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Case Report

Middle ear neuroendocrine tumour-case report of a rare tumour of the temporal bone

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ABSTRACT

Several conditions are known to present as masses in the middle ear medial to an intact tympanic membrane. Reddish masses that blanch on seigelization are usually paragangliomas. Sometimes, a high rising jugular bulb or an aberrant carotid artery can also be seen. Whitish masses are usually congenital cholesteatomas, tympanosclerosis, cartilage grafts that have been surgically placed, and rarely middle ear adenomas (MEA). An elderly male presented with decreased hearing, which was revealed to be secondary to a pale mass in the middle ear, displacing the intact tympanic membrane laterally. He underwent biopsy followed by complete excision of the mass, with preservation of the intact ossicular chain. There no evidence of recurrence on follow-up. Histopathology revealed the mass to be a MEA, also known as middle ear neuroendocrine tumour (MeNET). Middle ear neuroendocrine tumours are rare tumours. We describe the clinical presentation and management of this tumour.

Keywords Middle ear neuroendocrine tumour, MeNET, MEA, carcinoid tumour, Neuroendocrine adenoma of middle ear, NAME

INTRODUCTION

A number of lesions can present with a mass behind an intact tympanic membrane, with varied symptomatology. Middle ear neuroendocrine tumour is a rare disease that originates in the middle ear mucosa.[1] It occurs over a wide age range, has no gender predilection.[2] Common presenting complaints are hearing loss and ear fullness on the affected side.[3] It may mimic a paraganglioma or a congenital cholesteatoma on clinical examination.[1,4]

CASE REPORT

A 70-year-old male presented with complaints of decreased hearing in right ear. On oto-endoscopic examination, a pale mass was seen occupying the middle ear, displacing the tympanic membrane laterally with protrusion postero-superiorly into the right external auditory canal, as seen in Figure 1.

The patient was subjected to high resolution computed tomography of the temporal bone, which showed a soft tissue density lesion occupying the entire middle ear and mastoid, and also obstructing the Eustachian tube orifice. The ossicular chain was intact (Figure 2). The patient underwent a biopsy under local anaesthesia to know the nature of the swelling.

Biopsy revealed an unencapsulated tumour composed of cells arranged in nests, trabeculae and glandular pattern, which were surrounded by fibrosis. The tumour cells had hyperchromatic nucleus with eosinophilic cytoplasm, suggestive of a of neuroendocrine adenoma of middle ear (Figure 3).

A simple mastoidectomy with epitympanotomy for clearance of the mass through a post aural approach was done. First, the tympanic membrane was carefully separated from the middle ear mass. Even though the mass was fragile, it was cleared from the Eustachian tube orifice. The mass was meticulously completely elevated off the mastoid antrum and from epitympanum where it was surrounding the incus body and the malleus head. The resected mass was sent again for histopathologic examination (HPE). Since the ossicular chain which was intact and mobile, it was retained. Type I tympanoplasty was done for hearing reconstruction. Histopathologic examination of the specimen showed similar findings as the initial biopsy, confirming the diagnosis.

Postoperatively, the patient perceived an improvement in hearing, which was evidenced by closure of 33 dB air-bone gap that was present preoperatively. The patient was followed up for next 6 years till when he was free of disease.

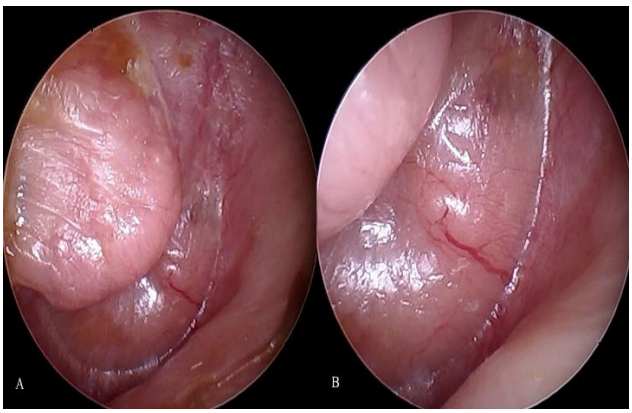

Figure 1 (A and B): Oto-endoscopy showing a pale mass occupying the middle ear, displacing the tympanic membrane laterally.

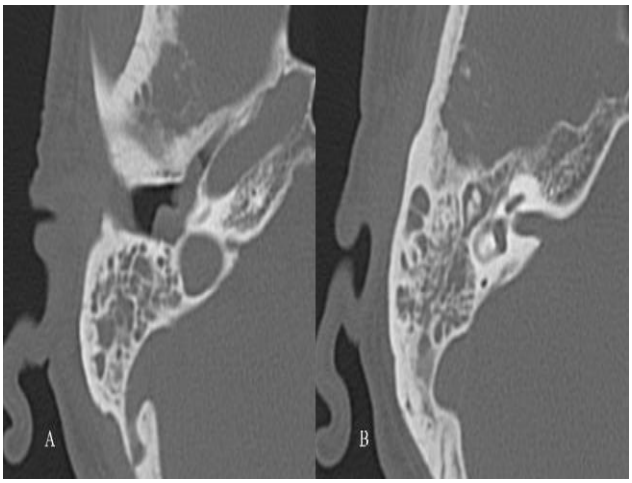

Figure 2 (A and B): HRCT Temporal bone shows: soft tissue density lesion occupying the entire middle ear and mastoid, and also obstructing the Eustachian tube orifice. Intact ossicular chain.

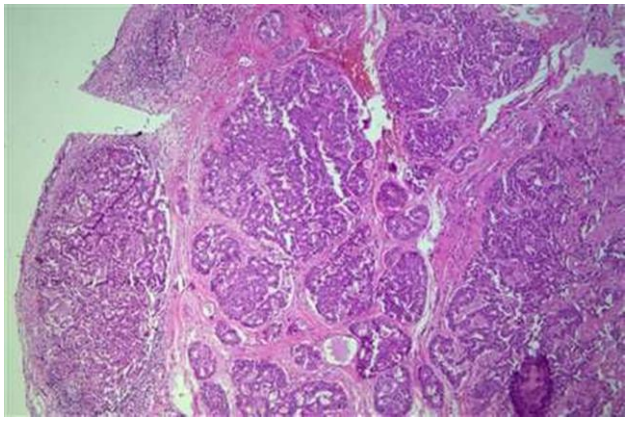

Figure 3: HPE of unencapsulated tumour composed of cells arranged in nests, trabeculae and glandular pattern, which were surrounded by fibrosis.

DISCUSSION

MEA was originally described by Hyams and Michaels in 1976 as a rare benign tumour derived from the lining epithelium of the middle ear mucosa that displays both exocrine and neuroendocrine differentiation.[5] It has since then been assigned several names MEA tous tumour (MEAT), neuroendocrine adenomas of the middle ear (NAME), middle ear carcinoid tumour.[2,3,6] As per the 5th edition of the World health organization classification of head and neck tumours, MEA has been renamed middle ear neuroendocrine tumour (MeNET) according to the nomenclature of neuroendocrine tumours at other sites.[7]

This is an uncommon tumour (<2% of ear tumours). These tumours show no sex predilection, and they usually affect middle-aged patients. Tumours are usually smaller than 1cm in their greatest dimension, and all involve the middle ear, but extension into the EAC (via the tympanic membrane), mastoid bone, and eustachian tube is not uncommon.[3,8] The clinical presentation often includes unilateral conductive hearing loss, aural fullness, tinnitus, discharge, otitis media, and bleeding. The overwhelming majority of MEA s do not invade or erode the temporal bone or infiltrate the facial nerve. Some patients experience facial paralysis.[1] Facial nerve paralysis and/or paresthesias may occur as a result of mass effect rather than nerve invasion.[3]

Microscopically, neuroendocrine adenomas of the middle ear are unencapsulated and "pseudo-invasive" with moderate cellularity. They display several growth characteristics: glandular and ribbon-like patterns, trabeculae, anastomosing cords, and solid sheets with variable cohesiveness. The predominant architectural pattern tends to vary among tumours and even within a single tumour. The cells are cuboidal to columnar and uniform in size, and with eosinophilic, finely granular, and homogenous cytoplasm.[3]

Even though MEAs are carcinoid tumours, our patient did not have any carcinoid symptoms. Carcinoid tumours are known to cause paraneoplastic systemic alterations caused by the secretion of hormonal products. This has been termed as "carcinoid syndrome" consisting of flushing, diarrhoea, sweating, wheezing, and abdominal pain. Some authors state that unlike carcinoid tumours of other organs, middle ear carcinoids do not manifest as typical carcinoid syndrome.[1] Although vasoactive compounds have been identified in carcinoid tumours of middle ear, only 1 report of carcinoid syndrome from a neuroendocrine tumour of middle ear exists. Lack of systemic changes associated with those tumours of the middle ear may be more a function of their relatively small size (lung tumours presenting with carcinoid symptoms tend to be large (>3.5 cm) rather than lack of biologic potential.[9]

On CT or MRI, a middle ear soft tissue lesion clearly showing significant enhancement after contrast medium injection is suggestive of a MEA. It can be differentiated from a paraganglioma on the basis that these lesions will be from the Jacobson nerve or its branches.[6]

The middle ear neuroendocrine tumour was previously considered to be benign.[2,5,10-12] However, recent studies have reported these tumours to be benign or malignant, tumour with a potentially malignant clinical behavior.[6,13]

Metastases involved the cervical lymph nodes in one patient and intra-parotid lymph nodes in three patients. This cervical lymph node metastasis was successfully treated with radiation. One of the patients who developed metastasis to the parotid also had intracranial and infratemporal fossa extension.[9,11]

Complete surgical resection is usually curative. Hearing can often be preserved. It has been suggested that if the tumour is intimately related to the ossicles, removal of the ossicles reduces the possibility of tumour recurrence.[14]

Recurrences or re-growths of neuroendocrine tumour may occur in as many as 20% of cases. They have been noticed after a varying interval of 1 year to 44 years.[11,13]

CONCLUSION

Middle ear neuroendocrine tumours are rare, low-grade malignant lesions with benign cell morphology. They have a tendency to reoccur locally, with a risk of metastasis. Complete resection of the tumour with long term follow up is recommended to be assured of a disease-free status.

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