meningiomas have distinctive histological findings, it is easy to diagnose them. But for rare types, as in the present case, ectopic sites meningiomas can pose as a diagnostic challenge for pathologists. Some may argue that biopsy of tympanic/middle ear lesion during myringotomy may lead to earlier diagnosis but the risk of damaging any vascular lesion or the facial nerve is high.

Treatment of choice for atypical meningiomas remains surgical excision. Where complete removal of the meningioma is not possible, radiotherapy is often warranted. It is also recommended that long

term follow up of patients be done together with a neurologist to exclude recurrence. Patients and their family members also need to be well educated about the prognosis of this disease and informed to not ignore any associated symptoms.

## Conclusion

Despite its rarity in otolaryngology, Primary Atypical Meningioma in middle ear is very likely to be misdiagnosed. Conventional tests such as Pure Tone Audiometry, Tympanometry and CT scans are helpful but not enough for diagnosis of this disease. So far, histopathological examination has been the only method to confirm the nature of the middle ear lesion. Once confirmed, it is imperative to follow up the patient closely and regularly with serial imaging for the rest of her life as the rate of recurrence is high. Since primary meningiomas are very rare, care must be taken to differentiate between intracranial or extracranial lesions as well as benign and malignant tumors.

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