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Chondromyxoid fibroma of the mandible: A report of rare entity

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ABSTRACT

Chondromyxoid fibroma (CMF) is an exceedingly rare benign tumor of cartilaginous origin accounting for less than 1% of all primary bone tumors and less than 2% of benign bone tumors. Chondromyxoid fibroma occurs mostly in the long bones of appendicular skeleton, but occurrence in craniofacial and gnathic regions is even rare. 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. A case of CMF involving the mandible in a 50 year old female patient is presented.

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1. Introduction

Chondromyxoid fibroma (CMF) is a benign tumor of cartilagenous origin and rarest of all bone tumors.[1] The tumor was first described by Jaffe and Lichtenstein in 1948 who differentiated the histologic findings of CMF from that of chondrosarcoma and enchondroma.[2] The tumor accounts for less than 1% of primary bone tumors categorized by Unni and Inwards and 1.6% of their catalog of benign bone tumors.[3]

The tumor originates from the physeal cartilage plate remnant with tibia being the most frequently affected bone (80%).[4] Its occurrence in the craniofacial bones and gnathic regions is least common. It is a slow growing, sharply demarcated tumor, which sometimes may behave in an aggressive way destroying trabecular bone and extending into the soft tissues.[5] It is associated with high recurrence rate of 10–80% with <2% risk of malignant transformation.[4]

CMF is more aggressive in younger patients with mean age of approximately 30 years.[4] The age of diagnosis is a factor for increased recurrence rate, with suggestion that the reduced resistance of the pediatric thin cortices and spongiosa contributes to the aggressive behavior of the lesion. In contrast however, it has been reported that there was neither difference in the rate of recurrence and the age of diagnosis, nor was there any correlation between the histological features and increased tendency to recur.[4] Hereby, we present a case of chondromyxoid fibroma occurring in the mandible in a 50-year-old female patient.

2. Case report

A 50-year-old female presented with a swelling on the left side of the lower jaw since one year. Initially, the swelling was of peanut size associated with mild dull pain and gradually

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Fig. 1 – Extraoral view revealing gross facial asymmetry over the left side of lower mandibular region.

increased to the present size measuring approximately 2 × 2 × 0.5 cm. There was no contributory past medical and dental history. General physical examination revealed no abnormality.

Extraorally, gross facial asymmetry was seen over the left side of the lower mandibular region, and the swelling was approximately 3 × 2 cm in size, extending anteroposteriorly 4 cm posterior to the angle of the mouth till 2 cm away from the angle of the mandible and superoinferiorly from the level of corner of mouth till the lower border of mandible (Fig. 1). On inspection, the swelling was roughly spherical with well-defined margins and a smooth surface. On palpation, all the inspectory findings were confirmed and the swelling was firm in consistency, nonfluctuant, and nontender. Intraorally, the swelling could not be differentiated from the surrounding mucosa.

Orthopantamography (OPG) revealed radiolucency in the region of swelling, irt 46 and 47 (Fig. 2). Magnetic Resonance Image (MRI) revealed tumor mass as hypointense and isointense images with the surrounding muscles and heterogeneously hyperintense on T2-weighted images (Fig. 3).

Incisional biopsy was performed, which revealed well-circumscribed lesion of fibrous connective tissue arranged in the form of fascicles. Numerous aggregates of histiocytes were arranged within the fascicles. Moderate amount of vascularity was seen in the form of stag-horn pattern. Inflammatory component was minimal suggesting the diagnosis as benign fibrous histiocytoma.

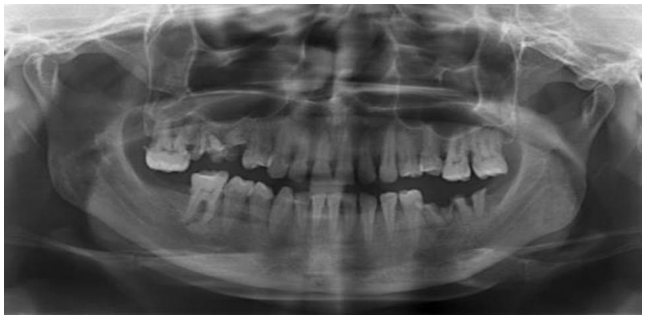


Fig. 2 – Orthopantamography revealing radiolucency in the region of swelling irt 46 and 47.

Complete surgical excision of the tumor was performed. Grossly, the tumor was approximately 2 × 2 × 0.5 cm, creamish white in color, and round to ovoid in shape, with irregular surface and firm consistency (Fig. 4). Histopathology demonstrated circumscribed lesion with numerous cells arranged in the form of fascicles. The cells are spindle-shaped proliferative fibroblasts embedded in the fibrous tissue with focal areas showing round cells. Intermixing with spindle cells, chondroid areas, and myxoid areas are also seen, suggesting the final diagnosis of chondromyxoid fibroma (Fig. 5).

3. Discussion

CMF is a rare benign and potentially aggressive bone tumor occurring primarily in the 1st to 2nd decades of life.[6] The common site of the tumor is the metaphysis adjacent to the epiphyseal growth plate. This is consistent with the hypothesis that the tumor arises from the remnants of cartilage at these sites.[7] CMF has a slight higher predilection for males. The male to female ratio is 2:1, but when cranium and facial bones are involved, male to female ratio is 1:2.[3] CMF has a predilection for the long bones with only 2% of the lesions involving craniofacial skeleton.[1,7] In the orofacial region particularly, this tumor occurs in the mandible. Till now, only 13 cases of mandibular CMF, 2 cases of CMF in the zygoma, and 3 cases of CMF in maxilla have been reported in the literature.[1,9] In this case, the patient was a female of 50 years age, and the lesion was located on the left side of the mandible.

No specific cause is known for CMF, but there is an association with certain chromosomal abnormalities.[1] Rearrangement of the long arm of chromosome 6 at bands q13 and q25 were the most frequent.[6]

Radiological appearance is characteristic, but is nonspecific with well-defined expansile radiolucent lytic lesion, which has scalloped and sclerotic borders with no intralesional calcifica-

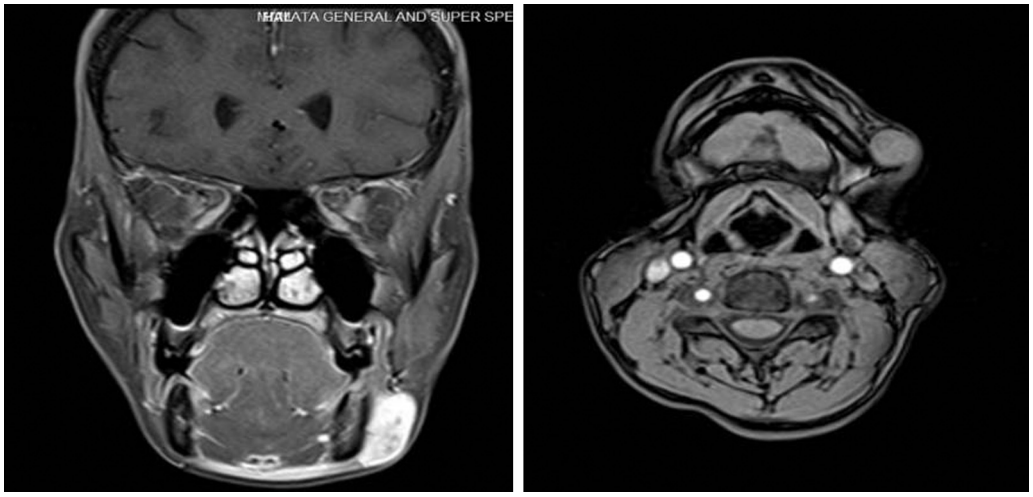


Fig. 3 – MRI coronal and axial view revealing tumor mass.



Fig. 4 – Grossing picture.

tions.[1,6] OPG revealed a radiolucent lesion in the region of swelling irt 46 and 47. MRI gives more detailed characterization with hypointense images on T1-weighted images and hyperintensity on T2-weighted images.[4] In this case, MRI disclosed benign soft tissue lesion.

Grossly, CMF reveals a well-circumscribed lobulated mass, which is firm in consistency. The cut surface of the lesion shows grayish blue areas resembling fibrocartilage. Glistening white areas are seen when chondroid component prevails and gray white gel-like area when myxoid component predominates.[1] In this case, white creamish gel-like areas were noticed amidst the fibrous tissue.

Histologically, the classical feature is lobular pattern with stellate or spindle cells in a myxoid background. A proportion of chondroid, myxoid, and fibrous tissue is seen arranged in lobules. These lobules have a hypocellular center and a hypercellular periphery. The hypocellular areas are made of spindle cells and collagenous fibrous tissue. The hypercellular areas are made of the chondroid, myxoid, or chondromyxoid areas. Hyaline cartilage foci are rare and only a few tumors

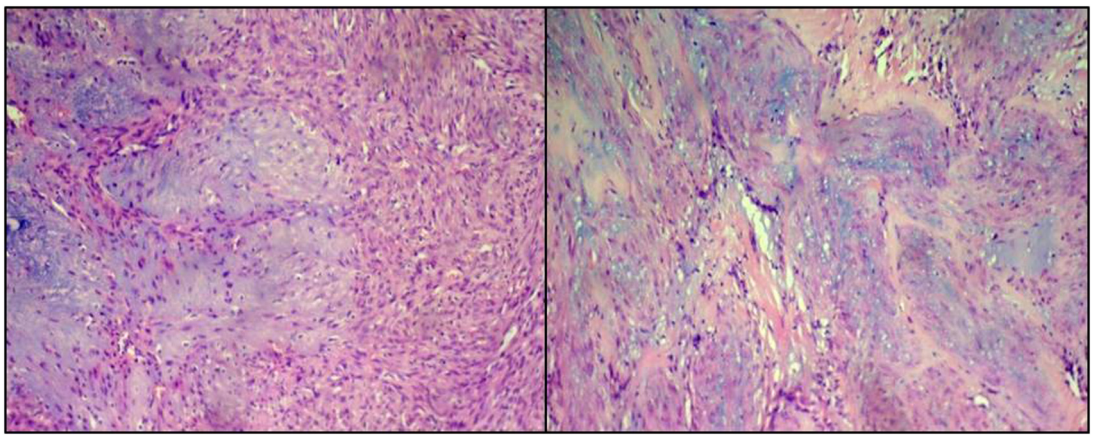


Fig. 5 – Histopathology revealing tumor cells arranged in the form of fascicles intermixed with chondroid and myxoid areas (H&E stain, 100× view).

Table 1 – Differential diagnosis of CMF.

Lesion	Chondromyxoid fibroma	Chondrosarcoma	Chondroblastoma	Fibrous dysplasia	Osteosarcoma	Chondroma
• Histological features	• Lobules of spindle-shaped or stellate cells.	• Well-formed hyaline matrix.	• Chondroblasts have eosinophilic, polygonal cytoplasm (not stellate as in CMF).	• Extreme myxoid change.	• Mitoses and nuclear atypia.	• Infiltrative margin.
	• Abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle-shaped or round cells.	• Absence of fibrous component.	• IHC is not helpful in distinguishing CMF from chondrosarcoma because both tumors express S-100, vimentin.	• Irregular woven bone.	• Diagnostic hallmark is osteiod formation, which is totally lacking in CMF.	• Large epithelial cells with eosinophilic or vacuolated cytoplasm.
		• IHC is not helpful in distinguishing CMF from chondrosarcoma because both tumors express S-100, vimentin.	• Calcification is a conspicuous feature unlike CMF.	• Loosely arranged fibrous stroma.		• Chordoma cells are typically arranged in cohesive nests and cords.
				• Chinese script pattern.		• Tumors express keratin and epithelial membrane antigen.
Radiographic features	• Lobulated outline with sharp margins and majority have a sclerotic rim.	• Osteolytic and intralesional calcifications.	• Cortical breach.	• Ground glass opacification with poorly defined margins.	• Mixed sclerotic or lytic lesion.	• CT scan – Well-circumscribed, lytic lesion.
	• Majority – Osteolytic and radiolucent.	• Endosteal scalloping.	• Endosteal scalloping.		• Tumor erodes through cortex causing elevation of periosteum.	• Expansile soft tissue mass.
		• Cortical remodeling.	• MRI is ideal.			• Intratumoral calcifications .
		• Moth-eaten appearance.				

show calcification and mineralizaton.[1] The present case also revealed spindle-shaped cells intermixed with chondroid and myxoid areas in the fibrous tissue stroma.

Diagnosis is based on location of the tumor, size of the tumor, age of the patient, and the radiological features. Differential diagnosis of CMF may include chondroblastoma, fibrous dysplasia, chondromyxoma-like chondroblastic variant of low-grade osteosarcoma, and high-grade myxoid chondrosarcoma (Table 1). A careful clinical correlation between radiographic and histopathological features decreases the risk of diagnostic errors. Wide excision of the tumor including a rim of normal bone is the treatment of choice for CMF patients.[8] Curettage alone has reported a recurrence rate of 25%.[1] Curettage combined with bone grafting decreased the recurrence rate to 7%.[2] In this case, the patient was treated by excision of the entire lesion. No recurrence is noticed till date on follow-up.

4. Conclusion

Rareness of the tumor makes their diagnosis difficult for both the clinician and the pathologist. Careful clinical management and follow-up are crucial for the successful outcome of the patients with this tumor.

Conflicts of interest

The authors have none to declare.

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