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Case report

Solitary central myofibroma of the maxilla: A case report

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ABSTRACT

Myofibroma is a benign mesenchymal neoplasm manifests as solitary lesion in soft tissue, rarely found in bone and internal organs. It is sometimes referred as myofibromatosis with appearance of multiple lesions which may involve visceral parts. Myofibroma is mainly diagnosed in infants and it is rarely develop in adults. It is asymptomatic curable lesion with clinical presentation of solitary\multiple nodules containing spindle cells. The bony lesions within the skull had a lytic or a cystic appearance which generates displacement and mobility of teeth as well as expansion of the mandible and swelling. There are few publications regarding the central (bony) type of myofibroma, all suggests solely involvement of the mandible, with no evident for maxillae origin. The following report is a unique case of isolated central maxillary myofibroma in a 9-year-old Caucasian female. Clinical, radiological and histological findings as well as deferential diagnosis are discussed.

1. Introduction

Myofibroma is an uncommon tumor typically originates from soft tissues and subcutaneous sites in the head and neck region, and rarely arises within the bone [1–7], as only a few sporadic cases were reported of this intrabony occurrence in the jaws [6,7,9–11].

The appearance of multiple myofibroma lesions is less common, and is often referred to as "myofibromatosis". Occasionally myofibromatosis may appear in a disseminated form, characterized by lesions found in: lungs, kidneys, pancreas, gastrointestinal tract and bone [9].

Myofibroma can afflict people at all ages from birth [1–8] to 84 years old [10]. Familial pattern of inheritance was suggested [8,11], however the exact etiology remains unknown [1,3,10].

Overall, this lesion is asymptomatic [7–10]. The most reported clinical presentation is swelling [1,6,9,11] and to a lesser extent the presence of an exophytic oral mass [6,9] (usually evolves on the alveolar ridge or gingiva due to cortical perforation), thus, displacement and crowding of teeth is a common clinical finding in central myofibroma [9]. Of note, no ulceration of the overlying oral mucosa was ever described in central myofibroma cases.

Allon et al. reported 4 cases of central mandibular myofibroma and reviewed the literature of similar cases. The latter study revealed 23 cases of central mandibular myofibroma, however no involvement of the maxillary bone has ever been reported [9]. Additional 3 cases of central myofibroma were published in the English literature [10,11] none of which were in the maxilla. Of note, Mynatt et al. published 24 cases of myofibroma in the peri-orbital region [12], of which 3 extended to the oral cavity by invading the maxilla, however this lesion did not originate from the maxilla as in this case. Moreover Foss and Ellis reported 4 maxillary cases, again none of which were central.

Overall, no report of central myofibroma in the maxilla was ever described, herein we report, for the first time a patient with solitary maxillary bone derived myofibroma.

2. Clinical presentation

Nine-year-old Caucasian female with a chief complain of intra oral mass over the right maxilla was presented to the Oral and Maxillofacial Surgery Department, Hadassah Medical Center, Jerusalem. The swelling was asymptomatic and was first noticed by her parents 3 months prior to her evaluation in our clinic due to the child's complain of a change in her bite; however, no medical consultation was done.

The swelling was firm and localized. It measured 3 × 2 cm, with pink intact mucosa that involved the right upper jaw including expansion of the buccal and palatal aspects. The maxillary teeth on the right side were displaced and slightly mobile however asymptomatic upon palpation or percussion (Fig. 1A and B). Panoramic X-ray and CT scan were preformed and diagnosed.

Comparison to the contra lateral side using the panoramic X-ray revealed a distally displaced upper right second molar and apically

☆ AsianAOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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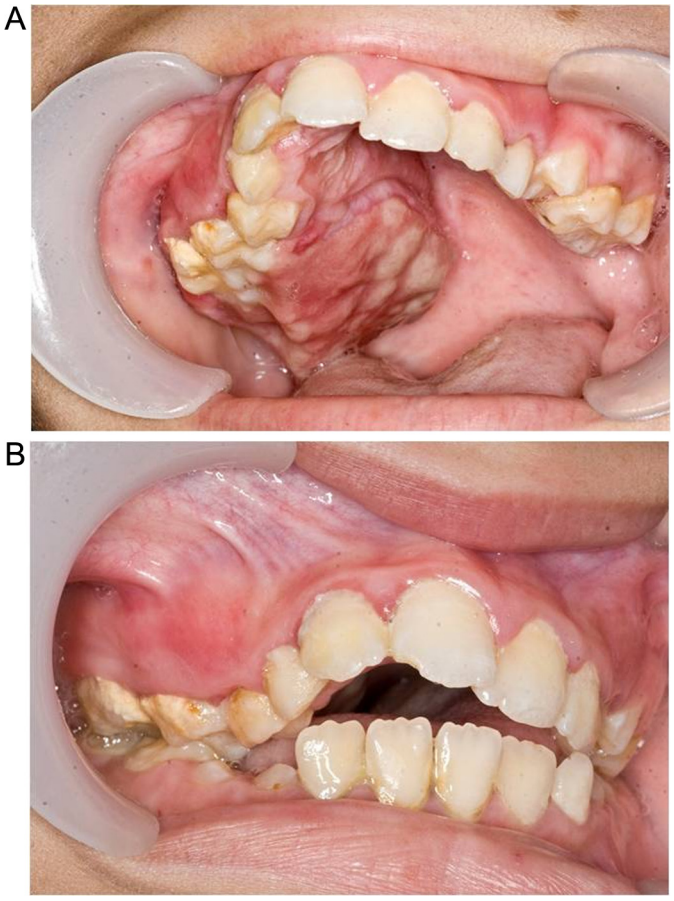

Fig. 1. A: Palatal swelling occupying the right maxilla. Notice the displaced teeth on the same side. B: Buccal swelling of the right maxilla.

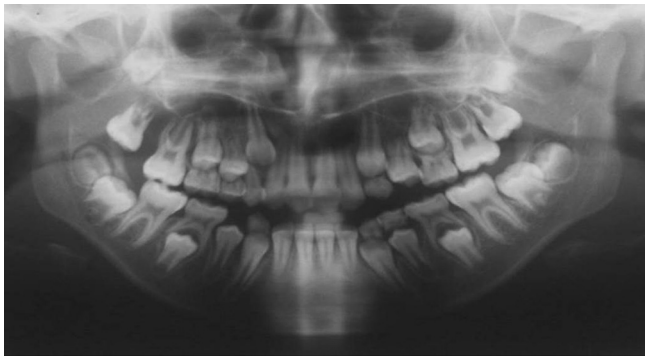

Fig. 2. Panoramic X-ray showing the distally displaced upper right second molar and the apically displaced right upper third molar tooth bud compared to the opposite side.

displaced third molar tooth bud, as well as "cloudiness" appearance of the upper right maxillary sinus (Fig. 2).

Noted throughout the CT scan, a low density lesion was surrounding the roots of the right upper maxillary teeth distally to the right lateral incisor, engendering an apical displacement of the right third molar tooth bud in addition to an obliteration of the right maxillary sinus. There was no involvement of the right orbital floor and the superior border of the lesion was in level with the middle concha (Fig. 3A–C).

Incisional biopsy was obtained from the palatal aspect of the mass and revealed fascicles of spindle shaped cells with pale pink cytoplasm, elongated, tapering nuclei with a vesicular chromatin and indiscernible nucleoli (light-stained areas, Fig. 4A). In

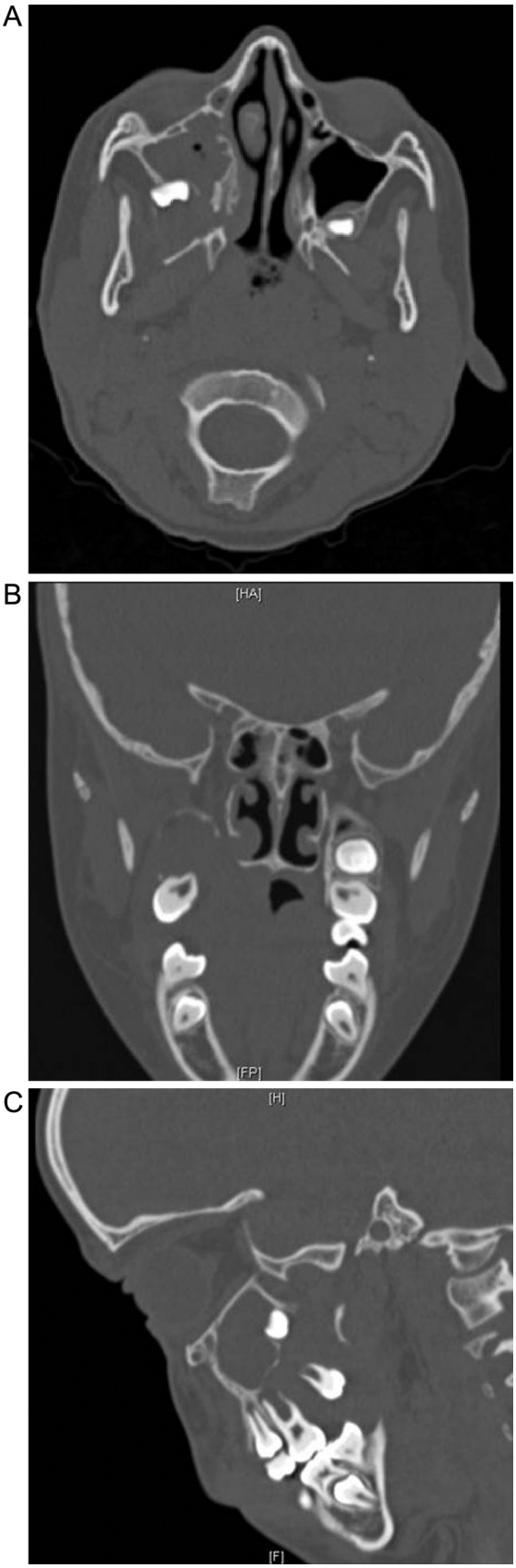

Fig. 3. A: CT scan axial cut showing obliteration of the right maxillary sinus with displaced upper right wisdom tooth. B: Coronal cut showing the apical border of the lesion. C: Sagittal cut showing the displaced second and third molars and the obliterated maxillary sinus.

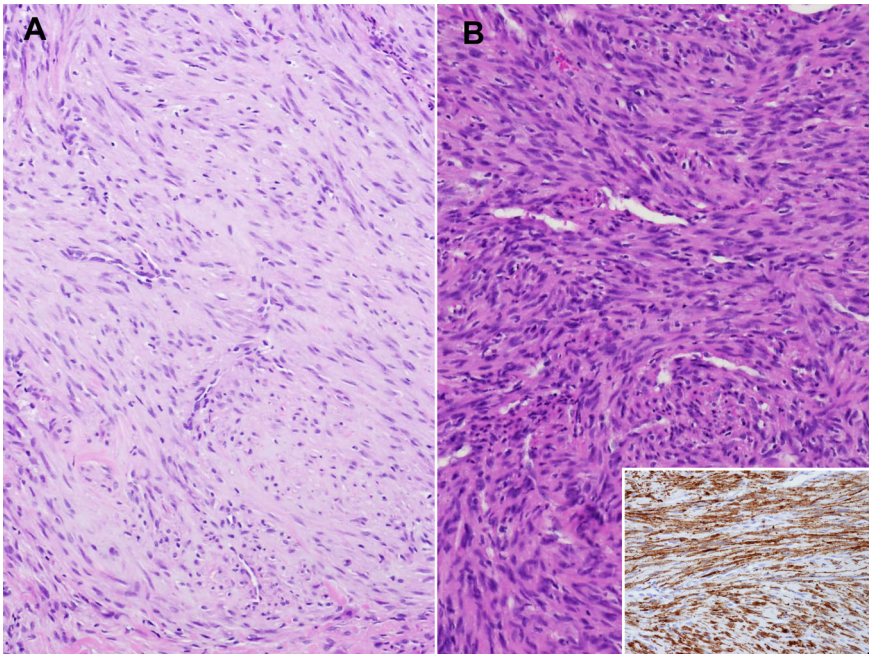

Fig. 4. A: A light-stained area consisting of bundles of spindle cells with abundant extracellular collageneous/chondroid-like matrix (hematoxylin and eosin, original magnification × 100). B: A dark-stained area comprising of groups of densely packed cells adjacent to hemangiopericytoma-like blood vessels (hematoxylin and eosin, original magnification × 100). (Inset) Bundles of tumor cells, intensely positive for alpha smooth muscle actin (original magnification × 200).

addition, areas of less well differentiated rounded and polygonal cells with slightly hyperchromatic nuclei were scattered throughout the lesion (dark-stained areas, Fig. 4B). Both these cell types were arranged around thin-walled, irregularly branching, hemangioperycytoma-like blood vessels. No nuclear atypia or pleomorphism was observed. The mitotic activity was low (2MF/50 HPF), and there was no evidence of necrosis, calcifications or stromal hyalinization. The tumor cells stained for alpha smooth muscle-actin (Fig. 4, inset), and did not stain for desmin, h-caldesmon, CD34, CD99 and Factor XIIIa. These finding were compatible with central myofibroma (Fig. 4).

3. Discussion

Based on the clinical and radiological findings, differential diagnosis (DD) should include: ameloblastic fibroma, central giant cell granuloma, keratocystic odontogenic tumor (KOT), unicystic ameloblastoma, juvenile fibromatosis and myofibroma. Moreover, multilocular appearance characterizes less common DDs including central vascular lesion and aneurismal bone cyst. Ill-defined borders of the more aggressive lesions can imply a malignancy bone tumor and therefore should be taken in consideration.

Several aspects were addressed for further supporting the selected most probable differential diagnoses. With regard to the nature of the lesion, a benign rather than a malignant lesion was favored, as no symptoms were reported and the child was in general good health. A malignant lesion in a child would be expected to be associated with deterioration in the patient health status over time. Regarding the origin of the lesion, peripheral or central (jawbones), we assumed that the expansion of the cortical bone, the extension of the lesion toward the maxillary sinus and the fact that there was no soft tissue hypertrophy indicates a central origin.

We rejected all the lesions which are rarely seen in the patient's group of age such as the odontogenic tumors: ameloblastic fibroma, KOT and unicystic ameloblastoma. Moreover aneurysmal bone cyst is considered favored in the second decade and therefore was also rejected.

The whereabouts of this lesion, situated on the maxilla, decreases the probability that this lesion is: ameloblastic fibroma, KOT or unicystic ameloblastoma, since they are all commonly found in the mandibular molar and ramus area. The diagnosis of Juvenile Fibromatosis was rejected due to the lack of skin bumps that frequently appear on the hands, neck, scalp, ears, and nose. Although the gingiva was hypertrophic on the involved region, the lack of joints deformities and physical limitation made the latter differential diagnosis unlikely to be positive.

The lesion is also unsuited with myofibroma prognosis since it rarely originates from the maxillary bone, supporting this is the high incidents reported of myofibroma appearance in the mandible.

The radiographic appearance of a multilocular lesion with displaced teeth is considered to be a non-specific finding and therefore could match any of the lesions listed on the DD list.

In summary, the final entities that were considered part of our differential diagnosis were: ameloblastic fibroma, central giant cell granuloma and central vascular lesion.

3.1. Imaging

Using a plane x-ray imaging, previous publications described central myofibroma as a lesion with unilocular and radiolucent appearance; cases of multilocular myofibroma were described as well [9]. Since most cases of central myofibroma were reported in the mandible, the x-ray of choice was panorex [1,6,7,9–11]. It was also mentioned that the borders of the central myofibroma lesions are either well or ill defined and perforation and\or expansion of the cortical bone can be witnessed as well [9].

Another appropriate evaluation tool for such lesions is the CT scan, demonstrating myofibroma as a low density lesion [7,10,11]. The panoramic X-ray imaging technique was less useful compared to previous mandibular reported cases, and since this lesion occupied the maxilla alone, it leaves us both plane films and CT imaging as our radiologic evaluation tools.

In the last decades, MRI was used for diagnosing myofibroma. Such work of utilizing MRI in evaluation of myofibroma within the

mandible was presented in Shibuya et al.'s article. The lesion typically presents with low and high signal intensity on T1 and T2 weighted spin echo machine, respectively [10]. Shibuya et al. also used a whole body scan of fluorodeoxyglucose positron emission (FDG-PET) and noticed a spot with increased uptake, assumed to be the site of the lesion [10].

In general, the improvement in imaging modalities and the wide usage of CT scan made it easier to diagnose and limit the DDs for this group of lesions. Thus, it is not surprising to find numerous reports of this rare lesion since the year 2000.

In this case study, the usage of the CT scan demonstrates: displacement of the right maxillary teeth, "cloudiness" of the right maxillary sinus, superior border of the lesion and its unilocularity appearance (Fig. 3), thus, helping to determine the surgical borders. Moreover, involvement of the internal organs was ruled out after a total body CT was preformed and examined.

3.2. Histopathology

Histological differential diagnosis of this tumor includes a spectrum of non-odontogenic neoplasms of mesenchymal origin such as: leiomyoma, solitary fibrous tumor/soft tissue hemangioperycytoma, benign fibrous histiocytoma, inflammatory myofibroblastic tumor and low grade myofibrosarcoma.

Leiomyomas pose one of the most difficult histological differential diagnoses chiefly due to similarity of individual cell morphology (spindle cells with pale pink cytoplasm) and general tumor architecture (fascicular growth pattern) [11]. These cells lack biphasic pattern (more primitive rounded cells with hyperchromatic nuclei) and do not present a pericytic vascular pattern. Leiomyoma cells stained with desmin and h-caldesmon, whereas myofibroma cells showed negative staining [11].

Solitary fibrous tumor/soft tissue hemangioperycytoma figures prominently in DD due to its possible primitive cells component and the characteristic pericytic vascular pattern, both seen also in myofibroma. The immune profile of the former demonstrates reactivity for CD34 and CD99, as oppose to myofibroma which is negative for both.

Benign fibrous histiocytomas demonstrate fascicular growth pattern of spindle cells and occasionally hemangiopericytoma-like areas. The fibroblast-like spindle cells are only part of a heterogeneous cell population that includes also plump "histiocytic" cells, xanthoma cells and a chronic inflammatory infiltrate. Histiocytomas are immune-reactive to CD68 and Factor XIIIa, which were negative in our case.

Inflammatory myofibroblastic tumors are characterized by a prominent inflammatory infiltrate composed of evenly distributed lymphocytes, plasma cells and spindled or histiocytoid tumor cells, in a variably myxoid or collagenous stroma; however the tumor lack both primitive cells and pericytic vascular pattern seen in myofibroma tumors. In addition, the lesion described herein is devoid of any significant inflammatory component and do not present histiocytoid nuclear features.

Low grade myofibroblastic sarcomas were described in pediatric population [12] and may affect oral cavity [13]. They may resemble myofibroma due to their fascicular growth pattern and myofibroblastic immunophenotype. In contrast to myofibroma these are diffusely infiltrative destructive lesions that show mild or worse nuclear atypia.

3.3. Treatment

Conservative resection was determined to be the treatment of choice based on the previously described central cases and the fact that no recurrences have ever been demonstrated in past studies [1,6–10,14,15]. The patient was admitted for surgical intervention which included a partial maxillectomy under general anesthesia and peripheral osteoectomy in order to make sure that the margins were free of tumor remnants.

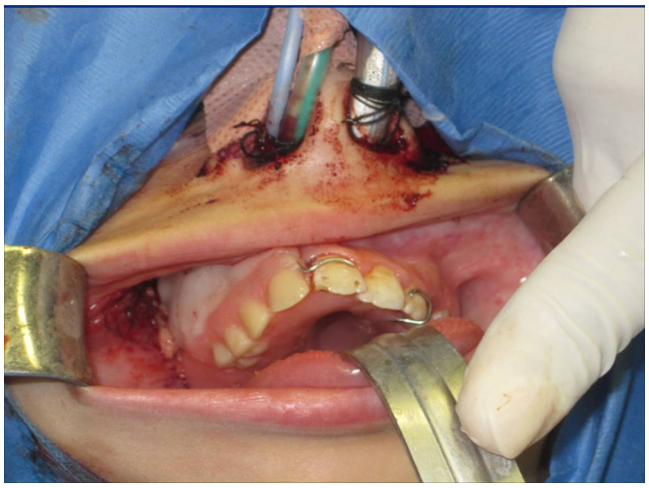

Fig. 5. Pre-fabricated obturator was adjusted to the maxillary defect using soft liner.

The maxillary defect was filled with iodoform gauze dressing and a prefabricated obturator (Fig. 5). The post-operative course was uneventful and 3 days later the patient was discharged and followed periodically. The iodoform gauze was pulled out a week later and the obturator had to be resealed and refitted using soft and hard lining materials several times based on the contraction of the surrounded tissues. In future, this obturator should be readjust to fit the growth of the maxilla which ends at the age of 15 [16].

Immediate reconstruction minimizes the deformity created by the tumor resection and prevents wound contraction and displacement of bony segments [17]. Nonetheless, this one stage approach has disadvantages including long operation time, blood loss and a high infection rate [18]. Moreover the ability to perform follow up and the capability to maintain a direct view of the involved area (to detect recurrences of the tumor) is limited using this modality. Since the staged protocol was previously reported and compared to the one stage protocol with successful results [18], the decision was made to treat this case with a staged protocol starting with a resection and using of an obturator as the first approach for treatment. The future stages will include reconstruction of the missing part with autogeneous bone graft, dental implants and lastly prosthodontics reconstruction.

Eight months post-operative until submission of this report, the clinical follow up demonstrated no recurrence signs of the tumor and the surgical site was totally healed (Fig. 6).

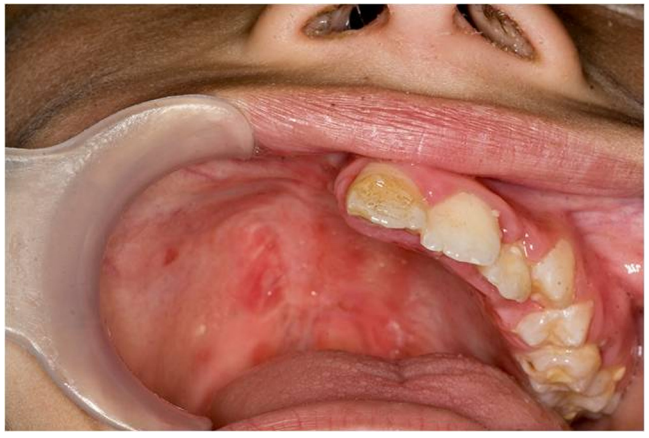

Fig. 6. Normal healing 8 month post surgery.

In conclusion, central myofibroma is a typical tumor of childhood and adolescents, previously described solely in the mandible. The innovation of this work derives from the fact it depicts a case in which myofibroma originates from the maxilla. The differential diagnosis consist of a wide list of benign and aggressive lesions, including jaw cysts which are seen in the dental office regularly by the general dentist; therefore, increased awareness of such an entity within the maxilla may lower the possibility of inaccurate diagnosis.

The treatment of choice is surgery with a preference for staged protocol in cases of segmental resections of either the mandible or the maxilla as presented in the current case.

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