CASE REPORT

Myxoid Neurofibromas of the External Ear Canal: A Case Report

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SUMMARY

Myxoid neurofibroma is benign tumours of perineural cell origin that arise from elements in the peripheral nervous system. We report a case of a 60-year-old female patient presented with history of right ear mass which was slowly growing. Her primary complaint was cosmetic deformity but hearing loss was also present. The mass was excised and histologically revealed a myxoid neurofibroma. To the authors' knowledge, this is the first time that a myxoid neurofibroma arising from external auditory canal is reported. It should be included in the differential diagnosis of a mass originating from this location.

KEY WORDS:
Neurofibroma, External ear canal

INTRODUCTION

The external auditory canal (EAC) can be the site of development of different benign tumours but rarely neurofibromas. Neurofibromas are relatively common lesions arising from elements in the peripheral nervous system. Therefore, they may develop anywhere in the body either just under the surface of the skin or deeper within the body. If they occur in EAC then deformity of the ear is usually the main symptoms with functional impairment in the form of hearing loss which is caused by obstruction of the EAC or invasion of the middle and inner ear. There were a few published reports on neurofibromas affecting EAC but to the authors' knowledge, a case of myxoid neurofibroma has never been reported in English literatures.[1, 2]

CASE REPORT

A 60-year-old female patient presented with 20-year history of right ear mass which was slow growing. There was history of diminished hearing from right ear that was progressive in nature but without any pain or discharge.

There was no past history of otitis externa, otitis media or trauma .There was no history of headache, vertigo, vomiting and fever or any complaint from nose or throat.

On examination of the right ear, there was a pale lobulated mass arising from the EAC (figure 1). It was a solitary nodule, skin-colour, firm in consistency and painless. The size of the external part was 2.5x3cm. The pinna, post auricular region and the mastoid area were normal and non-tender. The tympanic membrane could not be appreciated due to the mass completely obstructing the canal.

On tuning fork tests, Rinne test was negative in the right ear and positive in the left ear. Weber was lateralized to the right ear thus indicating conductive hearing loss in the right ear. X-ray both mastoids (lateral oblique view) showed normally pneumatised mastoid air cells bilaterally. Temporal bone CT scan showed the mass arising from right EAC occupies whole length of the canal and limited medially by the right tympanic membrane. Middle ear ossicles and inner ear structure were normal.

Under general anaesthesia whole of the mass was excised through a combination of postauricular and peraural approach. The mass was noted to be arising from the skin of postero-inferior wall of EAC with the length of 3.5cm (figure 1).

Microscopic description showed that the lesion was composed of proliferation of spindle cells arranged in haphazard manner within an extensive myxoid stroma (figure 2). Scattered s100 protein positive spindle cells were present (figure 2).

DISCUSSION

Peripheral nerve sheath tumours can be divided into benign and malignant. The two major benign categories are neurofibroma and schwannoma while a malignant form is malignant peripheral nerve sheath tumour.[3] Each benign category can be a part of neurofibromatosis type 1 or von Recklinghausen's syndrome which is characterised by cafe au lait skin patches, Lisch's nodules, axillary freckling, and fibroma molluscum but solitary neurofibroma is a localised neurofibroma which is usually not associated with neurofibromatosis. Even though they are usually benign, but some cause local destruction secondary to pressure effects. Variants of Solitary neurofibroma include cutaneous lipomatous, collagenous, epithelioid, granular, pigmented, denritic cell and myxoid neurofibromas.[4]

As in our case, Myxoid neurofibroma (MN) presents as a painless, slow growing and firm in consistency solitary nodule. Based on the histological and immunohistochemical findings, MN confirmed with abundant mucin in the matrix

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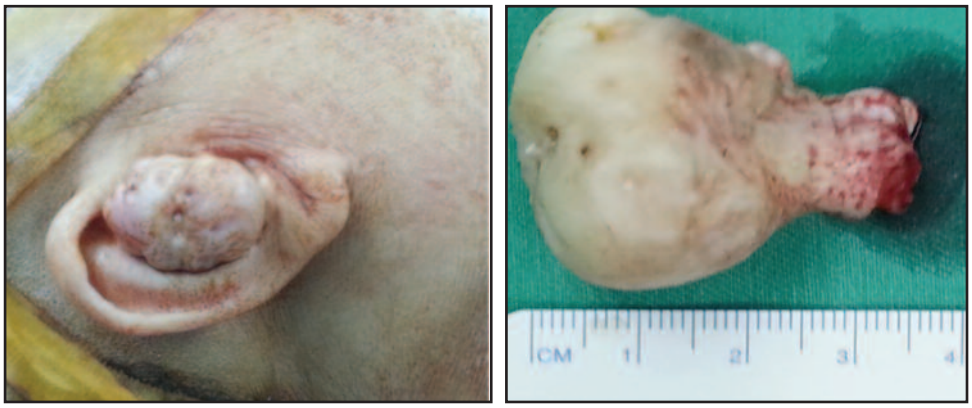

Fig. 1: A lobulated mass at the entrance of the right external auditory canal (left) and the mass after removal (right).

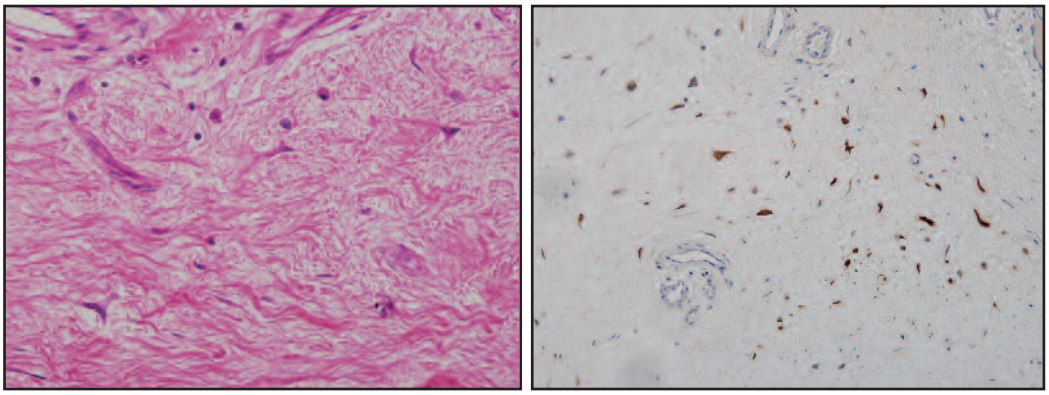

Fig. 2: Proliferation of spindle cells arranged as haphazard manner within an extensive myxoid stroma with scattered mast cells and blood vessels (X20 magnifications) (left) and scattered s100 protein positive spindle cells (x20 magnification) (right).

which makes a positive immunohisto-chemical staining for S-100 protein.

Neurofibromas should be considered in the differential diagnosis of other benign or malignant tumours of the external ear canal. CT scan is required for identifying the true extension and benign nature of this lesion. The treatment is total excision of tumour but most of the time for cosmetic or diagnostic reasons. However, it may cause functional impairment if it occurs in certain area such as in EAC.

CONCLUSION

To the authors' knowledge, this is the first time that Myxoid neurofibroma arising from EAC is reported. It should be included in the differential diagnosis of a mass originating from this location.

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