Case Report

Odontogenic Fibromyxoma of the Maxilla: A Case Report and Review of the Literature

Eva-Maria Dietrich,1 Styliani Papaemmanouil,2 Giorgos Koloutsos,1 Hlias Antoniades,1 and Konstantinos Antoniades1

1 Oral and Maxillofacial Surgery Department, General Hospital “G. Papanikolaou”, Thessaloniki, 57010 Eksoci, Greece
2 Department of Pathology, General Hospital “G. Papanikolaou”, Thessaloniki, 57010 Eksoci, Greece

Correspondence should be addressed to Eva-Maria Dietrich, dietrich@auth.gr

Received 11 January 2011; Revised 21 February 2011; Accepted 28 February 2011

Copyright © 2011 Eva-Maria Dietrich et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Fibromyxoma represents a rare benign neoplasm that mostly affects the posterior region of the mandible. Here, we report the case of a 46-year-old male with a swelling of the right maxilla. After proper diagnosis, he was treated with enucleation and curettage of the tumor. The defect was filled with a pedicled buccal fat pad flap. The mesenchymal origin from the dental follicle of the fibromyxoma is the most plausible explanation. Radiological examination with MRI, CT, and conventional radiography contributes to the differential diagnosis from other benign tumors, such as the ameloblastoma. Its management is surgical and comprises enucleation and curettage or en bloc resection. Patients must be monitored for at least two years postoperatively in order to diagnose possible recurrence. According to the literature, the maxilla is a rare location of a fibromyxoma and, to our knowledge, our case is the 30th presented case of a fibromyxoma of the maxilla.

1. Introduction

Odontogenic fibromyxoma represents a rare slow-growing benign neoplasm, usually occurring in the 2nd and 3rd decades of life, rarely in children or adults over 50 years of age [1, 2]. It is described as a myxoma with abundant collagen fibres. Myxomas in general represent from 2.3% to 17.7% of all odontogenic tumors with fibromyxomas representing a small number of all myxomas [3]. Their size varies and in case of multilocular myxomas it may reach 4 cm [4]. They do not metastasize to the lymphatics [5]. Main sign is the swelling of the affected region and the displacement of dentition, with pain occurring less frequently mostly in cases of soft tissue myxomas [6]. Paresthesia, hypesthesia, anesthesia, or negative results of the vital tests during clinical examination are very rare [4, 7]. Although the origin of a myxoma is still obscure, an origination from the dental follicle seems to be the most reasonable explanation [1].

The aim of this case report and review of the literature is to present the rarity of a fibromyxoma of the maxilla, the contribution of the radiological examination to the differential diagnosis, and the importance of a meticulous enucleation in order to prevent recurrence.

2. Case Presentation

A 46-year-old male was referred to the outpatient department of the Oral and Maxillofacial Surgery Clinic of the General Hospital “G. Papanikolaou” of Thessaloniki, with a swelling of the right maxilla. The swelling occurred 8 months prior to the consultation. Facial and mucosal numbness, pain, or tooth mobility was absent.

The medical anamnesis of the patient did not reveal anything in relation to the pathological condition. Radiological investigation by means of a panoramic radiograph was not helpful in diagnosing the lesion. Waters’ view revealed complete obstruction of the right maxillary sinus. The swelling occurred 8 months prior to the consultation. Facial and mucosal numbness, pain, or tooth mobility was absent.

The medical anamnesis of the patient did not reveal anything in relation to the pathological condition. Radiological investigation by means of a panoramic radiograph was not helpful in diagnosing the lesion. Waters’ view revealed complete obstruction of the right maxillary sinus. The swelling occurred 8 months prior to the consultation. Facial and mucosal numbness, pain, or tooth mobility was absent.

The medical anamnesis of the patient did not reveal anything in relation to the pathological condition. Radiological investigation by means of a panoramic radiograph was not helpful in diagnosing the lesion. Waters’ view revealed complete obstruction of the right maxillary sinus. The swelling occurred 8 months prior to the consultation. Facial and mucosal numbness, pain, or tooth mobility was absent.
of the walls of the right maxillary sinus, obstruction with low density tissue of the whole cavity, and local erosion of the walls (Figure 1). The intravenous administration of contrast agent showed no enhancement of the lesion. Involvement of the floor of the left maxillary sinus, partial obstruction of the ethmoid sinus, and slight thickening of the mucosa of the left frontal sinus are indicative of a secondary sinusitis. The nasopharynx and lateral pharyngeal spaces were normal.

The lesion was approached by means of a lateral rhinotomy incision, with enucleation and curettage of the tumor. The lesion had a solid consistency and was totally resected (Figures 2 and 3). The defect was filled with a pedicled buccal fat pad flap.

The histopathological examination revealed randomly stellate, oval, and spindle-shaped cells in a myxoid stroma (Figure 4). Septa of residual lamellar bone and odontogenic myxoma were present into the marrow space in a pseudomalignant pattern (Figure 4). Immunohistochemical examination by means of Ki-67 labeling index revealed a low rate of cell mitosis.

Two years postoperatively, the patient shows no signs of recurrence. His rehabilitation period was uneventful and he gained complete function soon after surgery.

In order to prove evidence of the rarity of a fibromyxoma of the maxilla and the frequency of recurrence, a literature search was carried out using Pubmed. Search terms included ≪fibromyxoma≫ and ≪myxoma≫. Exclusion criteria were not relevant papers, interviews, books’ and conferences’ abstracts, comments, replies to author and to editor, and
unsupported opinion of an expert. 43 articles met our
criteria. In order to record only reports of fibromyxoma and
not myxoma, the articles were further sorted, in order to
include those reports of fibromyxomas that were mentioned
under the general term myxoma. Finally, 19 articles met all
criteria and were chosen for further evaluation (Table 1)
[8–26].

3. Discussion

Myxoma/fibromyxoma is a rare odontogenic neoplasm.
Fibromyxoma is classified as a specific type of myxoma
with a higher fibrous/myxoid tissue ratio than myxoma.
There is a discrepancy regarding the reports of fibromyxoma,
as many of them are classified under the general term
“myxoma”, making the review of the literature difficult.
According to Dutz and Stout, the term myxoma was first
used by Virchow in 1863, but the term fibromyxoma
was described by Marcove et al. in 1964 who reported
extragnathic locations of fibromyxoma [27, 28]. We use the
term myxoma/fibromyxoma as it is being used in many
histopathological books in order to describe myxomas of the
jaw bones. The review of the literature for previous reports
of fibromyxoma was based on case reports that clearly report
a “fibromyxoma”.

Myxomas/fibromyxomas are usually located intraorally
most often in the posterior regions of the mandible, its angle
and ramus and rarely extraorally [6, 29]. The maxilla and
anterior region of the mandible are rarely affected. The lesion
can be diffused or well defined, uni- or multilocular. It is
characterized by a mucous or gelatinous grayish-white tissue
that replaces the spongy bone and displaces the cortical plates
of the jaws [1]. Root displacement and resorption may be
present [1]. It may refer to hard and also to soft tissues.

Previous theories stress that the lesion derives from the
nerve sheath or is the result of degeneration of fibromas,
lipomas and so forth, due to the chronic irritation and the
degenerative processes following tissue anoxemia [26].
Recent studies advocate that myxomas/fibromyxomas arise
from the mesenchymatous tissue of the dental follicle, thus
being described as odontogenic with fibroblasts playing the
major role in cell dispersal [1]. This explanation fails to
describe soft tissue myxomas [7]. They probably arise from
supportive structures of the teeth like the gingiva and the
periodontal ligament [7].

Histopathological characteristics of the myxoma/fibro-
myxoma are the hypocellularity, the presence of stellate,
spindle-shaped cells into a loose myxoid extracellular matrix
with cells presenting with thin, long cytoplasmic prolifera-
tions that give to the tissue characteristics of immature
mesenchyme [30]. The fibromyxoid lesion may present loci
of calcification or ossification and a higher amount of
hyaluronic acid, collagen fibres and vessels than a typical myxoma [14].

Myxomas are diagnosed with radiological, histological,
and histochemical investigation. The radiological investiga-
tion reveals homogenous radiolucencies or sclerotic trabec-
ulations with different appearances, like “honeycomb”, “soap
bubble”, and “tennis racket” [31]. In our case, the lesion
appeared as a large radiolucent area with no trabeculations.

Radiological examination plays a crucial role for the
differential diagnosis of myxomas/fibromyxomas and also
between benign myxomas and malignant neoplasms with
myxomatous tissue. In Magnetic Resonance Imaging (MRI),
the lesion shows low-signal intensity in T1 and high-signal
intensity in T2 [5]. In contrast, Kawai et al. advocate that
the high-signal is shown in T1 and not in T2 [31]. These
discrepancies may be related to the ratio of fibrous/myxoid
tissue, the viscosity, the concentration of proteins, the
presence of haemorrhage and the hypocellularity [5, 31].
Immunohistochemical examination uses antibodies against
specific biological substances of neuronal, muscular, epithe-

dial, and mesenchymal tissues. The evaluation of the presence
of vimentin, an intermediate filament of the cytoskeleton
characterises mesenchymal tissues, thus also myxomas [1].
Fibromyxomas also contain a high amount of hyaluronic acid
[32].

During the process of differential diagnosis pathological
conditions that should be included are ameloblastoma,
central haemangioma, fibrous dysplasia, odontogenic cysts,
aneurysmal cysts, central gigantocytic granuloma, metastatic
neoplasms, well-differentiated liposarcoma, and other rare
entities like desmoplastic fibroma [5, 33].

The main pathological condition that may lead to
difficulties in diagnosis is the ameloblastoma, especially
when the bony septa are curviform [3]. An important
characteristic for differential diagnosis is the fact that when a
contrast agent (Gd-DTPA) is being administered, in case
of the ameloblastoma the MRI shows strong enhancement of
the solid portion of the tumor, in contrast to the myxoma
that shows homogenous high signal intensity [3]. It is also
important to mention that root displacement and resorption
is not unique in ameloblastoma.

The treatment of the fibromyxoma is surgical and
involves enucleation and curettage. The avoidance of recur-
rence is strongly related to the complete resection of the
lesion. The patient should be monitored for at least two
years after the surgical intervention due to the higher rate of
recurrence during this period [5].

Myxomas/fibromyxomas show a recurrence rate between
25% [2] and 43% [1]. This is strongly related to the nature of the
lesion, presenting without a sheath, thus making the
complete removal difficult. Other odontogenic tumors,
like the keratocyst or the ameloblastoma show a higher
recurrence rate of 30% [34]–58,3% [35] and 55%–90%,
respectively [35]. The frequency of recurrence of a fibromyx-
oma of the jaws is higher than that of any other bone thus
having a poorer prognosis [36].

It is stressed that complete resection and peripheral
osteotomy is the treatment of choice depending on the
size and behaviour of the tumor and results in a lower
rate of recurrence [6, 7, 33, 37]. Simon et al. suggest that
radical resection with a margin of 1,5–2 cm of healthy bone
is the treatment of choice [6]. Small bony defects of the maxilla, under 5 cm, can be reconstructed by means of a pedicled buccal fat pad flap (BFP) [38, 39]. Greater bony defects require the positioning of an obturator prior to the reconstruction with a graft.

In conclusion, the maxilla is a rare location of a fibromyxoma. The radiological examination by means of CT and MRI plays an important role in the diagnosis of a fibromyxoma and in the differential diagnosis from other pathological entities such as the ameloblastoma. Its management is surgical and ranges from enucleation and curettage to complete resection and peripheral osteotomy according to its size. Patients must be monitored for at least two years postoperatively in order to diagnose possible recurrence.

**Conflict of Interests**

We disclose any financial and personal relationships with other people or organisations that could inappropriately influence or bias our work. The authors did not have any writing assistance in this paper.

**References**

[1] L. Lo Muzio, P. Nocini, G. Favia, M. Procaccini, and M. D. Mignogna, "Odontogenic myxoma of the jaws: a clinical, radiologic, immunohistochemical, and ultrastructural study," *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontics*, vol. 82, no. 4, pp. 426–433, 1996.

[2] R. N. Aquilino, F. M. Tuji, N. L. M. Eid, O. F. Molina, H. Y. Joo, and F. H. Neto, "Odontogenic myxoma in the maxilla: a case report and characteristics on CT and MR," *Oral Oncology Extra*, vol. 42, no. 4, pp. 133–136, 2006.

[3] A. Mosqueda-Taylor, C. Ledesma-Montes, S. Caballero-Sandoval, J. Portilla-Robertson, L. M. R. G. Rivera, and A. Menezes-Garcia, "Odontogenic tumors in Mexico: a collaborative retrospective study of 349 cases," *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontics*, vol. 84, no. 6, pp. 672–675, 1997.
