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Peripheral fibromyxoma of the maxilla: report of a rare case

Włókniakośluzak obwodowy szczęki: opis rzadkiego przypadku

Garma Maroua[1], Hentati Hajer[1], Bellalah Ahlem[2], Selmi Jamil[1]

[1] **Oral Medicine and Oral Surgery Department, University Clinic of Dental Medicine, Faculty of Dental Medicine, Laboratory of Oral Health and Maxillofacial Rehabilitation, University of Monastir, Tunisia**
Head: *Selmi Jamil*, Clinical academic Professor

[2] **Anatomical Pathology and Cytology Department, Fattouma Bourguiba University Hospital, Faculty of Medicine, Monastir, Tunisia**
Head: *Zakhama Abdelfetteh*, Clinical academic Professor

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Summary

Odontogenic fibromyxoma is a benign odontogenic mesenchymal tumour of the jaws. It affects people in the second to the fourth decade of life with a female predominance. Jaws are rarely affected by this lesion with a mandible predilection. This tumour represents a subgroup of myxoma which are characterized by primitive mesenchymal cells proliferation producing an amorphous mucoid-rich intercellular matrix. The collagen fibre content is higher in the case of fibromyxoma, hence his name. This odontogenic lesion is subdivided into central fibromyxoma and peripheral one. The intraosseous type is more common and easier to diagnose, however, the extraosseous variety is extremely rare and presents always a diagnostic challenge. That was the motive behind this study which aimed to report a case of a peripheral fibromyxoma involving the right maxilla diagnosed in an adult male patient, so that the clinical and histological features of this tumour can be highlighted. The authors also propose the diagnostic approach, detail the treatment option and the follow up procedures.

Streszczenie

Włókniakośluzak zębopochodny jest łagodnym, zębopochodnym nowotworem mezenchymalnym szczęk. Dotyka osoby w drugiej–czwartej dekadzie życia, z przewagą kobiet. Zmiany te rzadko obejmują szczęki, ze skłonnością do występowania w żuchwie. Guz ten reprezentuje podgrupę śluzaka, która charakteryzuje się proliferacją prymitywnych komórek mezenchymalnych, tworząc amorficzną macierz międzykomórkową bogatą w śluz. Zawartość włókien kolagenowych jest większa w przypadku włókniaka, stąd jego nazwa. Ta zmiana zębopochodna dzieli się na włókniakośluzaka centralnego i zmiany obwodowe. Typ śródkostny jest bardziej powszechny i łatwiejszy do zdiagnozowania, jednakże odmiana zewnątrzkostna jest niezwykle rzadka i zawsze stanowi wyzwanie diagnostyczne. Stanowiło to motywację do przygotowania tego artykułu, którego celem było opisanie przypadku włókniakośluzaka obwodowego obejmującego prawą szczękę, zdiagnozowanego u dorosłego pacjenta płci męskiej, w celu uwypuklenia cech klinicznych i histologicznych tego nowotworu. Autorzy zaproponowali również podejście diagnostyczne, uszczegółowienia opcji leczenia i postępowanie pozabiegowe.

Introduction

Odontogenic fibromyxoma or myxofibroma is a benign odontogenic tumour of the jaws of mesenchymal origin which was first described by Virchow in 1863.[1-4] It represents a subgroup of myxoma, while the World Health Organization uses the term myxoma and fibromyxoma interchangeably.[1,4] In fact, both are characterized by the proliferation of primitive mesenchymal cells producing an amorphous mucoid-rich intercellular matrix,[4] but according to the literature fibromyxoma tends to contain more collagen fibers than myxoma.[5]

This neoplasm is relatively rare, it represents 1 to 17.7 % of odontogenic tumours.[6,7]

In 75 %, it occurs in the second to the fourth decade of life with an age range of 1 to 73 years.[3,4] It is very rare in children under 10 years and adults over 50 years.[2,8]

The mandibular localization is more frequent than the maxillary one with a predilection in the molar and premolar region.[9] This tumour represents a female predominance with a female to male ratio of 1.5/1.[8,10]

Fibromyxoma may present an extra- or intra-osseous localization in the jaws.[11] The central localization is widely reported in the literature while the peripheral one is very exceptional.[3,11] This fact prompted this study, which reports a very rare case of peripheral fibromyxoma of the maxilla, and thus highlights its clinical, radiological and histological aspects and details the therapeutic approach.

Case report

A 66-year-old male patient presented to the oral surgery department with the chief complaint of a growth that had been gradually enlarging for the past two years. The swelling was painless, which may explain the delay in consultation until the lesion increased in size causing a discomfort upon eating and swallowing as the patient was biting on the lesion during mastication.

His familial and past medical history was noncontributory.

The head and neck exam was negative for any lymphadenopathy.

The intra-oral examination showed an expansile growth measuring 2 cm in greater axis involving the right maxilla in the site of the upper right first molar which was clinically absent. This lesion was non-fluctuant, soft to firm in consistency without any tenderness on palpation. The mucosa over the swelling was normal. The growth consisted of two parts, vestibular and palatal, separated by a groove reflecting the imprint of the antagonistic arch during occlusion (Figure 1a, Figure 1b).

The cone beam computed tomography, which was recommended by the patient's dentist

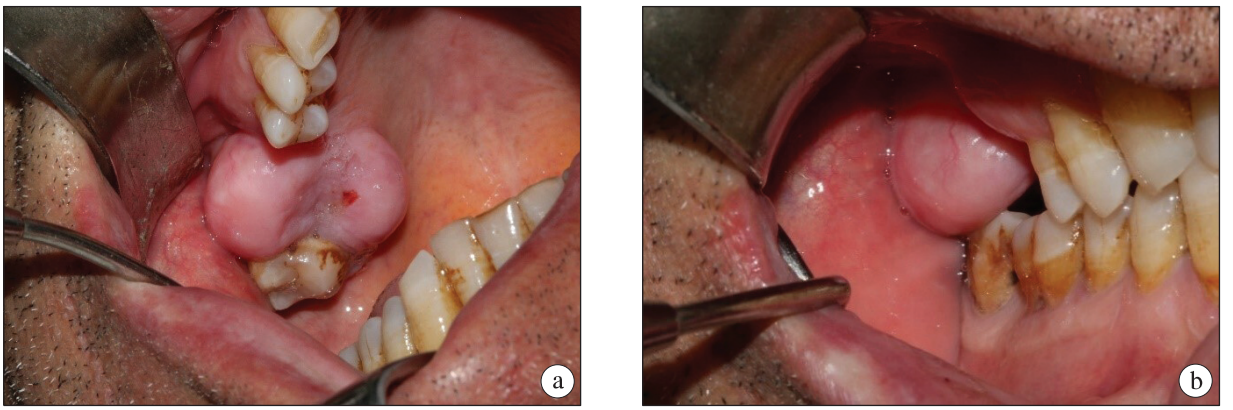

Fig. 1 a,b. Expansile nodule involving the right maxilla with a palatal expansion, covered by a normal mucosa.

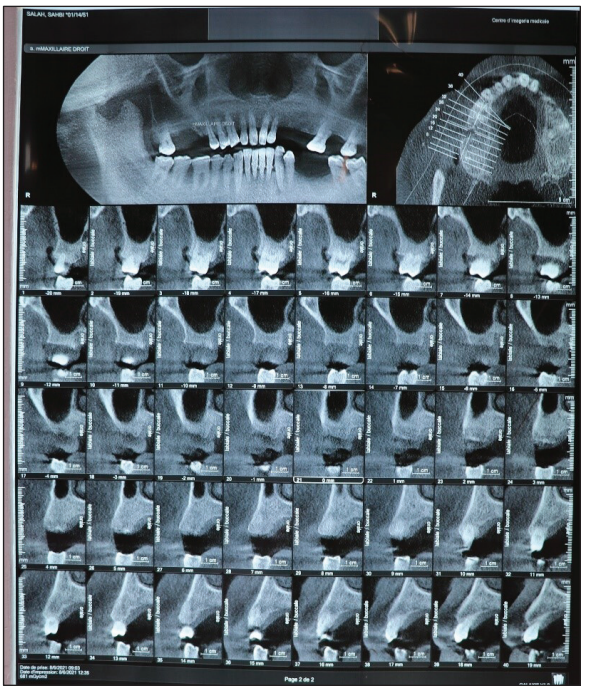

Fig. 2. Cone beam computed tomography: absence of the right maxillary first molar, no bone loss or anatomical structures displacement.

showed the absence of the tooth 16, no bone loss or anatomical structures displacement, which confirms the peripheral aspect of the lesion (Figure 2).

According to the patient the tooth 16 had been removed five years previously.

The biological examinations included complete blood count, a hemostasis check-up, parathormone, vit D, phosphorous and calcium serum levels; all the results were within the normal limits.

The patient underwent an incisional biopsy of the lesion under local anesthesia with in-depth curettage (Figure 3a, Figure 3b).

Histological examination of the specimen confirmed the diagnosis of peripheral fibromyxoma (Figure 4).

Immunochemically, cells were negative from CD34, PS 100, L'EMA and AML.

The patient continued regular follow-ups

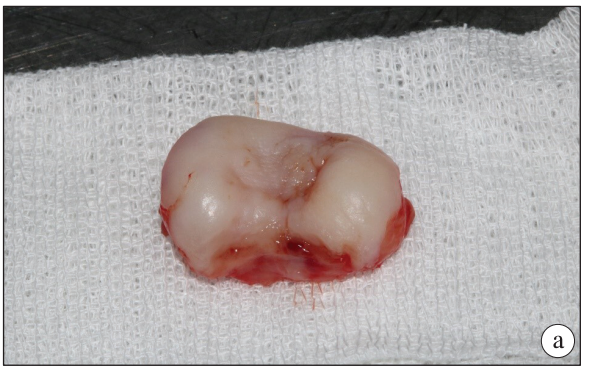

Fig. 3 a. Specimen.

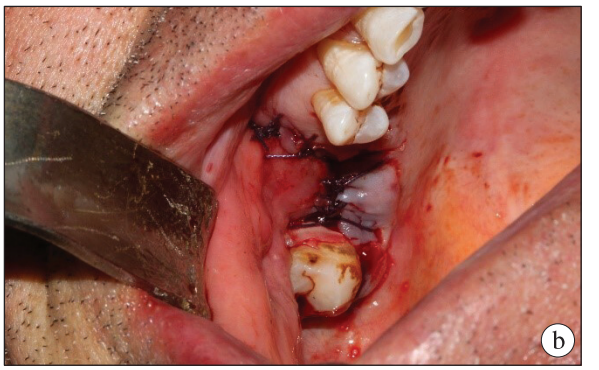

Fig. 3 b. Postoperative view: suturing the mucosa.

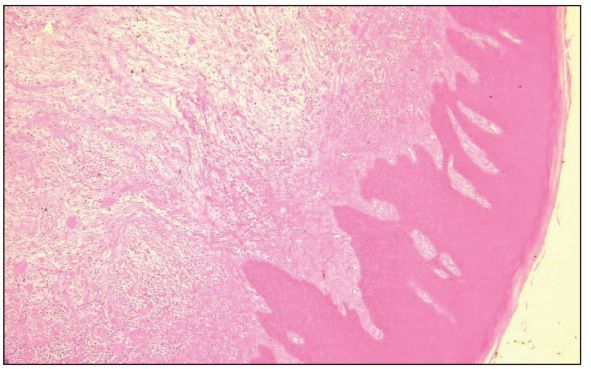

Fig. 4. The polyp is lined by an acanthotic squamous epithelium. The axis is formed by a proliferation of fibroblastic cells without atypia on a myxoid background (HE stains x 100).

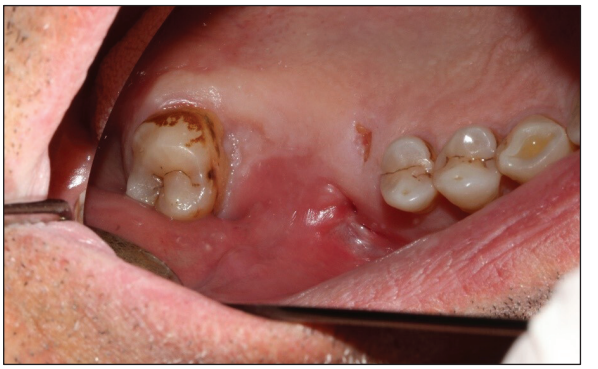

Fig. 5. Complete healing of the mucosa.

which were uneventful with no signs of recurrence (Figure 5).

Discussion

Fibromyxoma is a benign rare but not uncommon mesenchymal tumour affecting patients in the second to fourth decade of life with a female predilection.[3,11] It can occur in bony and soft tissues and rarely in the head and neck, where it presents a better prognosis than fibromyxoma involving the long bones.[3,10,12] In the jaws it occurs more frequently in the mandible: posterior mandible, anterior mandible than maxilla in order of frequency.[6,11] in fact, the maxilla area is the less frequent site but the more aggressive ones, hence it may involve the maxillary sinus, the zygoma or the orbits causing destruction of the maxilla and the anterior skull base.[3,13-17]

This lesion is classified among benign odontogenic tumours of ectomesenchymal origin with or without odontogenic epithelium.[7,16,18]

The odontogenic origin is supported by the fact that the greater portion of the fibromyxoma involving the jaws is derived from embryonic tissues such as dental follicle, dental papilla and the periodontal ligaments.[4,6,7,10] Also, it is backed by the evidence that fibromyxoma of the jaws occurs mostly in tooth-bearing areas as was the case of the patient reported here.[10,19]

Different terms are reported in the literature: myxoma, fibromyxoma, myxomatous tumour or myxofibroma. Actually, it is a category of myxoma, according to the World Health Organization,[1] classified as fibromyxoma depending on the amount of collagen fibers which must be considerable in the myxoid stroma in comparison with collagen fibers in the case of myxoma.[5,20-22]

For both lesions, the pathognomonic feature is the proliferation of mesenchymal cells producing an amorphous mucoid-rich intercellular matrix.[2,5,23]

This tumour, asymptomatic in character, grows slowly, hence the delayed diagnosis in the majority of reported cases. Nevertheless, this lesion is known as a locally invasive tumour that justifies the checking of molecular immunohistochemical markers during histological examination.[10]

Fibromyxoma is divided into central, or intraosseous lesions, and soft tissues or extraosseous lesions.[11,20] The central type is more frequently reported and easier to diagnose thanks to its intraosseous manifestations, which can be detected on X-ray examination. The peripheral type is extremely rare;[4,24] it appears as epulis-like lesion posing a diagnostic challenge,[20,25] and it is often misdiagnosed as epulis.

Clinically, the extraosseous type consists of painless soft tissue bulknodule growing peripherally next to the bony cortex.[11] This lesion represents a chief complaint when the swelling increases in size, thus interfering with mastication, occlusion, or aesthetics as was the case of the reported patient.

For this peripheral variant, no bone resorption is noted except a minimal peripheral resorption or local bone erosion in rare cases.[11]

Histologically, this extra-osseous lesion is characterized by the proliferation of spindle shape, stellate, round or polygonal hyalin cells in an amorphous mucoid matrix that contain a varied amount of collagen fibers.[10,11,20] Odontogenic fibromyxoma lesions are unencapsulated, without a clear boundary between pathologic and healthy tissue.[7,25]

All the type of cells present in this tumour are tested for some specific immunohistochemical markers.[11] Vimentin is typically expressed for all fibromyxoma indicating a mesenchymal origin.[4,11,20] This lesion is generally negative for S-100, CD34, SMA and CD86 as was the case of the patient reported here.[4,26]

It is crucial to determine the type of fibromyxoma, whether central or peripheral,

since it influences the decision regarding treatment.

The treatment of choice is surgical intervention ranging from local excision to en bloc resection.[3,11]

For the peripheral type, as reported here, the treatment consists in complete removal of the tumour including the involved tooth, adjacent soft tissue and the peripheral bone with thorough curettage of the adjacent bone. Conservative approach is widely discouraged due to the higher rate of recurrence for this lesion, which is between 25 to 43%.[3,7,10,11]

Recurrence is more common in young people, especially before puberty and before growth termination.[6]

This outcome is related to the type of treatment, the age and the fact that peripheral fibromyxoma presents ill-defined margin from the surrounding tissues and is non-encapsulated.[4,6]

Therefore, a long-term follow-up is required.[6,7,10,11]

Conclusion

Peripheral fibromyxoma of the maxilla is an extremely rare benign tumour. It represents a locally invasive lesion with a high level of recurrence that requires a careful diagnostic and therapeutic approach and a continuous clinical follow-up.

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Adres autorów: Oral Medicine and Oral Surgery Department, University Clinic of Dental Medicine, Faculty of Dental Medicine, Laboratory of Oral Health and Maxillofacial Rehabilitation, University of Monastir, Monastir, 5000, Tunisia.