International Journal of Otorhinolaryngology and Head and Neck Surgery
Galagali JR et al. Int J Otorhinolaryngol Head Neck Surg. 2016 Jan;2(1):53-55
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20160071

Fibromyxoma of temporal bone: a case report

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Received: 22 October 2015
Accepted: 05 December 2015

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ABSTRACT

Tumours of temporal bone are rare. This case is described to highlight the clinical presentation and management of this relatively rare neoplasm.

Keywords: Fibromyxoma, Temporal bone, Abducens nerve palsy

INTRODUCTION

Tumors of temporal bone are rare. Commonly encountered tumours include glomus jugulare, adenoma, adenocarcinoma, osteoma, fibrosarcoma, neurofibroma, meningioma, osteoclastoma, acoustic neuroma & squamous cell carcinomas.[1] Fibromyxoma of temporal bone is a rare entity and review of literature has revealed only one case.[2] Fibromyxoma of petrous apex has also been reported.[3,4] This case is described to highlight the clinical presentation and management of this relatively rare neoplasm.

CASE REPORT

A 20 yrs old male was referred to a tertiary care hospital neurology center with history of loss of consciousness associated with left sided headache of two months duration. Headache was throbbing in nature, associated with frequent vomiting with history of blurring of vision of left eye of one month duration. Patient had an attack of loss of consciousness lasting for 30 min with drooling of saliva and was thought to have had an attack of seizure and hence referred to the neurophysician. There was no history of frank seizures, hypertension, diabetes mellitus or tuberculosis in the past. No past history of otorrhoea, otalgia, tinnitus or facial palsy was offered by the patient. He was referred to the ENT surgeon to rule out any possible ENT focus.

ENT examination revealed bilateral intact and mobile tympanic membranes (TM) with no hearing loss. Otoneurological exam was normal. Next day patient developed diplopia (Figure 1) following abducens nerve palsy left ? Gradenigo's syndrome and was treated with injectable antibiotics and other supportive therapy. Routine investigations & biochemical parameters were normal. Pure tone audiometry showed normal AC & BC thresholds within 20 dB. Contrast enhancing CT scan brain showed destructive mass / lytic lesion in left mastoid ? Congenital Cholesteatoma (Figure 2). The patient was taken up for tympanomastoid exploration under general anaesthesia and modified radical mastoidectomy with type III tympanoplasty with complete excision of tumor mass from mastoid cavity was done.

Intra-operative findings

After performing cortical mastoidectomy, to our surprise, a jelly like mass was noticed in the mastoid cavity (Figure 3), which did not resemble cholesteatoma or granulation & did not bleed on touch. It was seen eroding the mastoid bone. The tumor mass was seen extending from mastoid cavity to epitympanum, petrous apex

medially and to mastoid tip inferiorly. The tumor mass was excised in toto & sent for HPE. Facial bridge was broken & the ridge lowered. The chorda tympanic nerve was spared. The temporalis fascia was placed below the remnant of tympanic membrane and over the stapes head and facial ridge.

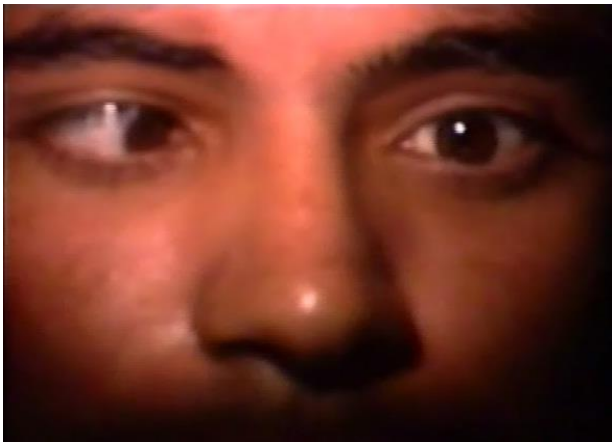

Figure 1: Lateral rectus palsy (preoperative).

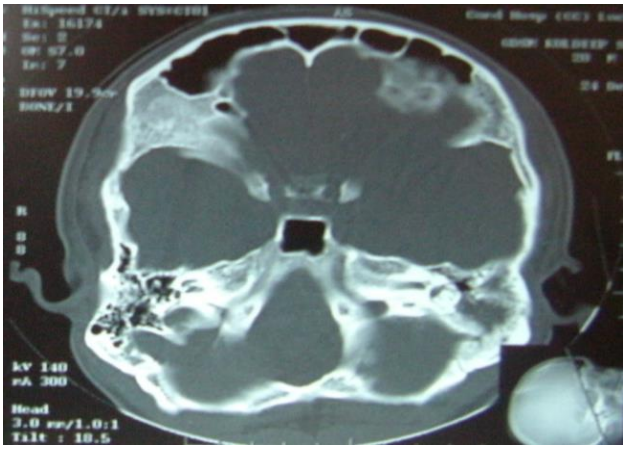

Figure 2: CECT scan brain showed destructive mass / lytic lesion in left mastoid.

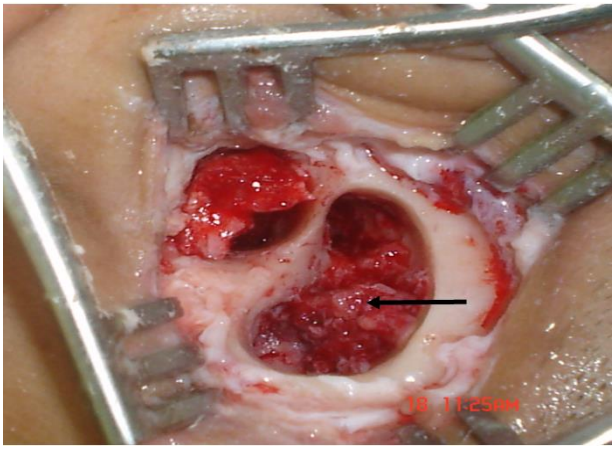

Figure 3: A jelly like mass noticed in the mastoid cavity (Black Arrow).

Post operatively, the patient was continued of injectable antibiotics for a period of 14 days and systemic steroid, inj Wymesone, was given for a period of 05 days. Oral antibiotics were continued for another 14 days. Post aural sutures were removed on 7th post operative day. The headache reduced completely by 7th post operative day & the abducens nerve palsy (left) started improving. By 15th post operative day, diplopia persisted only when eye was in left gaze. Patient was further given oral steroids for 21 days in tapering dose with supportive therapy.

Histopathological examination confirmed the diagnosis as Benign Fibromyxoma of Temporal bone (Lt) (Figure 4).

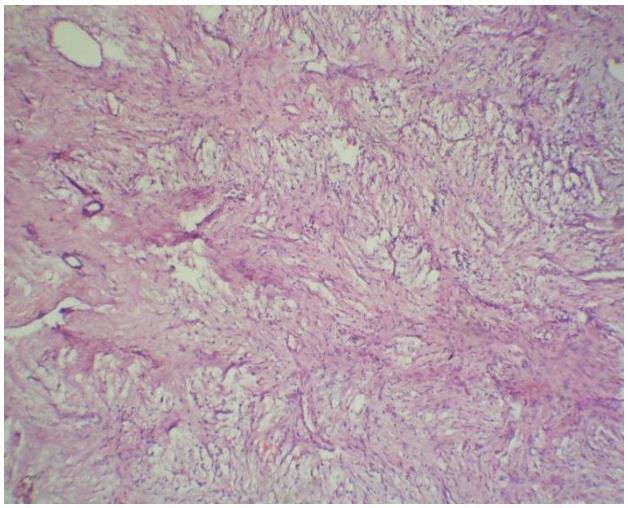

Figure 4: HPE slide of the mass suggestive of benign Fibromyxoma.

Follow up

Patient remains asymptomatic with normal eye movements (Figure 5) and no evidence of recurrence of disease after 38 months, as on 06 Mar 09 on CT scan. However has been lost to follow up thereafter.

Figure 5: Improvement in lateral rectus palsy (Lt) postop.

DISCUSSION

Myxomas are benign neoplasms derived from primitive mesenchyme. Initially coined by Virchow in 1871, the term myxoma traditionally has been used to describe tumors that resemble the mucinous appearance of the umbilical cord.[5] Fibromyxoma of the temporal bone is a primary bone neoplasm which is a benign, locally infiltrative, usually involving skull bones, mandible and long bones & never reported to metastasize. Fibromyxomas are soft, smooth, jelly like tumors resembling Wharton's jelly. The tumor is weakly vascularized. Within the temporal bone the tumor leads to destruction of bone by pressure effect & resembles destruction by cholesteatoma. Clinically the tumor is slow growing which causes circumscribed osteolysis of bone. Histopathological examination reveals tissue composed of stellate & spindle shaped cells, set in a myxoid stroma with variable degrees of fibrosis with small isophorphic nuclei without mitosis.[6] Areas of connective tissue with loose collagen fibres may be seen forming border to the myxomatous zones. There are no specific cellular elements like chondroblasts or lipoblasts. Although it can give the impression radiologically of an aggressive growing tumor, it is a benign lesion for which conservative surgical excision of tumor is sufficient & not curettage. Follow up is required as recurrence is likely.

Funding: No funding sources

Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Galagali JR, Mahajan SB, Magu SK. Fibromyxoma of temporal bone: a case report. Int J Otorhinolaryngol Head Neck Surg 2016;2:53-5.