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# Surprise Lesion In A Suspected Cholesteatoma

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***Abstract***
*Middle ear adenoma is a rare, benign tumour and accounts for minor proportion of middle ear glandular neoplasms. We report a case of middle ear adenoma in a 39/male came with complaints of right ear discharge on & off for one year. The case was clinically diagnosed as right unsafe ear after imaging study revealed the features of right ear cholesteatoma and the patient was operated on modified radical mastoidectomy. On microscopy, sections showed nonspecific inflammatory granulation tissue and there were few glandular proliferations lined by single to stratified layers of cuboidal to columnar epithelium with periodic acid shiff stain positive mucin. The immunohistochemical markers demonstrates positivity for cytokeratin and negativity for chromogranin. With all the Hematoxylin and Eosin, Periodic acid shiff stain and Immunohistochemical markers evaluation, the middle ear adenoma was diagnosed.*
***Keywords***: *middle ear adenoma (MEA), middle ear neuroendocrine tumor (MeNETs), neuroendocrine adenoma (NEA), immunohistochemistry (IHC), periodic acid shiff stain (PAS), World health organization (WHO)*

## INTRODUCTION

Middle ear adenoma is one of the rare benign glandular neoplasms of the middle ear, constitutes 2% to 4% of ear tumors overall [1]. It can have both exocrine and neuroendocrine component [2]. In 2024, World Health Organization recognized it as a middle ear neuroendocrine tumors according to [3]. There is no gender predominance. It has a wide range age distribution, but is prevalent in the 3rd to 5th span of decades [4]. Few studies have shown that it could have both exocrine and neuroendocrine differentiation. Its rarity and lack of proper pathogenesis had led to debatable classifications. Thus, histopathological and immunohistochemical examination are important for definitive diagnosis [1].

In this report, we present a case that was clinically and radiologically suspected to be right ear cholesteatoma but was ultimately diagnosed as middle ear adenoma exhibiting exocrine features on histopathology.

**Case report**

A 39-year-old man came to Outpatient Department with complaints of right ear discharge on and off for one year. The discharge was purulent, foul smelling and occasionally blood tinged. These complaints were aggravated by head washing and relieved by pain medications. A history of hearing impairment in the same ear was present. Clinical examination of the right ear revealed purulent discharge, attic perforation and congested margins. With all the clinical findings, the case was diagnosed as right ear cholesteatoma after radiological correlation, and the patient underwent modified radical mastoidectomy. Tissue bits from right mastoid region- attic, antrum and cavity - were submitted for histologic examination. Grossly, the specimen comprised multiple gray-white to gray-brown soft tissue bits collectively amounting to 0.5cc. The tissues were all embedded. Microscopically, the haematoxylin and eosin sections showed predominantly nonspecific inflammatory granulation tissue, few fragments of squamous epithelium and few areas of glandular proliferations. The glands were lined by single to stratified layers of cuboidal to columnar epithelium with round to oval nuclei and a moderate amount of eosinophilic cytoplasm [Figure 1A,1B]. Few glands were noted to contain mucin. The periodic acid shiff stain demonstrates mucin positivity [Figure 2]. Subsequently, the immunohistochemical markers cytokeratin and chromogranin were applied for further evaluation. Cytokeratin shows positivity in the glandular epithelium and chromogranin shows negativity [Figure 3A,3B]. With all the haematoxylin and eosin, periodic acid shiff stain and immunohistochemical markers evaluation, the middle ear adenoma was diagnosed.

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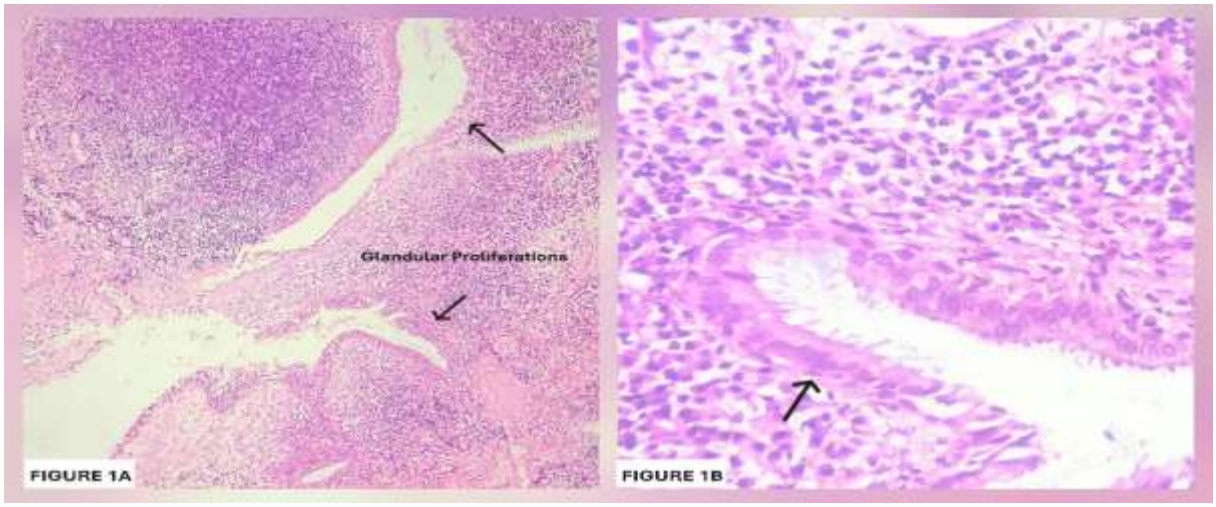


**Figure 1:**
**A) Microscopy examination showing glandular proliferations [H & E stain,100x]**
**B) Glands are lined by cuboidal to columnar epithelium [H & E stain,400x].**

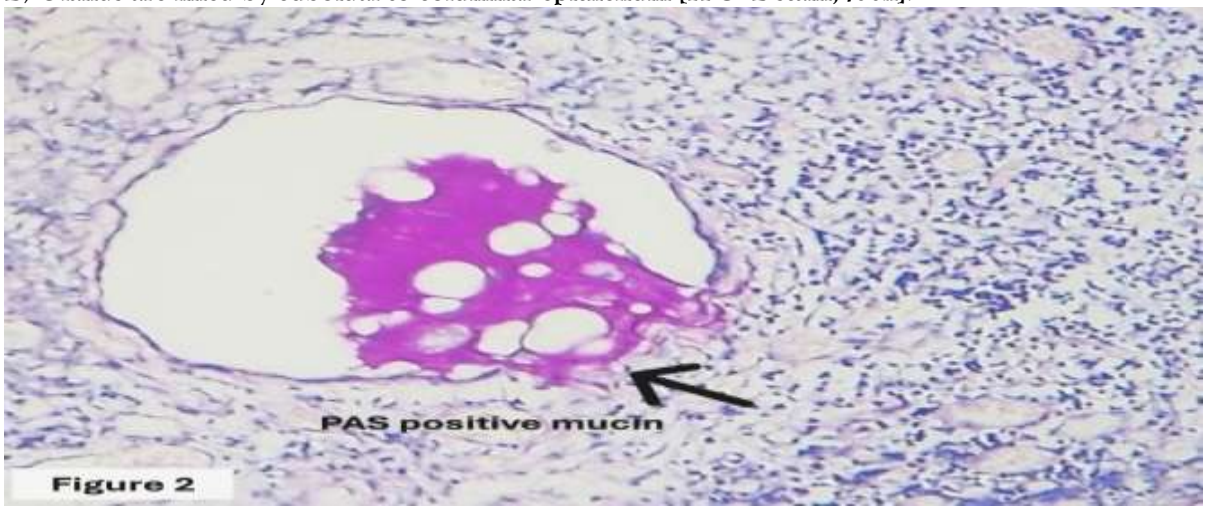


**Figure 2: PAS positive mucin [PAS stain, 100x]**

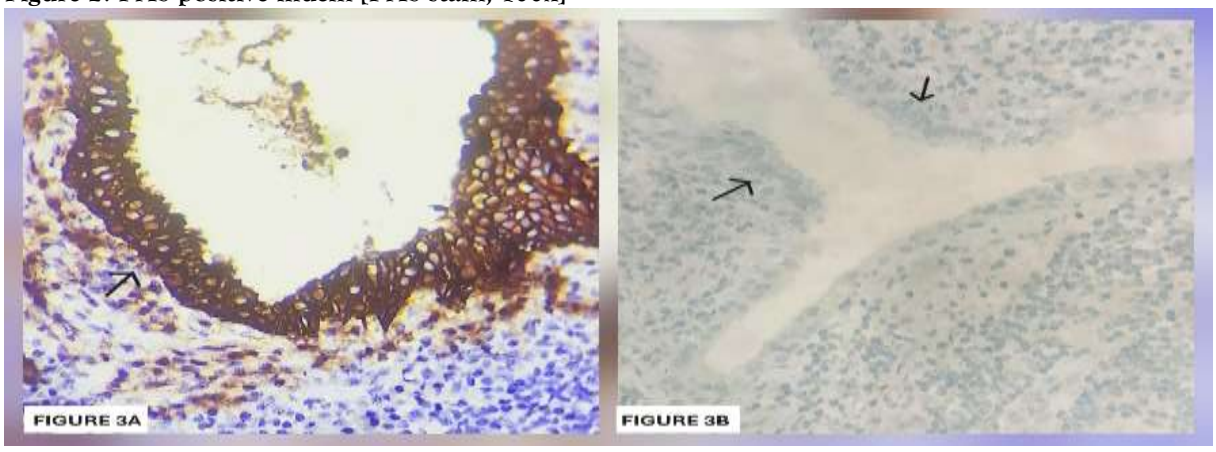


**Figure 3:**
**A) IHC for cytokeratin showing strong cytoplasmic positivity [IHC,100x].**
**B) IHC for chromogranin showing negative staining [IHC,100x].**

## DISCUSSION

MEA is rare benign middle ear glandular neoplasm of comprises about less than 2% to 4% of all ear tumours [1]. It is now recognized as MeNETs according to WHO [3]. It can have both epithelial and neuroendocrine component [2]. There is no gender predominance and has a broad age distribution, but

it is said to be common in the 3rd to 5th span of decade [4]. MEA was first discovered in 1976 as an adenomatous benign middle ear neoplasm with their unique characteristics by Hyams & Michaels in a series of 20 cases obtained from 1950 to 1970 [5]. Also, Derlacki and Barney described as a series of 3 cases. In 1980, Murphy et al described MEA as a "Carcinoid Tumor" because of the presence of neuroendocrine component [2]. Structurally, the tympanum is a small, air-filled spaces and contains ossicles (malleus, incus, and stapes) in the temporal bone and important for auditory signal transmission. The neuroendocrine characteristics in the epithelial lining is not usually found in the middle ear cavity [2]. The pathogenesis of MEA or MeNETs remains unclear and the potential factors would be genetic, persistent inflammation, and abnormal neuroendocrine cell differentiation [6]. Symptomatically, the complaints include ear fullness (mass), conducting hearing loss, infection, pain, discharge, tinnitus and dizziness [2,7]. Rarely, patients experience facial paralysis [4]. The otologic examination reveal mass, discharge, perforation depending upon the presentation. The audiometric evaluation shows conductive hearing loss [7]. Either CT and MRI are recommended for suspected MEA cases [2,6]. Usually, the radiology reveals non-destructive mass lesions in the middle ear; mostly with no bone destruction [4,6].
Grossly, MEAs are usually well circumscribed, unencapsulated, soft or rubbery white, grey, or reddish-brown mass. They are mostly avascular in nature. They may destruct the middle ear bony parts and the ossicular chain too [7]. Microscopically, MEAs show glandular, trabecular, solid, infiltrative and organoid pattern [8,5,4]. The lesion is composed of cuboidal -to-columnar epithelial cells [7] with a moderate amount of eosinophilic cytoplasm [7,8]. The nuclei are round to oval exhibit minimal pleomorphism and chromatin may show "salt and pepper" pattern correlating with neuroendocrine differentiation [MeNETs] and usually inconspicuous nucleoli [5,2,7]. Otherwise, the chromatin appears fine and even [7]. The other morphological patterns are plasmacytoid, flattened, irregular and spindled [4,7]. Mitosis is usually rare [5]. MEAs produce mucin, which is positive for PAS, Alcian blue, and mucicarmine stains [5,8]. However, the mainstay of treatment is surgical excision includes extirpation of tumor with auditory ossicles, if affected. Surgery should be done according to clinical and radiological correlation [9]. Few classification systems were proposed for MEA; however, the widely used one is 2009 classification by Saliba and Evrard. They categorized MEA into 3 types: NEA with positive IHC markers & no metastasis (type I), MEA with negative IHC marker & no metastasis (type II), and NEA tumor with positive IHC markers & metastasis (type III) [2,10]. In the WHO 2024 classification of head and neck tumours, MEA has been reclassified as MeNETs [3]. The epithelial or exocrine component of MEA is positive for cytokeratin (89.6% - 90%), CK7 (89.6%- 90%), CAM 5.2 (81.3%) and CK20- focally weak positive (6%). The neuroendocrine component is positive for chromogranin (87.5% - 80%), NSE (50%), synaptophysin (31.3%), and serotonin (25%) [7,1]. Usually Ki67 is low; around 2% -3% [6,11]. The differential diagnoses of middle ear MEA include various benign tumours & lesions - paraganglioma, schwannoma, Schneiderian-type mucosal papilloma, cholesteatoma, CSOM, choristoma, and hamartoma [4,5]. There are literatures saying MEA or MeNETs usually do not recur or metastasis [2,4]. Only one case report has been said to metastasize in cervical lymph node [2]. Rarely, they may have aggressive malignant features with higher Ki67 indices, which are always challenging for clear surgical margins and additional therapies needed [6]. But in recent literature, it's been said that around 22% recurrence is noted and 9 % metastasis noted of all MEAs. Also, the most predominant metastatic sites are the cervical lymph nodes, liver, and bones [12]. In MEA or MeNETs around 2 years follow up needed especially in case of suspected malignancy as they may recur or metastasize [2,6]. The follow up should include otoscopy, audiometry, with or without CT are recommended [4]. They also occur in paediatric age groups which are extremely rare. There are 4 paediatric cases (<18 years old) with MeNETs have been documented. Three patients underwent complete surgical excision and had 1-year follow-up [11].

## CONCLUSION

In our case, the tumour demonstrated cytokeratin positivity and chromogranin negativity, one of the key markers for neuroendocrine differentiation. Hence, the diagnosis of middle ear adenoma favoured than the middle ear neuroendocrine tumour, based on haematoxylin and eosin findings, periodic acid shiff positive mucin and immunohistochemical markers evaluation. Since MEA are diagnostically challenging and the surgical management is strongly recommended with ossicular chain removal to ensure a complete

excision and proper 2 years follow up period. To diagnose middle ear adenoma, it is very mandatory to correlate clinical presentation, radiology, histopathology and immunohistochemistry findings for definitive diagnosis.

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