CASE REPORT

Chondromyxoid fibroma of zygoma: A rare case report

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ABSTRACT

Chondromyxoid fibroma (CMF) is a rare benign mesenchymal tumor of the bone. Clinically, it is characterized by a lobular growth pattern and histologically by chondroid and myxoid differentiation. The tumor is rare in the craniofacial bones with only 2% of all reported cases. Extragnathic location in the facial skeleton is extremely rare. Most of the cases reported either originate from gnathic sites or in the cranium. A case of CMF in a 3½-year-old male is presented here, which arose from the root of zygomatic arch. A detailed clinical history and histopathological picture of one more case is added to the literature. It is important to document such cases so that better light can be shed on future reviews and conclusions. This shall facilitate better treatment approaches and prognosis. This case is the first reported case of involvement of the zygomatic arch in a pediatric patient.

Key words: Benign, cartilage, chondromyxoid fibroma, lobulated, myxoid, paediatric, zygomatic tumor

INTRODUCTION

Chondromyxoid fibroma is a separate and distinct benign tumor of cartilaginous origin, which involves craniofacial bones.[1] This rare tumor of the bone was originally described by Jaffey and Lischtenstein in 1948[2,3] accounting for less than 0.4-1% of all bone tumors.[1,2] CMF contains variable amounts of chondroid, fibromatoid and myxoid components. The presence of tissue resembling hyaline cartilage in the tumor justifies its classification as a cartilaginous tumor.[3] Most of the authors feel that the designation of fibromyxoid chondroma may fit the tumor better, as it gives an indication of histological derivation along with the description of the lesional components.[4]

This tumor has been defined as "a benign tumor characterized by lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle-shaped or round cells with varying number of multinucleated giant cells of different sizes" by the World Health Organization (WHO).[5]

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CMF occurs mostly in the long bones of appendicular skeleton, but occurrence in craniofacial and gnathic regions is rare.[2] This tumor is more common in the second decade of life with a history of slow growth and gradually develops symptoms over a period of months to years duration. The common symptoms are pain, swelling and local restriction of movement. Occasional cases are asymptomatic. Radiographically, it is a radiolucent lesion, which frequently has a lobulated outline with defined margins and usually with a sclerotic rim except in the cases of gnathic origin, suggesting a benign lesion. Intra-lesional calcified matrix is very rare.[6]

Histopathological diagnosis is sometimes straightforward due to the biphasic pattern but it can be challenging when it is similar to chondrogenic sarcoma.

CASE REPORT

A 3½-year-old Indian boy visited a private dental clinic with a complaint of a small painless mass in the region of the left zygomatic arch [Figure 1]. The mass was noticed by the mother to be slow growing over the last 6 months but was to be slow growing over the preceding 6 months but was asymptomatic.

Extra-oral examination revealed a firm, immobile, oval swelling measuring 2.5 × 2.0 cm in dimension located in the left pre-auricular area. The swelling was non-tender and the skin above the swelling appeared normal and was not fixed to the underlying lesion.

The head and neck examination was non-contributory, with absence of cervical lymphadenopathy. An examination of the ear, nose, throat and eyes revealed no abnormality.

A computed tomographic (CT) scan revealed a bony lesion on the root of zygomatic arch, extending from the zygomatic extension of the temporal bone [Figure 2].

Under general anesthesia, the tumor was approached with a pre-auricular incision. The tumor was dissected from the underlying tissue with ease and a lesion of 2.0 × 1.5 cm, was scooped out en block. The underlying bone had a raw and eroded surface, which was curetted well and the surgical site was closed.

On macroscopic examination, the tumor was solid and lobulated [Figure 3]. The cut-section revealed a well circumscribed, lobulated growth pattern. White semi-translucent glistening areas were clearly seen, which were soft and gel-like, while peripheral areas appeared fibrous. The lesion appeared well demarcated but lacked a capsular tissue. An area of firm to gritty tissue was also noticed during sectioning.

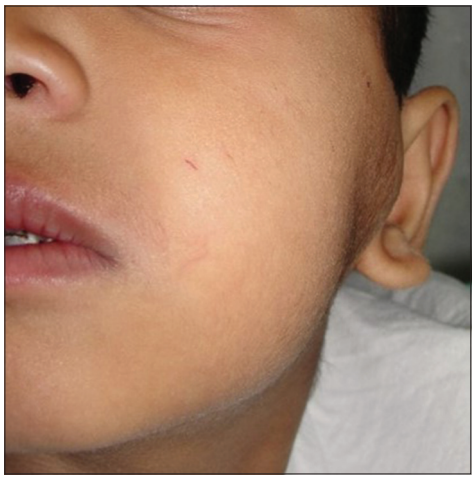

Figure 1: Extra oral photograph revealed a pre-auricular swelling on the left side of the face

Histopathology

On microscopic examination, the lesional tissue showed lobules with a hypocellular center showing myxochondroid appearance separated and surrounded by peripheral hypercellular areas. The tumor cells within the lobules were spindle and stellate-shaped with eosinophilic cytoplasm [Figure 4]. These cells were embedded in basophilic matrix showing myxoid change. Most of the areas showed delicate collagen fibers while few areas showed mature collagen fibers. A few foci of liquefactive myxoid areas were also seen.

The chondroid areas showed tumor cells residing in spherical and oval lacunar spaces surrounded by hyaline-type matrix [Figure 5]. The chondromyxoid areas featured a conspicuous vascularity around early attempts of primitive cartilaginous lacunae formation. There was no evidence of mitotic figures or pleomorphism in the tumor cells [Figures 6 and 7].

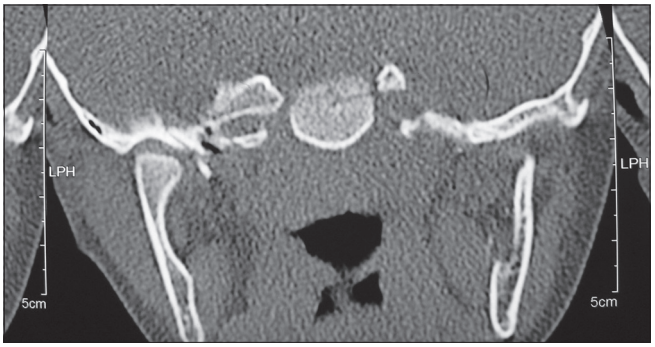

Figure 2: Computed tomography scan revealed radiodensity around the ramus of the mandible on left side

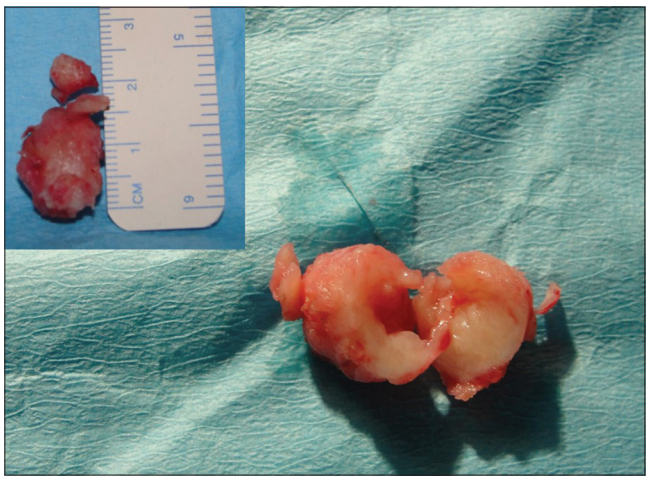

Figure 3: Gross image showing a lobulated mass with a glistening jelly-like surface and consistency

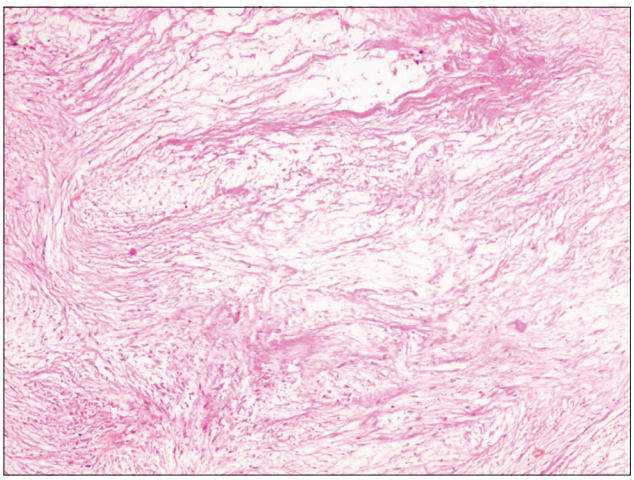

Figure 4: Myxoid areas with loose fibrillar collagen and inflammatory cell infiltrate (H&E stain, ×40)

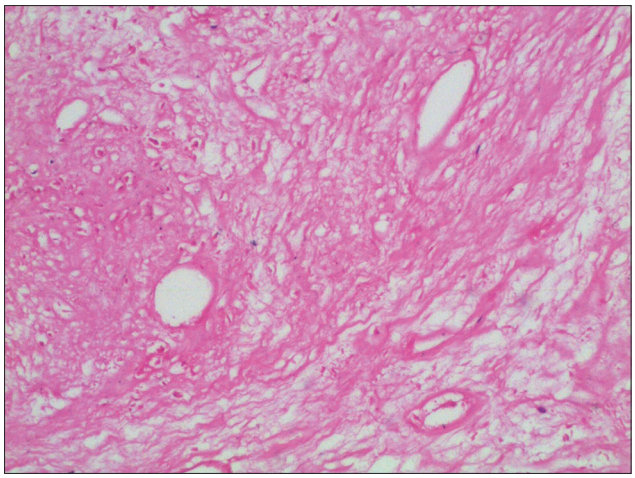

Figure 5: Foci of chondroid differentiation in a myxoid background with fibrillary collagen and few blood vessels (H&E stain, ×100)

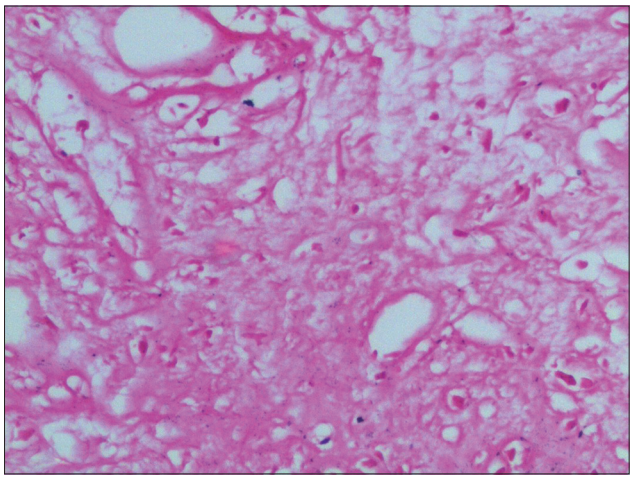

Figure 6: Foci of chondroid differentiation showing chondrocytes in lacunae and few blood vessels (H&E stain, ×200)

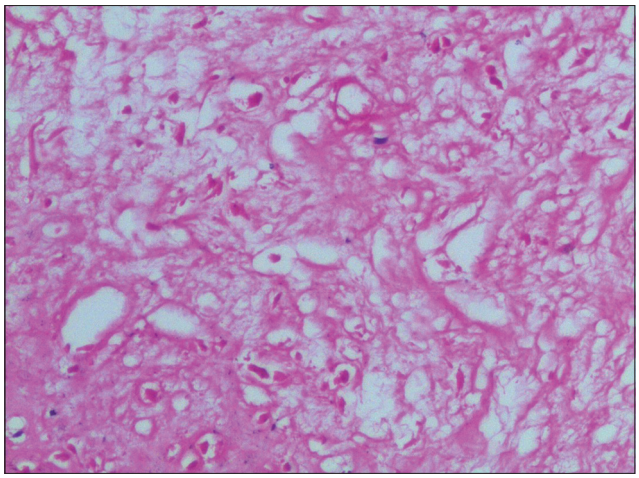

Figure 7: Myxoid component and chondroid differentiation showing chondrocytes in lacunae and few blood vessels seen (H&E stain, ×200)

A final diagnosis of CMF was arrived at with the help of CT scan, gross findings and histopathological picture.

DISCUSSION

Histopathologically, CMF is a rare benign tumor arising from cartilage forming mesenchymal tissue with a unique lobulated growth pattern and biphasic tissue distribution. Lischtenstein and Jaffey recognized this entity, which was named as CMF in 1948[4,7] and they reviewed 358 cases of CMF in different bones of the body evidently showing that none of the bone was immune to the occurrence of CMF.[1,2] The tumor accounts for 1% of all bone tumors[1] and approximately 2/3rd of all cases of CMF occur in the long bones of the body. Only 2% of cases of CMF involves facial and cranial skeleton. The craniofacial skeleton involved by CMF are maxilla, mandible, frontal, orbital floor, ethmoid, parietal, petrous, sphenoid, pterygopalatine fossa, mastoid, occipital and zygoma bones of skull.[2] The first reported case of CMF in the jaw bones was by Srivastava in 1955.[6] To our knowledge, approximately 35 intracranial cases and 25 cases involving the jaw bones have been reported, but only 2 cases involving the extragnathic site in the facial skeleton exclusive of cranium are reported.[2] Only two cases of CMF in the zygoma have been reported till date in English literature.[1,2]

In the reported cases, the age range varied from 1st to 8th decade of life with a peak in the 2nd decade of life. One case of congenital occurrence was noted. The female to male ratio is 2:1 when the cranium and facial bones are involved.[8]

The lesion commonly presents itself as a painful, slow growing mass with a span of 6 months to 2 years.[1,2] Radiologically, a non-specific appearance of radiolucent, lytic lesion is generally seen with partial or complete cortical erosion. Intra-lesional calcification and dense opacities are rarely seen. In our case, the lesion was totally symptomless and was noticed as an unesthetic bulge near zygomatic arch region. CT scan showed a lesion of 2.0 × 1.5 cm at the root of zygoma and a provisional diagnosis of osteoma was arrived at and lesion was surgically excised.

Although no specific cause is known for CMF, some authors have noted an association with certain chromosomal abnormalities. In a study of 4 patients with CMF, Granter and colleagues found that all of the subjects had a clonal rearrangement of chromosome 6.[9] Each of these rearrangements involved band 6q13, which has not been associated with other bone tumors.

On gross examination, the tumor was a well circumscribed, lobulated mass, firm, rubbery to soft in consistency. The cut surface of the lesion showed opalescent gray-blue areas resembling fibrocartilage. Glistening, homogenously white areas are seen when chondroid component prevail and gray-white and gel-like when the myxoid component predominates.[4] Predominant white gel-like areas were well appreciated in the lesional mass in lobules with intervening fibrous tissue in our case. A small area of firm to gritty tissue was also noticed during sectioning. Well lobulated and

circumscribed lesional mass was seen with lobular growth pattern.

Microscopically, a great amount of variability in presentation might be seen.[4] Histologically, a classical feature is a lobular pattern with stellate or spindle cells in a myxoid background. A proportion of chondroid, myxoid and fibrous tissue is seen arranged in lobules. The lobules have a hypocellular center and a hypercellular periphery. The hypocelluar areas are made of spindle cells and collagenous fibrous tissue. The hypercellular areas are made of the chondroid, myxoid or chondromyxoid areas. Frank hyaline cartilage foci are rare and only a few tumors show calcification and mineralization.[1,4] Liquefactive myxoid changes are also sparsely evident.

Histopathologically, CMF resembles chondroblastoma and chondrosarcoma.[1,2,4] A distinction between these tumors is vital. Misdiagnosis rates of 22-28% are reported.[2] Some features like radiographic presentation, histological presence of homogenous hypercellularity, pleomorphism and liquefactive changes of the matrix are helpful in drawing a distinction between CMF and chondrosarcoma.[3] Features like site of the lesion, along with histological features of calcification and periosteal reaction help in differentiating chondroblastoma from CMF. Lack of atypical mitotic figures in significant numbers and plump bizarre cells with multiple nuclei are useful criteria for distinguishing CMF from chondrosarcoma.[10]

Microscopic features in this case revealed a clear lobular pattern. The borders of the tumor mass were clear and well demarcated but unencapsulated. The tumor was made of chondroid, myxoid and fibrous tissue both in mature and immature forms. The periphery of the lobes were hypercellular and was made up of spindle-shaped cells with few areas of collagen formation. The central core areas were hypocellular and were made of chondromyxoid and myxoid areas. A good distribution of vascular capillaries was seen in the tumor mass. The chondroid areas were numerous and showed proper lacunae formation with chondrocytes entrapped within them. The mineralization was poor and incomplete. Areas with attempted cartilage formation were also seen. Based on these findings a diagnosis of CMF was made. Lack of hypercellularity in a uniform pattern, absence of pleomorphism, mitotic figures and liquefaction sites ruled out chondrosarcoma. Size, location of the tumor, growth pattern and age of the patient along with scan images helped in the confirmation of the diagnosis.

The treatment of choice for CMF is en bloc excision with normal bone margin. Recurrence rate of 25% has been reported and more so after a conservative curettage. When this tumor is small and localized in the facial skeleton, many authors recommend curettage followed by a strict follow-up to avoid cosmetic and functional sequelae of bone resection.[1,6] Malignant transformation of these cases is rare with no documented cases.[2,3] In the present case, the tumor neatly shelled out exposing the raw bone surface, showing an area of bone involvement. The involvement of a facial bone, site and size justified a conservative approach in our case.

The patient was asymptomatic and did not show any evidence of recurrence in a follow-up period of 14 months after surgery.

CONCLUSION

CMF is a rare benign tumor of the bone with a negligible malignant transformation rate. These tumors share the gross and histological presentations with a more aggressive and malignant tumor, chondrosarcoma. A detailed knowledge of the lesion will enormously help in proper diagnosis, choosing the right treatment modality leading to a better prognosis. A right conservative approach should be taken which shall be beneficiary to the patient.

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