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Case Report

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Unmasking a Diagnostic Dilemma: A Rare Cartilaginous Tumor in the Maxilla

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Abstract

Tumors characterized by the production of chondroid matrix, at least focally, are grouped under cartilaginous tumors. It ranges from benign lesions to highly aggressive neoplasms. Benign cartilaginous tumors are usually asymptomatic, with overlapping histopathological features that pose diagnostic challenges for pathologists. A male patient in his late twenties presented with a painless swelling in the right palatal region for three months. The patient had a history of nasal congestion and breathing difficulties. On examination, a unilateral lesion of size 6x4x2.5 cm extended antero-posteriorly from the canine to the maxillary tuberosity and mediolaterally from the right alveolus to the mid-palatine raphe. The surface was lobulated without any ulceration and no evidence of tooth mobility. The orthopantomogram (OPG) revealed an unilocular, well-defined lesion with a sclerotic border. Computed tomography (CT) showed a lytic lesion with destruction of the maxillary sinus walls. The key differentiating feature in histopathology is the lobular arrangement of hyaline cartilage supported by a myxoid background with matrix calcification, without any evidence of tissue dysplasia. Immunohistochemistry findings were consistent with the diagnosis of Chondromyxoid Fibroma (CMF), effectively differentiating it from malignant counterparts such as chondrosarcoma. The subtotal maxillectomy (Transoral approach) was performed under general anesthesia. The current case discussed a benign cartilaginous tumor mimicking malignancy, which has an incidence of less than 1% of all bone tumors, highlighting the diagnostic complexity and importance of histopathological confirmation.

Keywords: benign cartilaginous tumor, chondroid matrix, chondromyxoid fibroma (CMF)

Introduction

Cartilaginous tumors share a common characteristic feature of the production of chondroid matrix at least at a focus, and these range from benign to highly lethal neoplasms.[1] It is categorized based on its neoplastic or non-neoplastic nature, composition (purely cartilaginous or not), and site of occurrence (intraosseous, juxtacortical, or soft tissue). The WHO in 2022 classified cartilaginous tumors into benign (enchondroma, osteochondroma, chondroblastoma, chondromyxoid fibroma, and synovial chondromatosis) and malignant lesions (chondrosarcoma and the chondroid variant of chordoma).[2,3] Benign cartilaginous tumors are usually asymptomatic, slow growing, locally invasive, and the histopathological features of these lesions mimic each other. It is difficult to excise the complete lesion, and en-bloc resection is usually recommended to overcome recurrence. Malignant transformation of benign cartilaginous lesions like enchondroma and osteochondroma never develops, and only 1-5% deteriorates to chondrosarcoma. These lesions in the head and neck are rare, posing significant diagnostic challenges due to their morphological heterogeneity because of the mesenchymal cell multipotency during differentiation.[4] Hereby, we present a rare case of Chondromyxoid Fibroma (CMF) in the maxilla of a young adult male to highlight the diagnostic complexity and importance of histopathological confirmation.

Case Report

A male patient in his late twenties presented with a swelling in the upper jaw present for three months. The swelling was insidious in onset, gradually increasing in size. The patient reported nasal congestion, sinusitis, and difficulty breathing. Extraordinarily, mild facial asymmetry was observed in the right maxillary region, with obliteration of the naso-jugal groove. Intraorally, a unilateral lesion measuring 6x4x2.5 cm was noted on the right palatal shelf, extending antero-posteriorly from the canine to the maxillary tuberosity and mediolaterally from the right alveolus to the mid-palatine raphe. The surface was lobulated, with no evidence of surface ulceration or tooth mobility (Figure 1). Gingival recession with root exposure was noted in the right lateral incisor and canine, without occlusal discrepancy. The lesion was firm to hard on palpation and mildly tender over the right molar area. Based on clinical features, a provisional diagnosis of ossifying fibroma was made, with differential diagnoses including minor salivary gland neoplasm, odontogenic tumor, fibrous dysplasia, osteosarcoma, and chondrosarcoma.

The orthopantomogram (OPG) revealed a unilocular, well-defined lytic lesion (radiolucent with mild opacity), with a sclerotic border in the right maxilla extending from mesial aspect right maxillary first premolar to distal aspect of right maxillary third molar. The upper limit extended to the superior wall of the maxillary sinus. There is no evidence of cortication of bone. The root apices of first and second maxillary molars and interdental bone appeared to be resorbed (Figure 2A). The 3D reconstructed image of computed tomographic (CT) image reveals loss of trabecular pattern of alveolar bone extending from the facial aspect of right maxillary first premolar till the maxillary tuberosity. The soft tissue window contrast enhanced axial section showed focal destructive mass extending from the infra-zygomatic surface to the posterior maxilla, with nasal septum deviation and matrix calcification (Figure 2B). Radiological differential diagnoses included fibrous dysplasia, non-ossifying fibroma, and benign cartilaginous tumor.

Incisional biopsy showed a greyish-white soft tissue measuring 0.8x0.9 cm, firm to hard in consistency. Gritty consistency was noted during grossing. The cut surface showed whitish-coloured lobules with hemorrhagic areas. The histopathology revealed lobular arrangements of hyaline cartilage supported by a myxoid background. Cells were situated in lacunae, arranged singly or in clusters, with occasional multinucleation. Peripheral fibrous condensation and Basophilic stippled calcifications were noted, with no evidence of mitotic activity or necrosis (Figure 3). Based on the clinical, radiographical and histopathological features, the lesion was consistent with chondromyxoid fibroma. Immunohistochemical analysis was performed to confirm the diagnosis and rule out malignant differentials. The tumor cells showed strong cytoplasmic positivity for S-100 and SOX9, indicating chondrogenic differentiation, while diffuse vimentin positivity confirmed their mesenchymal origin. The Ki-67 proliferative index was low (<5%), supporting a benign nature, and CD34 and Bcl-2 negativity ruled out vascular or aggressive neoplasms. Additionally, p53 expression was absent, further supporting the benign histopathological profile of the lesion. These findings were consistent with

Chondromyxoid Fibroma (CMF), effectively differentiating it from potential malignant counterparts such as chondrosarcoma. The choice of treatment was under general anesthesia to avoid recurrence. The excisional specimen was consistent with the histopathological findings of the incised specimen (Figure 4). Post-operatively, there was no evidence of recurrences for the past 1 year, but the patient had issues of difficulty with mastication and aesthetic concerns that were corrected by prosthetic replacement. (Figure 5)

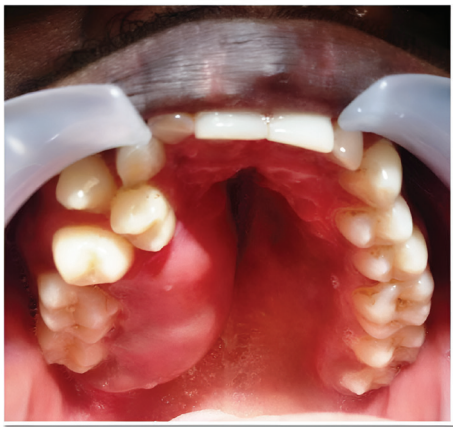

Figure 1: Intra-oral swelling extending mediolaterally from the right alveolus to the mid-palatine raphe.

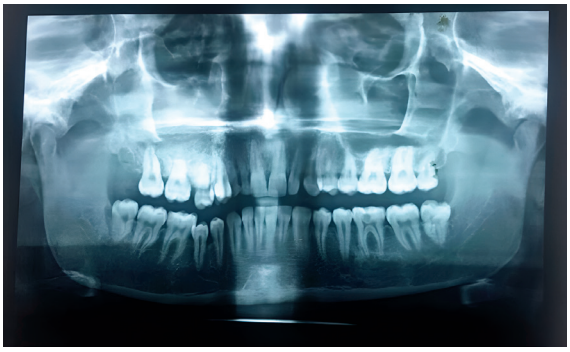

Figure 2A: A well-defined, unilocular lytic (radiolucent with mild internal opacity) lesion with a sclerotic border is noted in the right maxilla. The lesion extends from the mesial aspect of the right maxillary first premolar to the maxillary tuberosity. Superiorly, it reaches the superior wall of the maxillary sinus. The root apices of the first and second maxillary molars, as well as the surrounding interdental bone, appear affected

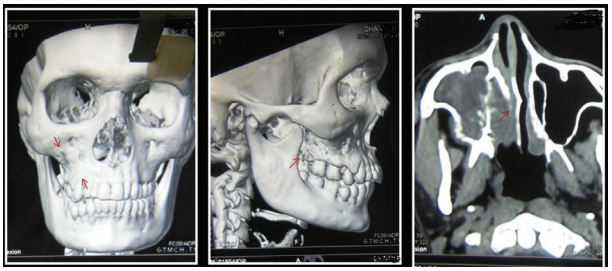

Figure 2B: 3D reconstructed CT images reveal loss of the normal trabecular pattern in the alveolar bone from the facial aspect of the right maxillary first premolar to the right maxillary third molar, suggesting involvement of a pathological lesion in this region (left). A focally destructive mass is observed extending from the infrazygomatic surface toward the posterior surface of the maxilla (middle). A diffuse lesion is also noted within the left maxillary sinus with deviation of the nasal septum (right).

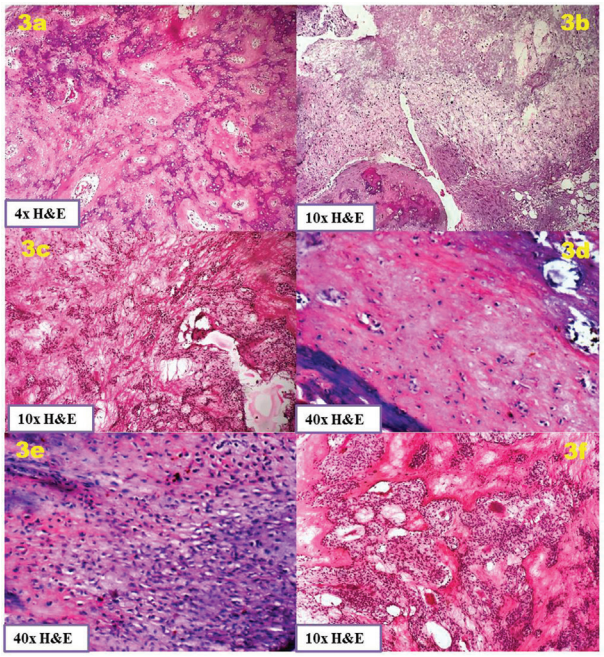

Figure 3: (3a) Photomicrograph revealed lobular pattern of arrangement of hyaline cartilage supported by a fibroid and myxoid background; (3b) predominant myxomatous areas with stellate shaped cells; (3c) exhibits fibroblastic proliferation with round to oval shaped cells. [10x objective, 10x eyepiece & H&E]; (3d) central lobular areas exhibit cartilaginous matrix with round to oval nuclei along with basophilic stippled calcifications; (3e) showing predominant areas of chondroid differentiation with well-formed lacunar spaces and chondrocytes residing in lacunae was evident [40x objective, 10x eyepiece & H&E]; (3f) revealed confluent nodular architecture with intervening collagen fibers and evident of osteoclast- like giant cells [10x objective, 10x eyepiece & H&E].

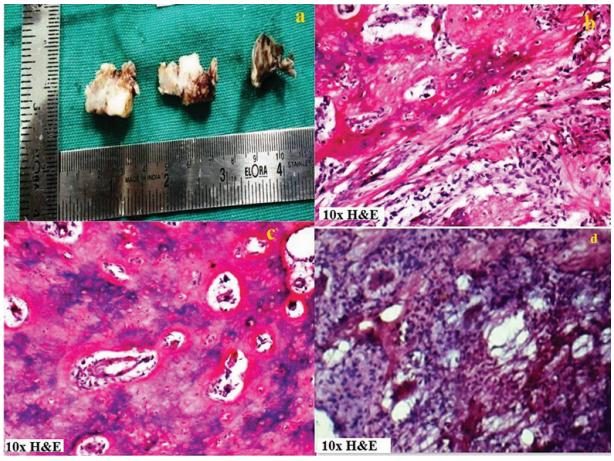

Figure 4: Grossing picture of excisional specimen. (a) and the photomicrograph showing lobular pattern of arrangement of hyaline cartilage supported by a myxoid background within this loosely arranged stellate shaped cells. Central lobular areas exhibits cartilaginous matrix showing round to oval nuclei; (b, c) The osteoclast-like giant cells are evident; (d) 10x objective, 10x eyepiece & H&E.

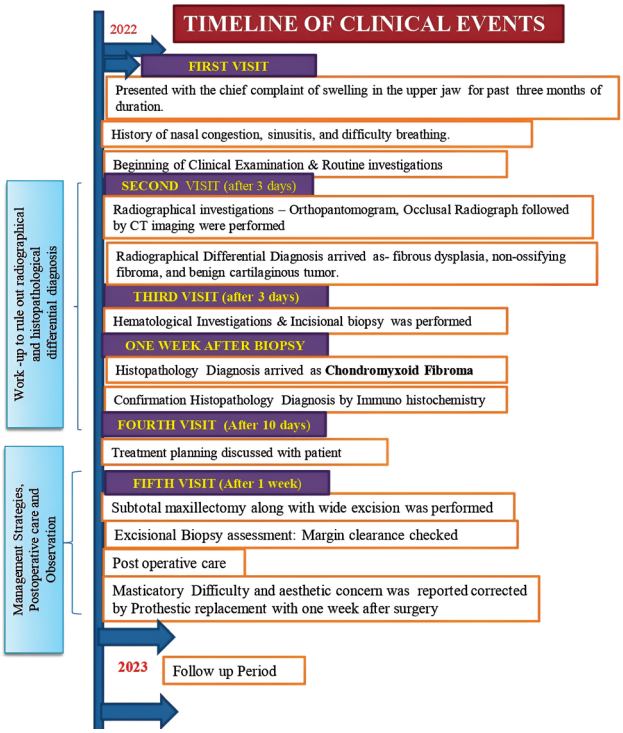

Figure 5: Clinical timeline events.

Discussion

CMF is a rare benign cartilaginous or chondrogenic tumor that predominantly affects the proximal metaphyseal region of long bones, such as the tibia, femur, and fibula, with approximately 25% occurring in flat bones.[3] It has characteristic histological features, including a lobular architectural pattern in a chondro-myxomatous background. Initially described by Jaffe and Lichtenstein in 1948, CMF was identified as a distinct entity that which may mimic chondrosarcoma histologically.[5] According to the World Health Organization (WHO), CMF is defined as a "benign tumor characterized by lobules of spindle-shaped or stellate cells proliferating within an abundant intercellular matrix that is myxoid or chondroid in nature."[6] It accounts for 1-1.8% of all bone tumors and 2-5% of tumors in the craniofacial region. The first case of CMF involving the anterior mandible in a young woman was reported by Srivatsava in 1955.[7] More recent literature highlights the rarity of CMF in the craniofacial region, with only 25 cases involving jawbones, 2 cases in the zygoma, 1 case in the pterygopalatine space, and 35 intracranial localizations reported.[3]

Despite its benign nature, CMF poses a diagnostic challenge due to similar lobular architecture, cellularity or calcification patterns with other cartilaginous tumors especially chondrosarcoma particularly when occurring in unusual locations. CMF primarily manifests in the second to third decade of life, with no significant sex predilection. This report underscores an exceptionally rare presentation of CMF in the maxilla of a young male, a location seldom reported in the literature. Cytogenetic analysis has revealed recurrent clonal abnormalities involving chromosome 6, particularly the rearrangement of the long arm at bands q13 and q25. These regions harbor genes such as BMP6, COL9A1, and COL10A1, along with oncogenes like FYN and ROS1, which are associated with cartilage development.[8,9] The repeated genetic rearrangements and over expression of glutamate receptor gene GRM1 are consider as key driver in the development of CMF. The comprehensive radiographical and histomorphological assessment combined with GRM1 immunohistochemistry helps us to distinguish the CMF from other histological mimics.[10]

Clinically, CMF typically presents as a painful, slow-growing swelling over a period of 6 months to 2

years. The pain is usually mild, intermittent, and dull in nature, as seen in this case. However, in rare sites like the hands or feet, painless swelling may be the primary complaint. Occasionally, the tumor may be asymptomatic and discovered incidentally on radiographs.[3,8] Radiologically, CMF appears as a radiolucent lesion, often eccentric, affecting the metaphyseal region with sharply demarcated and scalloped margins that aid in diagnosis and monitoring. The calcification of matrix is rare but more frequently observed in craniofacial CMF than in CMF affecting long bones.[8,9] Advanced imaging, such as CT and Magnetic Resonance Imaging (MRI), offers detailed insights into the extent of bony destruction and adjacent tissue involvement. MRI findings typically show a lobulated pattern with hypointense signals on T1-weighted images and heterogeneously hyperintense signals on T2-weighted images.[11] In this case, the lesion extended to the maxillary sinus, causing nasal congestion and difficulty breathing, with evident matrix calcification on CT.

Histopathological examination remains the gold standard for diagnosing CMF. The lesion typically exhibits hypocellular lobules of stellate or spindle-shaped cells set in a chondro-myxomatous intercellular matrix. These lobules are separated by a highly cellular connective tissue stroma, which contains spindle-shaped cells and thick fibrous bands, along with variable numbers of multinucleated giant cells.[3,9] Dahlin emphasized the presence of giant cells at the periphery of the chondroid lobules with plump, hyperchromatic nuclei as a characteristic feature of CMF.[12] Given its histological resemblance to other entities, differential diagnoses include chondroblastoma, chondrosarcoma, enchondroma, and aneurysmal bone cyst.[11] However, the unique histological features of CMF aid in distinguishing it from these lesions. IHC is a valuable tool in identifying the specific proteins in the diagnosis and differentiating CMF from other tumours.[13] In the present case, the tumor cells diffuse vimentin positivity followed by strong cytoplasmic positivity for S-100 and SOX9, indicating cartilaginous origin. The proliferative index of Ki-67 was low (<5%), supporting a benign nature of the lesion, and CD34 and Bcl-2 negativity ruled out vascular or aggressive neoplasms. These findings were consistent with Chondromyxoid Fibroma, effectively differentiating it from malignant counterparts such as chondrosarcoma.

Despite being benign, CMF can recur, particularly after incomplete excision, with recurrence rates reported as high as 25%.[14] The recurrence risk of CMF, depending on the treatment modalities, includes curettage, curettage with adjuvants like phenol applications and bone cementation, and surgical excision. The curettage alone showed a relatively high recurrence rate of up to 80%. The combination therapy with bone grafting reduces the recurrence rate by around 22%. The En-bloc resection with wide margins is a more aggressive approach with subsequent functional deficit and cosmetic concern, but preferred treatment of choice for a better prognosis and reduces the recurrence risk by 4%.[15] However, in the craniofacial region, complete excision is often challenging due to anatomical complexity and proximity to vital structures.[15,16] The additional factors tumor in younger age, tumor location, and surgical clear margins, increase the recurrence risk. In the present case, subtotal maxillectomy (Transoral approach) was performed without any complications. The absence of mitotic figures and necrosis in the excised tissue indicates a favorable prognosis. However, long-term follow-up with clinical and radiographic evaluations is essential due to the potential for recurrence.[16]

Conclusions

Diagnosing CMF in atypical locations, such as the craniofacial region, remains challenging due to its overlapping clinical and radiographic features with other benign and malignant tumors. The differential diagnosis includes ossifying fibroma, odontogenic tumors, fibrous dysplasia, or even malignancies like chondrosarcoma and osteosarcoma to be ruled out while diagnosing the lesion. Radiographically, CMF may mimic fibrous dysplasia or non-ossifying fibroma due to its well-defined, lytic appearance with a sclerotic border and mixed trabecular patterns. However, histopathology, as seen in this case, provides definitive confirmation of the diagnosis, with the absence of mitotic activity, necrosis, and dysplasia confirming its benign nature.

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or public health policy.

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Conflict of Interest

The authors declared that they have no conflict of interest.

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