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Case Report

# A Case Report of a CPA Meningioma Presenting as Middle Ear Mass With Middle Ear Effusion

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Middle ear (ME) benign tumors are rare, and among them is meningioma. An ME meningioma might be isolated or merely a lateral extension of a CPA meningioma. We report a case with presentation of ME effusion followed by the appearance of an aural polyp after repeated myringotomies. Computed tomography (CT) revealed a benign-looking ME and mastoid mass. After debulking and biopsy, it turned out to be a meningioma. However, when MRI was performed, a large CPA meningioma was detected. ME masses are rare; however, they might be encountered, and CT must be performed followed by biopsy or total removal. In case of detection of a tumor with probable intracranial connection as meningioma, an MRI should be performed to exclude intracranial extension.

**KEYWORDS:** Middle ear meningioma, CPA meningioma, mastoid meningioma

## INTRODUCTION

Meningioma is a relatively common benign intracranial tumor. On the other hand, its presence in the middle ear (ME) and mastoid cavity is rare and represents around 1% of temporal bone tumors.[1,2] It is rarely malignant. It is usually the result of lateral spread of an intracranial meningioma. However, it can arise even more rarely with no intracranial origin, primarily from the ME.[3] In the literature, it is mainly published in case reports or in small case series.[3] Publications of isolated primary middle ear meningioma (MEM) before the era of widespread MRI have been criticized by many to miss intracranial tumors.[1,4,5] There are many theories in the literature to explain isolated extracranial MEM. For instance, maturation of pluripotent cells, or sequestrated arachnoid cells resting along bony fusion lines,[4,6] or intracranial hypertension or trauma misplacing arachnoid cells into the ME.[2]

Usually, meningioma is more common in females and has been reported in association with other female malignancies.[7,8] The explanation could the presence of estrogen and progesterone receptors on the tumor surface.[8]

## CASE PRESENTATION

A written consent was taken from the patient for reporting her case and institutional board review was obtained. This was a 35-year-old female patient who complained of left-sided pulsatile tinnitus and hearing loss since April 2016. There was no history of vertigo, otorrhea, or otalgia.

She neglected her complaints and sought medical help only 2 years later. In April 2018, pure-tone audiometry was performed and the result showed a mild conductive hearing loss with an air bone gap (ABG) of 20 dB with speech discrimination score of a 100%, and a type B tympanometry with absent acoustic reflexes on the left side. Otoscopy revealed a dull tympanic membrane. The other side was normal. Nasopharyngoscopy was performed and was free.

The patient was managed as ME effusion and received medical treatment. However, her condition did not improve.

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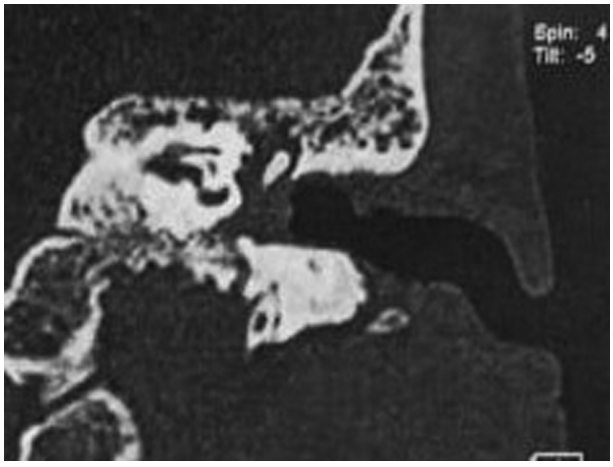


**Figure 1.** Preoperative CT coronal view of the left temporal bone on bone window showing middle ear mass.

In September 2018, left-side myringotomy and grommet tube insertion were decided. Four months later, the tube was extruded with no improvement of symptoms. The patient then skipped a few months of follow-up. She then presented to our department in August 2019, with the same complaints and audiological findings. Consequently, myringotomy was again attempted. Intra-operatively, granulation tissue was found in the ME cavity, and a biopsy was done, which revealed only nonspecific inflammation.

In October 2019, the patient still complained of persisting tinnitus, hearing loss, and additionally, non-positional dizziness and left-sided headache. A new audiological assessment was performed and revealed a modest increase in ABG (25 dB). Otoscopic examination revealed a dull, bluish, intact tympanic membrane with a large central bulge, seemingly resulting from an ME polyp. All cranial nerve functions including the facial nerve were normal.

Thin-cut multi-slice computed tomography (MSCT) was ordered (Figure 1). It revealed a nonspecific soft tissue mass occupying the ME and mastoid cavity with intact ossicular chain and dural and sigmoid sinus plates, and a free jugular foramen with no suspicion of intracranial lesion. Neither sclerosis nor hyperostosis were found.

In December 2019, a decision was made to perform an exploratory tympanotomy to obtain a better specimen for biopsy. This was performed via a post auricular approach. A polyp-like ME swelling bulging through an intact tympanic membrane was found. On entering the ME, a reddish firm soft tissue mass, filling the whole ME and spreading toward the mastoid antrum and cavity was found (Figure 2). The decision was made to perform maximum debulking of visible tumor using an intact canal wall mastoidectomy, posterior atticotomy, and posterior tympanotomy. The mass had occupied a significant portion of the mastoid cavity. The tumor was bloody but not as severely as usually encountered with paraganglioma cases. The ossicular chain was intact; however, after tumor dissection, there was an incudostapedial (IS) separation repaired by bone cement, and a perforation in the TM in an adherent part of the tumor which could not be dissected and was repaired by an underlay temporalis fascia graft. The tumor was grossly totally removed and sent for histopathological analysis. Neither could an obvious site of origin be detected, nor could and evidence of intracranial connection be found.

Post-operatively, the facial nerve was HB I as before surgery, graft was intact, and tinnitus had improved. Bone conduction was preserved. Unfortunately, an ABG persisted, which suggests failure of bone cement to fix the IS joint.

The biopsy revealed a meningothelial meningioma WHO grade 1. It was confirmed via immuno-histochemistry, namely epithelial membrane antigen (EMA), progesterone receptor, and vimentin (Figure 3). Consequently, an MRI of the cerebello-pontine angle was ordered to assess residual tumor and/or intracranial extension. A $3 \times 2.5 \times 1.3$ cm tumor was found occupying the cerebellopontine angle with a classical dural tail, highly suggestive of meningioma (Figure 4). Consequently, the patient was rescheduled for a removal via a transotic approach.

## DISCUSSION

Meningioma of the ME and mastoid bone are rare entities.[9] Only 2% of meningiomas occur extracranially.[8] In response to critics of the existence of isolated extracranial meningioma in the literature,[1,4,5] we performed an MRI for our patient, and a large CPA meningioma was actually found.

Meningioma has been reported as an isolated lesion in both mastoid[6] and ME.[1,3] In the present studied tumor, it was found in both cavities. It was a meningothelial type, WHO grade I, which is reportedly the most common type.[4] The patient in our study is typical

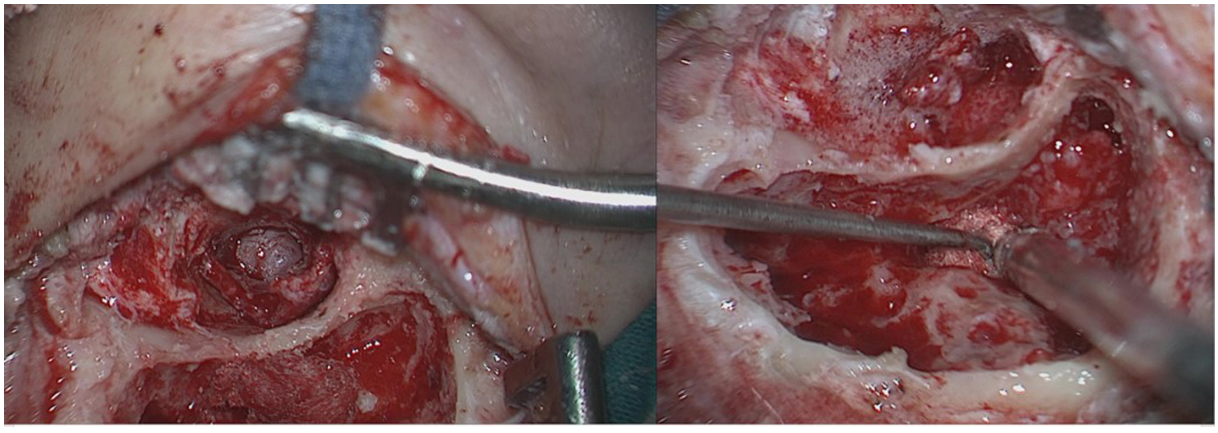

**Figure 2.** Left: Middle ear polyp bulging through left intact tympanic membrane. Right: A part of the mass filling the left mastoid antrum.

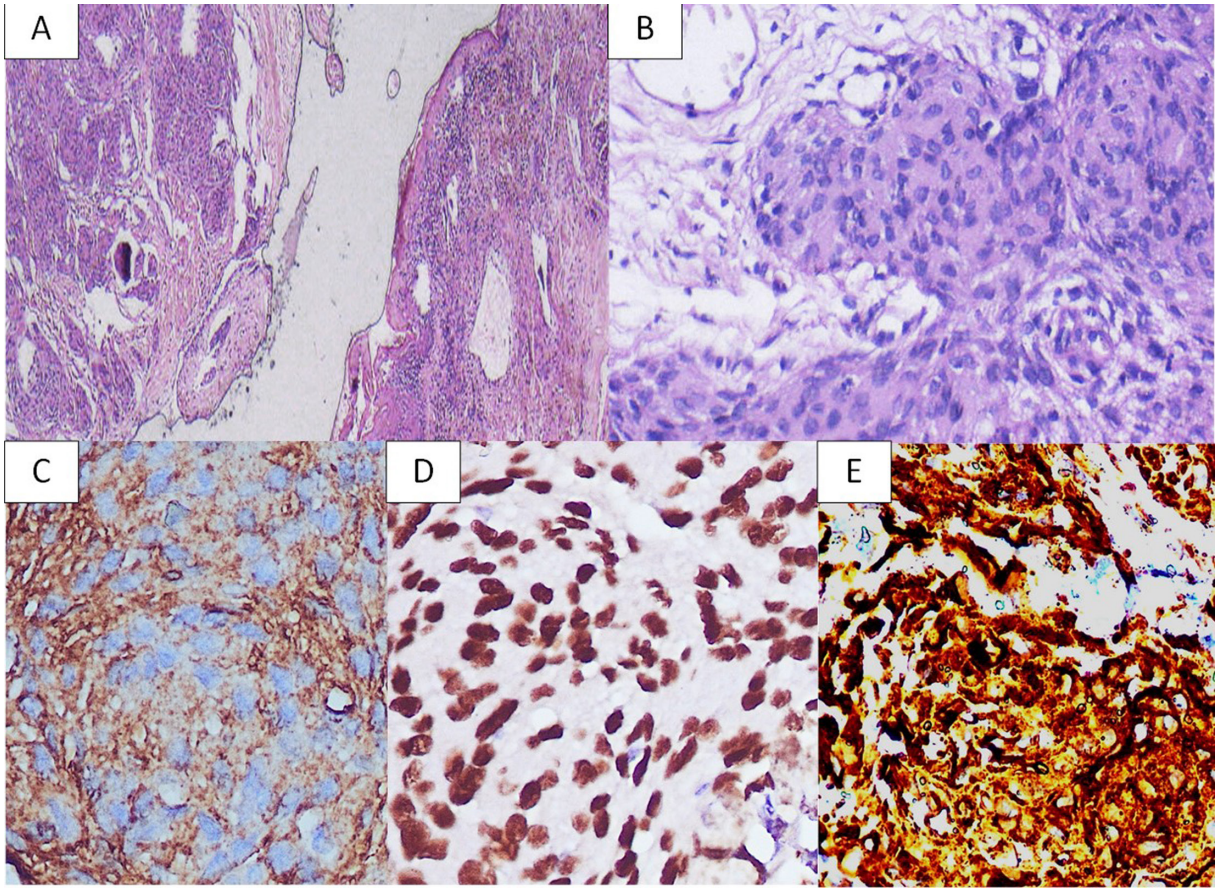


**Figure 3.** Tissue fragments from the middle ear and mastoid cavity showing meningothelial meningioma formed of masses and whorls (A, H&E, ×40). The masses are formed of syncytial cells with indistinct cell membrane, round uniform nuclei, and eosinophilic cytoplasm. Neither atypical nor anaplastic features are detected (B, H&E, ×400). The tumor cells show diffuse strong membranous staining for epithelial membrane antigen (EMA) (C, ×400), diffuse and strong nuclear staining for progesterone receptor (PR) (D, ×400), and diffuse strong cytoplasmic staining for vimentin (E, ×400).

epidemiologically. Since, meningiomas are reported to be more common in females with a mean age in the fourth decade.[7] However, a considerable CPA connection was found on imaging.

Therefore, benign tumors of the ME should be considered when dealing with ME polyps,[4] persistent ME effusion,[10] or when otoscopic findings are unusual.[8] MSCT should be ordered in such situations. In case the biopsy turns out to be a meningioma, immunohistochemistry is needed for confirmation. Consequently, an intracranial connection must be excluded using MRI,[6,7] since 20% of intracranial meningiomas have an extracranial spread.[1,4] The optimum treatment for most MEM would be total gross excision.[5] This is different from tumors with intracranial extension in which subtotal or near total resection to preserve nerve function or avoid major vessels and radiotherapy are occasionally considered as valid options in some patients.[8,10]

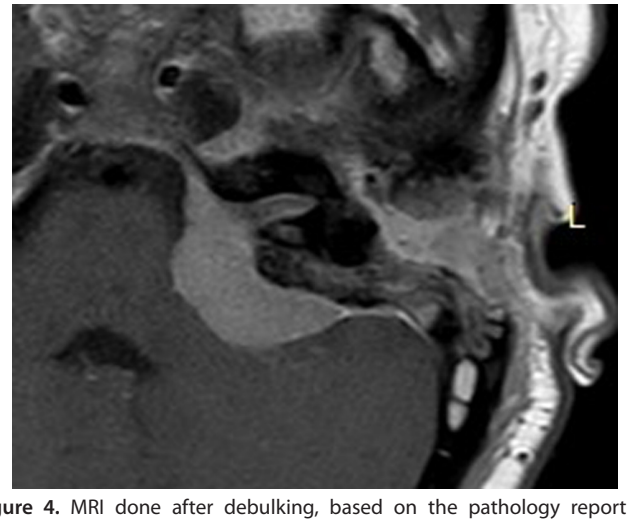

**Figure 4.** MRI done after debulking, based on the pathology report of meningioma. Axial T1 showing a left CPA mass with a classical dural tail, suggestive of meningioma.

In our center, MRI is not routinely performed for tumors of the ME with no apparent intracranial extension on computed tomography (CT) and no jugular foramen changes or clinical suspicion as cranial nerve palsies. Surgery would be indicated with the intent of complete tumor removal. The extent of the approach would be dictated by hearing level in both ears, fitness of the patient, and the extent of the tumor. In case of any suspicion of intracranial extension (clinically or CT), MRI is ordered to rule out jugular foramen or intracranial extension. In this particular case, the major part of the considerable delay in diagnosis was because the CPA component was asymptomatic. Moreover, the ME component was obscured both clinically and radiologically by the ME effusion. Based on this, the patient was treated for few months and grommets inserted repeatedly. The start of suspicion of tumor started only with the accidental discovery of

a polyp-like lesion when inserting myringotomy tubes, which led to the decision of biopsy and imaging.

## CONCLUSION

CPA meningioma may rarely present as ME effusion or ME mass or both. Biopsy and imaging in case of a polyp and/or resistant ME effusion is mandatory. MRI should be performed in tumors with suspicious intracranial origin, especially meningiomas.

**Informed Consent:** Written informed consent was obtained from all participants who participated in this study.

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