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CASE REPORT

Cartilage Analogue of Fibromatosis in the Maxillary Alveolar Bone

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Abstract Cartilage analogue of fibromatosis is a relatively common tumor in the palms and soles of young children and adolescence. The characteristic histological finding of the tumor is the differentiation toward cartilage formation within a background of fibromatosis-like growth. Therefore, the tumor may cause potential diagnostic problems in the maxillofacial bones where it has not been described. The purpose of the present report is to describe the imaging characteristics, histological and immunohistochemical features of an extremely rare case of cartilage analogue of fibromatosis in the maxillary alveolar bone of a 25-year-old female.

Keywords Cartilage · Fibromatosis · Chondrosarcoma · Myxoma

Introduction

Calcifying juvenile "aponeurotic" fibroma was first described as a distinct entity by Keasbey in 1953 under the terminology of "Juvenile aponeurotic fibroma (calcifying fibroma) in the palms and soles of young children [1]. It is also described under the terminology of calcifying analogue of fibromatosis (CAF) as it neither represented fibroma nor restricted to the palms and soles of the young children [2]. The tumor usually arises in deeper paraskeletal or fascial tissues, but without any connection to the underlying bone [1–4]. The CAF both radiographically and microscopically manifests stippled calcification [1, 2] and thus poses considerable diagnostic challenges in sites where it is not known to exist or described as yet in the literature. The purpose of the present paper is to report the imaging characteristics, histological and immunohistochemical features of an extremely rare case of cartilage analogue of fibromatosis in the maxillary alveolar bone of a 25-year-old female.

Case Report

A 25-year-old female presented with a swelling and associated mild pain of 5-month duration over the left maxillary alveolar process in the region of premolar and molars. She disclosed no significant past medical history, but her past dental history revealed she had undergone extraction of left maxillary third molar 7 months prior. On intraoral examination, a smooth surfaced swelling was noted around the premolar and molar region with displacement of the second molar (Fig. 1a). There was no ulceration, thrills or bruit. There was no bleeding on provocation or tenderness on palpation. No other significant changes were evident.

Conventional radiograph revealed a soft tissue shadow with irregular streaks of opacification in the edentulous third molar region with displacement of the second molar and obliteration of the lamina dura and periodontal ligament space (Fig. 1b). The axial and coronal sections of the computed tomography (bone window) showed a well-defined, soft tissue mass displaying discrete and stippled calcifications with destruction of the cortical plates and extension into the left maxillary sinus (Fig. 2a–d). The differential diagnosis included ossifying fibroma, calcifying epithelial odontogenic tumor, odontogenic myxoma

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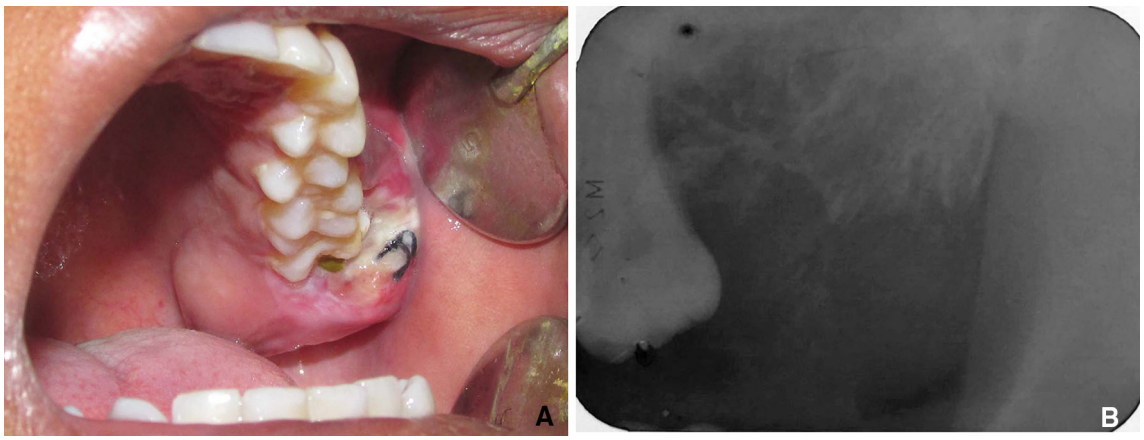

Fig. 1 **a** The lesional mass in the region of left maxillary molar tooth. **b** Reveals a soft tissue shadow with fine streaks of opacification without evidence of lamina dura or periodontal ligament space of the second molar

and chondrosarcoma. Incisional biopsy was performed which revealed a fibromyxomatous background with spindle- and stellate-shaped cells along with residual bone. The lesion was interpreted as odontogenic fibromyxoma. Based on the histological diagnosis, the extent of the lesion was assessed from the CT scan to mark the lesion for surgical removal. Bupivacaine 0.5% with adrenaline 1 in 200,000 was used for both buccal and palatal infiltration. A vestibular incision was made, and the mucoperiosteum was reflected using periosteal elevator to expose the anterior and lateral walls of the maxilla and as well as the zygomatic buttress. An osteotomy cut was made on the buccal side, and the vertical cut was made between the second premolar and molar. The palatal cut was completed through the buccal cut using an osteotome. The entire segment was removed along with lesion, and an obturator was placed. The surgical specimen was cleaned with normal saline and placed in formalin for histopathological evaluation.

On gross examination, the soft tissue mass appeared fibrous with no evidence of mucinous or myxoid change. The mass appeared to be attached to the second molar tooth (Fig. 3). Microscopic examination showed spindle-shaped cells arranged in a fascicular growth pattern within a background of fibrous to myxoid stroma. Interspersed within the tumoral mass were islands of fibrocartilage with calcification and residual or reactive bone formation. No pleomorphism or mitotic figures were noted (Fig. 4a, b). The microscopic features were consistent with cartilage analogue of fibromatosis. Immunohistochemistry was performed for markers such as cytokeratin, EMA, S100, vimentin, CD68 and CD99, but were negative except for positive reaction with vimentin (Fig. 4c). The postoperative event was uneventful, and the there was no recurrence since 2 years.

Discussion

The term calcifying juvenile aponeurotic fibroma was mainly adopted to distinguish it from other fibromas that it does not resemble [1]. Subsequently, Lichenstein et al. [2] proposed the term cartilage analogue of fibromatosis as the appropriate term and he believed that CAF is a tumor-like nodular proliferation of peculiar spindle connective tissue cells showing focal chondroid differentiation at sites of calcification and occasionally, even frank cartilage change. The latter finding is regarded as distinguishing feature of this entity [2].

The CAF evolves as a gradual, small, ill-defined, soft tissue mass without any apparent history of trauma. The tumor occurs over a wide age range (2–67 years) with a 2:1 male-to-female predilection [5]. Involvement of the head and neck is considered rare with only few cases reported in sites such as neck, scalp and medial pterygoid muscle and in relation to the jaw bone [4, 6].

The case described here appeared as a soft tissue lesion with fine streaks of calcification on conventional radiograph (Fig. 1b). On computed tomography, it appeared as a soft tissue density with calcification causing osteolysis of the alveolar bone and extension into the maxillary sinus (Fig. 2). However, in the original description [1], the tumor was described as a relatively radio-opaque soft tissue mass with fine stippling, but without associated bone involvement.

The characteristic microscopic findings in CAF are one of a fibromatosis-like growth but in which there is a tendency for chondroid or cartilage differentiation associated with mineralization [1, 2]. It has been observed that in any given tumor, depending upon the predominance of the cellularity, collagen fibers and foci of cells within the lacunae may lead to an erroneous impression of fibroma,

Fig. 2 **a**, **b** Axial sections (soft tissue window) reveal a soft tissue mass with scattered hyperdensity and cortical break. **c**, **d** Extension of the mass with stippled calcification into the maxillary sinus

fibromatosis, chondroma, pseudosarcoma and even chondrosarcoma or fibrosarcoma [2, 4, 5]. Although myxoid change has not been reported in the initial report by Keasbey [1] and Lichenstein [2], it is not uncommon to find loose and myxoid stroma [5, 7]. The latter finding leads to a diagnosis of fibromyxoma, based on the incisional biopsy sample, in the present case. However, the overall microscopic features of the present case are consistent with the description found in the literature for CAF (Fig. 4a, b). The immunohistochemical profile of the tumor would neither differentiate cell lineage nor distinguish it from other tumors [5]. Thus, the tumor cells may react non-specifically but consistently with vimentin, S100, CD68 and CD99, variably with EMA, SMA and CD34 and negatively with CK and desmin [5]. Immunohistochemistry in the current case revealed negative reaction except for vimentin (Fig. 4c).

The current case is interesting because it is the first case of CAF in the maxillary alveolar bone. Therefore, pertinent corroborative information is required in view of the

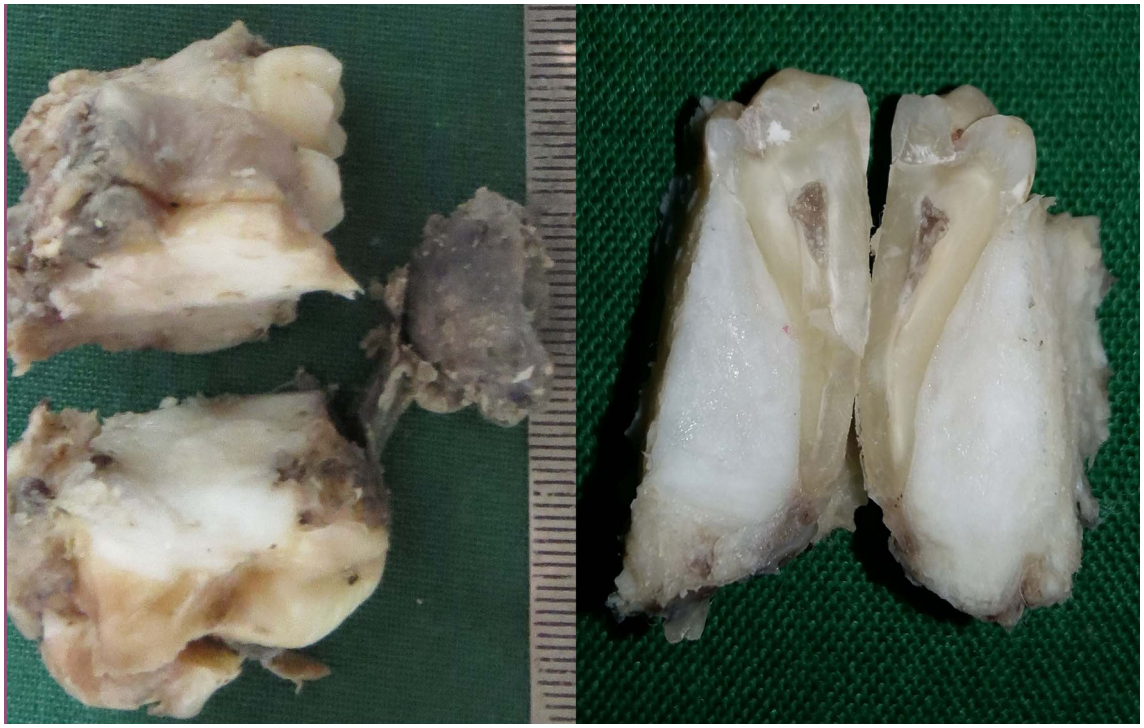

Fig. 3 Entire excisional specimen and the cut surface along with the attached tooth

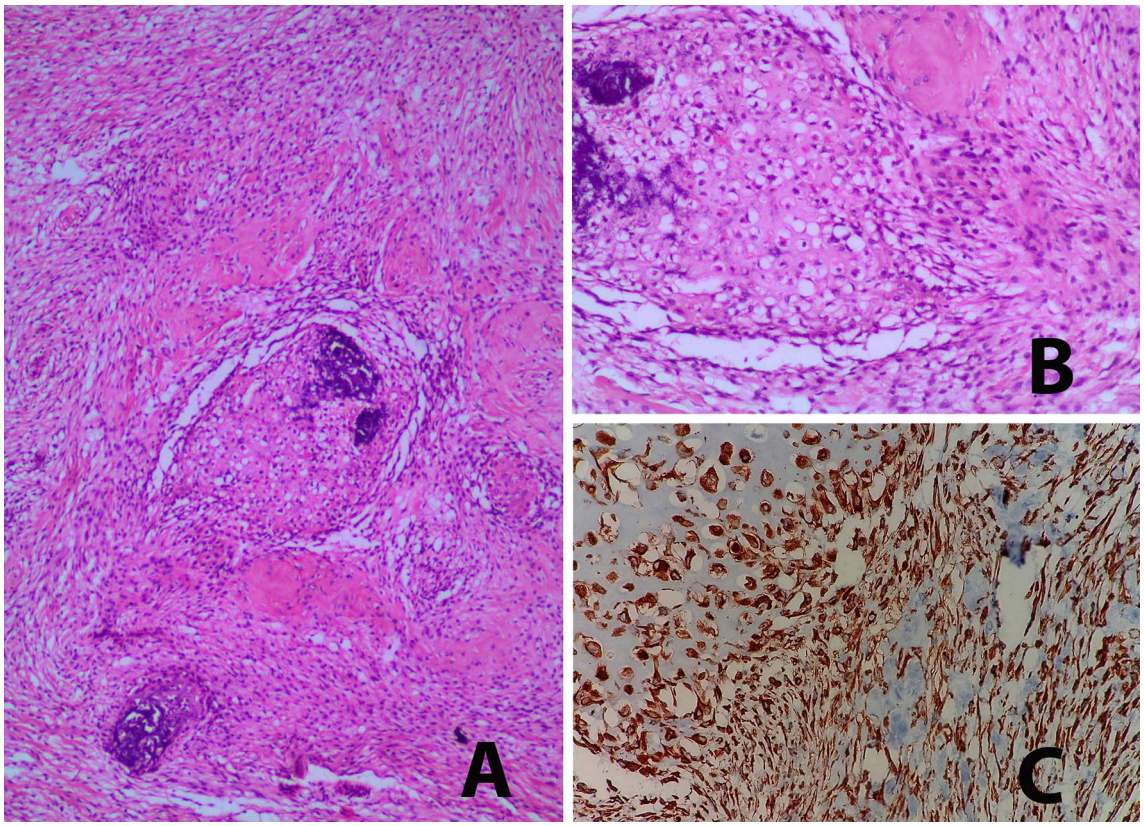

Fig. 4 **a** A fibromatosis-like growth pattern and nodules of fibrocartilage with secondary calcification, and the higher magnification of the cartilage is shown in **b**. **c** Positive reaction to vimentin in both spindle cells and in the fibrocartilage. [H & E, ×40, ×400, Vimentin, ×400]

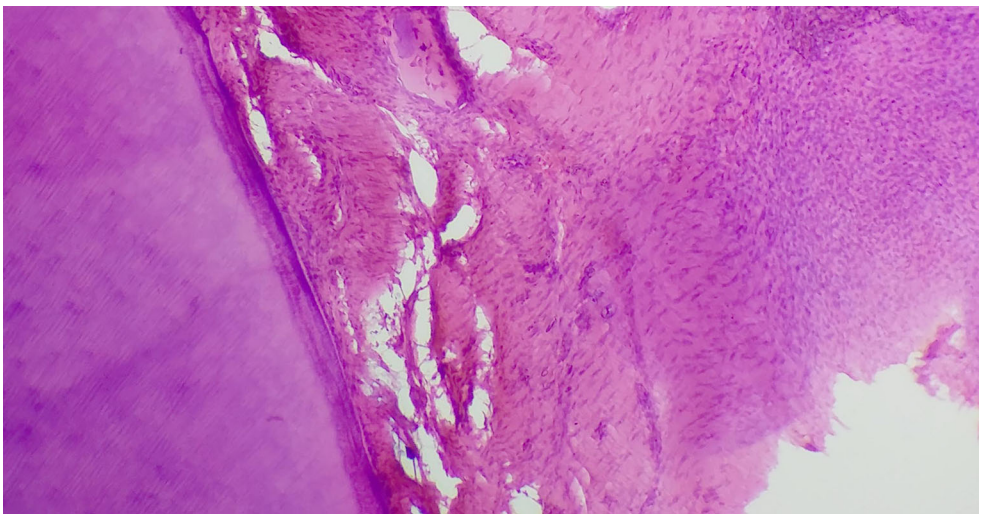

Fig. 5 Decalcified section of the specimen is shown in Fig. 3, which shows the transition of the lesion from the fibers of the periodontal ligament. [H & E, ×100]

reported lack of bone involvement in CAF [1, 2]. However, tumors consistent with CAF have been described in the cranial bones of dogs and also in the bones of humans [7–10]. It has been suggested by proponents of CAF that at the point of insertion of Sharpey's fibers into the bone or cartilage surfaces, there occurs subtle transition from a fibrous connective tissue composition to one of fibrocartilage [11]. The human periodontium is a specialized structure which supports the tooth in the socket through insertion of Sharpey's fibers between cementum of the tooth and alveolar bone. Further, according to Sicher's, as quoted by Everett et al. [12], *"There are so many undifferentiated cells in the periodontal ligament that I am not surprised that they may produce cartilage at one time or another."* The same view holds good for the current case as it appears to be attached to the root of the second molar tooth (Figs. 3, 5). In addition, review of the literature shows that it is not uncommon to find fibrocartilage in the human periodontium which occurs by the replacement of principal fibers of the periodontal ligament by fibrocartilage juxtaposed between the cementum and alveolar bone [12].

Although this tumor is believed to be a self-limited benign tumor, it has a tendency to recur after primary surgical excision [1]. Therefore, the recommended treatment is conservative surgical excision.

Conclusion

This report presents an extremely rare case of CAF in the maxillary alveolar bone that has the potential to be misdiagnosed as any other closely simulating tumor that may frequently occur in the jaws.

Compliance of Ethical Standards

Conflict of interest The author declares no conflict of interest.

Ethical Approval All procedures performed in this study involving human subject was in accordance with ethical standards of the institutional ethical committee.

Informed Consent Informed consent was obtained from the patient in this study.

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