INTRODUCTION

The first known documentation on middle ear adenoma as an uncommon lesion has been traced back to 1898 by Treitel, however, it was only as recently as 1976 that two separate articles, first by Hyams and Michaels and next by Derlacki and Barney, counted this lesion discretely as an adenomatous lesion. The first paper presented a case series of 20 cases collected over 20 years, between 1750 and 1970. The second paper was a series of 3 cases. While today they are recognized as middle ear adenoma, earlier these lesions were often described as ceruminoma, ceruminous adenoma, monomorphic adenoma and carcinoid tumour. As a result of their complex histological appearances, there are two schools of thought. One contends that carcinoids and middle ear adenomas are separate entities, while the other believes that they are variants of the same lesion.

Embryologically, the mucosa of middle ear develops from the foregut endocrine cells are known to be present in the bronchopulmonary and gastrointestinal tract of the foregut, which arises from the first pharyngeal pouch. It is, therefore, postulated that the middle ear adenoma may take its origin there.

They most often present with compromised hearing, ear fullness, ear ache and some less common symptoms. The differential diagnosis includes non-tumourous conditions like chronic otitis media, cholesteatoma and cancerous conditions like papillary adenocarcinoma or squamous cell carcinoma.

CASE REPORT

A 22 years old female was referred to Sligo General Hospital, Ireland. She presented with left sided deafness for a few months. She also complained of an intermittent bloody discharge from the same ear. There was no history of otalgia, tinnitus, dizziness or facial weakness. On examination under microscope revealed occlusive polyp in the left ear and a biopsy had been taken under general anaesthesia. Histopathology report described an adenoma / carcinoid tumour of the middle ear confirmed by positive immunohistochemical staining. CT temporal bones revealed the extension of the disease. The patient underwent left tympanotomy and excision of the tumour. The recurrence rate is low, but has also been reported years after treatment. At least one case of four recurrences and eventual systemic metastasis have been reported. Late recurrence, along with the metastatic potential, makes it important to follow-up the patients for years.

Key words: Neuroendocrine tumour of middle ear. Adenomatous middle ear tumour. Middle ear adenoma. Carcinoid tumour of the middle ear.
power view of the tumour revealed cords of uniform cells with some glandular differentiation suggestive of a middle ear adenoma / carcinoid tumour (Figures 1 and 2).

High resolution computed tomography of the temporal bone showed a soft tissue density mass present in the region of the left tympanic membrane. It was difficult to say if this opacification was actually in the external canal or in the middle ear. There was no definite evidence of bony erosion on CT (Figure 3). The middle ear ossicles appeared intact. The patient was referred to the neuro-otology clinic at Beaumont Hospital, Dublin, Ireland for a second opinion, where she underwent a left tympanotomy and excision of the tumour. Histopathology revealed a middle ear adenoma / carcinoid tumour. The patient was followed-up with the neuro-otology service at Beaumont Hospital Dublin, Ireland.

**DISCUSSION**

WHO classification as of 2005, considers middle ear adenoma as a benign tumour. It also considers adenomatoid tumours, neuroendocrine adenoma, and middle ear carcinoid as synonymous.\(^3\) There is another group, however, that wants it to be approached as a low grade malignancy for its metastatic character.\(^4\) The metastases have mostly been reported as local. Interestingly, despite a characteristic histological appearance, no matter where they are located, their presentation is prominently dependent upon their location.\(^3\) There is also no known familial predisposition to the development of the lesion.\(^1\)

Despite its rarity, there are a number of lesions for which middle ear adenoma may be a considered differential diagnosis. The most important non tumourous condition is chronic otitis media. The benign tumours are paraganglioma, schwannoma and schneiderian-type mucosal papilloma, cholesteatoma, choristoma and hamartoma.\(^1,5,6\) The primary malignant tumours in differential diagnosis are squamous cell carcinoma, rhabdomyosarcoma, and papillary adenocarcinoma.\(^1\)

Common presenting complaints with middle ear adenomas are hearing loss, a mass, and pain. Aoki et al.\(^4\) reported that the chief complaint is a hearing loss (> 90% cases), and upto a third complain of ear fullness, tinnitus and ear discharge.\(^4\) Hearing loss is mostly conductive. The tumour grows slowly, and the average disease duration is longer than 2 years. Morphologically, middle ear adenomas are mostly white, grey or rust brown in colour.\(^1\)

There is little to see on conventional CT or MRI scans, except an isodensity shadow equivalent to the density of otitis media and cholesteatoma. High resolution temporal CT usually shows a well circumscribed, soft tissue attenuation enhancing mass, without evidence of bone erosion.\(^4,7\) On MR imaging, the middle ear adenoma may show low to intermediate intensity on T1 weighted images, high on T2 weighted images, and enhancement following administration of gadolinium.\(^4,5\)

The diagnosis of middle ear adenoma is usually made on light microscopy confirmed on immuno-histochemical evaluation.\(^4\) Histological architecture could be trabecular, glandular or solid. The tumour cells could be cuboidal or cylindrical, with acidophilic cytoplasm which may appear plasmacytoid.\(^1\) The nuclei may appear round to oval with a finely stippled nuclear chromatin,\(^6\) a remarkable ‘salt and pepper’ chromatin pattern\(^1\) and inconspicuous nucleoli.

It is very important to know that frozen section histopathology of this tumour could be misleading. Burn and Pearl refer to a study of a series of 32 frozen section biopsies taken from the middle ear, within which the final diagnosis of middle ear adenoma could not be made on the initial microscopy on the frozen section. Out of the 32 initial diagnoses, 31 mistaken initial diagnoses were paraganglioma (21), chronic otitis media (6) and cholesteatoma (5).\(^1\) Definitive diagnosis is greatly assisted by immunohistochemical examination.\(^5\)

Immunohistochemical staining, however, is of little help in differentiating them from endolymphatic sac tumours as both present a similar spectrum of neuroectodermal staining characteristics.\(^9\)

A conservative local resection is generally considered sufficient for carcinoids of the small bowel. Since middle

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**Figure 1:** This low power view shows the trabecular appearance of the tumour with cords of cells in the submucosa.

**Figure 2:** A higher power view of the tumour showing cords of uniform cells with some glandular differentiation.

**Figure 3:** CT temporal bone showing an area of soft tissue opacification at the left tympanic membrane.
ear adenomas are also circumscribed, the same approach may be employed in the middle ear. Although there is no known established approach for treatment of middle ear adenoma, perhaps owing to a very limited incidence of the condition, complete removal by tympano-mastoidectomy or radical mastoidectomy has been favoured more.\textsuperscript{4}

Adjuvant radiotherapy has been prescribed in aggressive cases of middle ear adenoma, but there is limited evidence in favour of its clinical efficacy.\textsuperscript{4} At least one case of neo-adjuvant and later adjuvant radiotherapy has been also reported, which could not result in successful treatment and the tumour recurred four times, eventually metastasizing to the liver. The patient died during systemic chemotherapy and the total duration from diagnosis to death was about 11 years.\textsuperscript{2}

Aoki and colleagues reported that 6 out of 34 cases who had a surgical procedure of middle ear adenoma had a recurrence, the time interval averaging 200 months. They also observed that this interval was much longer (15 - 33 years), following tympano-mastoidectomy or radical mastoidectomy.\textsuperscript{4} They have, however, not stated the number of cases who were treated as such. Saliba and Evrard write that if the involved ossicular chain is not removed, the probability of recurrence is higher. They, therefore, suggest not just complete removal of the lesion but also the encased ossicles.\textsuperscript{10} These characteristics of the tumour, arguably call for a long duration of follow-up as has also been advised in literature.\textsuperscript{3}

The middle ear adenomas are benign tumours and not known to recur often.\textsuperscript{1} A recurrence is, however, known to have occurred after incomplete surgical removal. Also, an involvement of facial nerve has been quoted as a poor prognostic sign.\textsuperscript{1} The Ki-67 labelling index seems a reasonably reliable indicator of tumour grade, but a statistical cut off is not known as yet.\textsuperscript{4} Extension through the tympanic membrane has also been reported in the literature.\textsuperscript{1} Evidence suggests that incidence of recurrence is higher (14%) with transcanal tympano- tomy, than with radical mastoidectomy (9%). However, this evidence is not considered enough to suggest clinical superiority of one procedure over the other.\textsuperscript{10}

\section*{REFERENCES}
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