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Case Report

A Rare Case of Chondromyxoid Fibroma of Temporal Bone, Left Side

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Abstract

Chondromyxoid fibroma (CMF) itself is a rare neoplasm of bones. It is very rarely only seen, in skull bones, and if at all it affects the skull, usually in mandible or maxilla, and extremely rarely only affects the ear bone. This is a case report on this very rare benign neoplasm of Ear bone.

Keywords: Chondromyxofibroma; Benign Neoplasms of Ear

Introduction

Chondromyxoid fibroma (CMF) is a slow growing, rare, benign primary bone tumour of cartilaginous origin. It was first described by Jaffe and Lichenstein in 1948. It represents 0.4-0.5% of all the primary bone tumours and typically affects more males than females. Most common site of origin is metaphysis of long bones mainly around the knee joint (40%) but also occur in small bones of hands, feet, pelvis, vertebrae and ribs. The overall incidence of CMF in the craniofacial region ranges from 2-5% and most commonly occur in maxilla or mandible. CMF in the temporal bone is very rare. We are presenting such a rare case of CMF in the temporal bone, its detailed work up and management plan we adopted [3].

Case Report

The patient was 35year old man with no known comorbidities had complaints of aural fullness in left side since 2weeks.He had occasional tingling and numbness around left ear since 2 yrs. No h/o headache, hearing loss, facial pain, otalgia.

Tinnitus and diplopia. On examination there was a large pinkish swelling, arising from the floor and posterior wall of external auditory canal (Figure 1). There were no similar swellings in the body.

Figure 1: Otoendoscopic image.

We further evaluated with HRCT temporal bone which showed that the left mastoid process was replaced with irregular soft tissue/sclerosed trabecular matrix (inhomogeneous enhancement) measuring 28 mm AP, 25 mm CC, 25 mm.

Transverse and focal erosions of sigmoid sinus plate, small cortical defects along the outer aspect of the mastoid process and in the lateral part of the bony external auditory canal along the floor. Small soft tissue component was seen extending through the defect in the floor into the EAC (Figure 2).

Figure 2: HRCT temporal bone.

Then a provisional diagnosis of hemangioma in the left mastoid process with bone erosions and extension into EAC was made. Subsequent MRI brain also suggested hemangioma as the probable diagnosis. There was a well circumscribed, lobulated brightly enhancing T1 iso to hyperintense and T2/FLAIR altered signal intensity lesion involving the left mastoid air cells which abutting and indenting adjacent sigmoid sinus and erosion of the floor and posterior wall of EAC (Figure 3).

Figure 3: MRI Brain.

Then we decided to proceed the case with excision biopsy. Surgical exploration of mastoid and external canal, followed by excision of tumour was done. The tumour was highly vascular and it involved the tip and posterior aspect of the mastoid. Posterior canal wall was eroded and the tumour was projecting into the canal from the posterior wall. The same removed but the posterior wall defect was not closed due to severe bleeding as a result of injury to sigmoid sinus on attempted complete clearance of disease which was controlled by packing (Figure 4).

Figure 4: Intraoperative images.

Posterior wall reconstruction and removal of any residual tumour if present is planned later. Microscopically the section showed bony trabeculae and a lesion with lobulated architecture. The lobules are composed of hypocellular myxoid centres with peripheral hypercellular areas admixed with specks of coarse calcification. The hypocellular areas are composed of stellate to spindle cells admixed with hyaline cartilage in myxohyaline vascularized stroma. Based on the histopathological findings it was diagnosed as chondromyxoid fibroma (Figure 5).

Figure 5: Histopathology.

Discussion

CMF itself is a rare benign neoplasm of bone that represents less than 0.5% of all primary bone tumours and the common site of origin is metaphysis of long bones mainly around the knee joint. This rare neoplasm of bone rarely only affects the skull bones. When it affects skull bones, it most commonly occurs in maxilla or mandible. Extremely rarely affects the ear [1].

There is male preponderance with peak incidence in the second and third decade of life. The fact that cranial bones develop from endochondral tissue has led to the belief that CMF arises from cartilage or embryonic rests of cartilage, which can be found in areas of synchondrosis at the base of the skull and at suture sites. Due to the slow growth of the tumour symptoms are few and tend to be of long duration [2].

CT and MRI are useful modalities for preoperative diagnosis. CT scan findings of CMF are relatively homogenous, well circumscribed, osteolytic lesion with a wavy bony margin and foci of calcification. Intratumoral calcification is reported in craniofacial CMF. MRI is used to determine the extension of the lesion into the dura and intracranial space. CMF is typically hypointense (low signal) in T1 weighted and heterogeneously hyperintense (high signal) in T2 weighted imaging. The lesion is markedly enhanced after Gadolinium contrast administration. In this case radiological features were confusing with hemangioma and a definitive diagnosis could not be made. Therefore, biopsy was needed to achieve a definitive diagnosis [2].

CMF is characterized by lobules of spindle shaped or stellate cells with abundant myxoid or chondroid intercellular material with varying numbers of multinucleated giant cells of different sizes. Histopathological analysis revealed bony trabeculae and a lesion with lobulated architecture. The lobules are composed of hypocellular myxoid centres with peripheral hypercellular areas admixed with specks of coarse calcification. The hypocellular areas are composed of stellate to spindle cells admixed with hyaline cartilage in myxohyaline vascularized stroma [3].

In temporal bone lesions it is important to distinguish CMFs from other tumours such as chordoma, myxoidchondrosarcoma and facial schwannoma. Chordomas usually express epithelial antigens such as epithelial membrane and keratin. CMFs are not stained by antibodies against these proteins. It is important to distinguish between CMF and myxoidchondrosarcomas because their treatments are different. They have similar MRI features and both are positive for S-100 in immunohistochemical analysis. Hence it is very difficult to differentiate these two entities. It is crucial to differentiate between CMF and chondrosarcoma. Because the management and prognosis differ significantly. It was reported that about 22% of CMF cases were misdiagnosed as chondrosarcoma [4].

CMF should be managed by extensive focal or total resection of the affected bone to reduce the chances of recurrence. Recurrence rate after a simple curettage was reported as 12.5 to 25%.

Conclusion

CMF is a benign but potentially aggressive tumour. Involvement of the skull base especially temporal bone is very rare. Biopsy is necessary to exclude it from other lesions of the skull base with chondroid origin. Complete resection is the treatment of choice since the radiotherapy for inoperable or recurrent lesions carries the risk of malignant transformation.

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