Case Report :

Chondromyxoid Fibroma In Skull; An Unusual Site.

Saira Afeef Zia[1], Zainab Malik[1], Khurram Khaliq Bhinder[1], Ameena[2], Shaista Riaz[3]

1. Shifa International Hospital,Islamabad,Pakistan .
2. Lahore General Hospital, Lahore,Pakistan .
3. McGill University,Montreal,Canada .

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Corresponding Author:

Khurram Khaliq Bhinder,Resident Radiologist,
Shifa International Hospital, H-8, Islamabad,Pakistan.
kkbhinder@yahoo.com

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Abstract

Chondromyxoid fibroma are the rarest of the benign cartilaginous neoplasms. Mostly it involves metaphysis of long bones, however, rarely can be found in calvarium / skull base. We report a case of a 30 year-old female who was referred to the Radiology Department at Shifa International Hospital, Islamabad with history of progression and gradual loss of vision in unilateral eye. The patient had contrast enhanced MRI of brain which showed a heterogeneously enhancing mass lesion abutting the clivus, CP angle and brain stem with internal areas of necrosis. Differentials of chordoma, meningioma and chondrosarcoma were given. The patient then underwent surgery which confirmed the mass to be chondromyxoid fibroma.

Keywords:

Chondromyxoid Fibroma, Clivus, Cerebellopontine.

Introduction

Chondromyxoid fibroma (CMF) is infrequent benign cartilaginous tumor [1]. It represents 0.5% of all bone tumors ,with 71% cases involve long bones of lower limbs on epiphyseal part, most commonly in knee joint(40%) and 2% of CMF are present in skull. Chondromyxoid fibroma occurrence in temporal bone is a rare entity [2]. Chondromyxoid fibroma is characterised by lobules of spindle shaped or stellate cells with sufficient myxoid or chondroid intracellular material with multinucleate giant cells of varying sizes [3].

It is frequently diagnosed in the second and third decade of life, most commonly in male with male to female ratio 1.28:1. CMF is difficult to diagnosed as it is misdiagnosed with aneurysmal bone cysts, giant cell tumors (GCTs), chondrosarcoma, chondroblastoma and enchondroma [4].

In this paper, we report one case of chondromyxoid fibroma of temporal bone with clinical presentations, radiological

and histopathological studies.

Case presentation

30 years old with no comorbids presented to our emergency department with active complain of headache for last 3 years. She also started experiencing decreased vision over passage of time in her right eye. She had an episode of sudden loss of consciousness two weeks prior to admission and developed weakness in her right limbs. No associated symptom of nausea, vomiting, fits a nd memory disturbance was reported. Contrast ehanced MRI brain was ordered which showed extra axial appearing abnormal signal mass lesion with heterogenous post contrast enhancement and internal areas of hemorhage, lying posterior to and abutting the clivus,anterior to the brainstem and extending into bilateral CP angle cisterns,causing significant mass effect on the brain stem and posterior fossa structures.Signal abnormality in the left hemipons was reported due to mass effect. Extension of the lesion into the region of right cavernous sinus, mildly compressed and slightly displaced right ICA. Neurosurgery department was called and urgent right pterional craniotomy was done. Mass was excised and sent for biopsy to histopathological team, who reported, to be a chondromyxoid fibroma. This tumour was arranged in lobular architecture with fibrous bands separating these lobules. These lobules were composed of stellate cells in a chondromyxoid stroma and bony trabeculae and foci of calcification were also present. On immunochemistry, the specimen was positive for S-100 and negative for PR, EMA and PanCK. Non contrast CT scan brain post right pterional craniotomy was done to look for post-operative brain changes which revealed mild edema (Figure 1, 2 and 3).

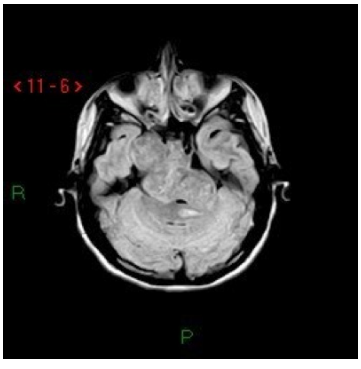


Figure 1

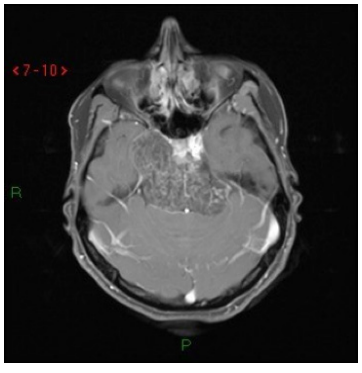


Figure 2



Figure 3

Discussion

Chondromyxoid fibroma is a slow growing benign chondrogenic tumor. Temporal bone is an extremely rare location of chondromyxoid fibroma [1].

Its pathogenesis involve neoplastic degeneration of residual embryonic elements or residual ectopic cartilaginous tissue of skull base sutures that disappear during endochondral ossification process [5]. CT and MRI are used for pre-operative diagnosis of CMF. CT imaging shows osteolytic lesions with marginal sclerosis. MRI is more effective than CT as it shows shows the extent of the disease. CMF tissue shows low density on T1 weighted imaging and non uniform high intensity on T2 weighted imaging [2].

The histolopathological results shows myxoid lesion containing paucicellular center, bland stroma cells, reactive boney spiculesand hyaline cartilage.CMF rarelyshows matrix calcifications in skull lesions. Lobular appearance,chondromyxoid stroma and fibrous tissue with multinucleated giant cells are characteristic features of chondromyxoid fibroma [3]. In our case, the biopsy findings include a tumour with lobular architecture with fibrous bands which were positive for S-100.

The clinical features varies according to the site of origin. Symptoms for skull based lesions include headache, neuralgia, facial pain,exophthalmos,convulsions and diplopia [6].In this case, patient had headache and decreasing vision loss with the passage of time.

Chondromyxoid fibroma is difficult to diagnose as chondrosarcomas, chondromas, chondroblastomas are closely related to CMF both in clinical presentations and histologically.

Patients with CMF presents with progressive pain with bone swellings and restricted range of movements in affected limb. These symptoms are also found in patients with chondrosarcomas [4].

CMF differs from chondrosarcoma by its lack of invasive growth pattern and observation of tumor tissue encasing preexisting bony trabeculae. Chondroma's infiltrative margin and tissue containing large epithelial cells with eosinophilic or vacoulated cytoplasm are features of chondroma that distinguish it from chondromyxoid fibroma [7].

The definitive treatment of CMF is surgical resection with resection of portion of surrounding normal bone to prevent recurrence and malignant conversion. The chances of recurrence and conversion to malignancy of CMF varies between 3% to 26% [6].

In our case clinical radiological and histopathological features led to diagnosis of chondromyxoid fibroma.

Conclusion

We report a rare case of chondromyxoid fibroma of temporal bone in a 30 years female which is a benign cartilaginous tumor. The clinical presentation varies according to the site. The detailed physical examination, investigations can decrease the chances of misdiagnosis and prompt surgical interventions is effective in the treatment.

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