Case Report

Myxofibroma of the maxilla, current concepts, and differential diagnosis

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Abstract

Odontogenic myxomas represent a small portion of all odontogenic tumors. A myxoma of the bone is a rare lesion that occurs almost exclusively in the jaws. An odontogenic myxoma has a variable clinical and radiological appearance, and it should be considered in the differential diagnosis of radiolucent and mixed radiolucent–radiopaque lesions of both jaws in all age groups. Myxomas consist of an accumulation of mucoid ground substance with little collagen, the amount of which determines whether it is called a myxofibroma. This paper presents the case of a 39-year-old male with a solid whitish red, nonulcerative, nontender expansion of both the buccal and palatal sides of the right upper alveolar bone. Results of a radiological examination revealed a unilocular radiolucency with cortical expansion and displacement of both the right upper second premolar and the first molar. The lesion was totally excised, and the histopathological examination showed a myxofibroma. Healing was uneventful, and there was no recurrence 12 months after surgical excision. Complete removal of the tumor, leaving no remnants attached to the soft tissue or bone, should be considered because of the well-known potential of myxofibromas to recur.

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Introduction

Myxomas are tumors usually seen in the left atrium of the heart, skin, subcutaneous tissues, and centrally in the bones.1 According to the histological classification of odontogenic tumors by Pindborg and Kramer, myxomas and myxofibromas are benign tumors that infiltrate and consist
wholly or partly of a myxoid stroma containing loosely arranged fusiform and stellate cells with more or less long anastomosing processes. The World Health Organization classifies these tumors as benign odontogenic neoplasms consisting of rounded and angular cells lying in an abundant myxoid stroma.

Myxomas represent 3–6% of all odontogenic tumors. These slow-growing tumors consist of an accumulation of mucoid ground substance with little collagen, the amount of which determines whether they are called myxofibromas. A myxofibroma of the bone is a rare lesion, which occurs almost exclusively in the jaws. In some cases, a myxoma is an aggressive tumor capable of extensive local infiltration and bone destruction, and it can spread into adjacent soft tissues. Although the mandible and maxilla are the two most common sites of head and neck myxomas, they have also been reported in the parotid glands, nasal cavity, paranasal sinuses, nasopharynx, and eyelids. The mandible is involved more often than the maxilla, and most reports show a slight predilection for females. The angle of the jaw, ramus, and adjacent molar region are most commonly affected. The anterior mandible is involved less frequently than the posterior mandible and ramus. Myxomas usually occur in the 2nd–4th decades of life, with a peak in the 3rd decade. Because they are benign tumors with a slowly progressive course, surgical options vary from conservative approaches, such as curettage or enucleation, to more aggressive lesions requiring a local anesthetic or en bloc resection.

This article presents a case of a myxofibroma, briefly reviews the pertinent literature, and suggests possible steps for the differential diagnosis.

Case presentation

A 39-year-old male was referred to the Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Istanbul University, Istanbul, Turkey, for evaluation of an expansion in the posterior right maxilla. The mass was slowly growing, and the patient was referred to our hospital by a private dentist.

The clinical examination revealed a solid, whitish red, nonulcerative, nontender fixed swelling that had a hard consistency and measured 2 cm × 2 cm. The swelling was located in the right posterior maxilla, and involved a part of the buccal mucosa that approached both the buccal and palatal sides of the alveolar bone. The texture and color of the overlying skin were normal. The patient could recall no pertinent traumatic or medical history.

A radiological examination showed a well-defined pear-shaped unilocular radiolucency with cortical expansion and displacement of both the premolar and the molar. The radiolucency had clearly defined borders. The superior surface was slightly scalloped and had displaced the floor of the sinus (Fig. 1). No destruction of the root of the premolar or the molar was seen. An axial computed tomographic examination showed both buccal and palatal expansion of the lesion extending from the first premolar to the second molar and the exact borders (Fig. 2). The regional lymph nodes were not palpable.

Discussion

When a myxofibroma is seen in the jaw, it is presumed to be associated with the dental anlage because of its close similarity to the mesenchymal portion of the tooth germ (i.e., the dental follicle, papillae, or periodontal ligament).
However, there are different theories regarding the role of the odontogenic epithelial component frequently found within myxomatous stroma on histological examinations. Many investigators discussed whether the origin of a myxofibroma of the jaws is osteogenic or odontogenic.⁶,⁷

Myxofibromas of the head and neck are rare. Myxofibromas can be divided into central and peripheral types,³ with the peripheral type being rarer. It is important to determine the type, because it is correlated with the treatment. Myxofibromas grow slowly without causing any symptoms. Because they enlarge painlessly, they can reach a considerable size prior to being noticed. Lesions can expand the bone, but they perforate the cortex only if they reach a great size. They are locally aggressive lesions, which tend to recur if they are treated too conservatively. Central types can proliferate in the jaw causing bulging of the bone cortex and ultimately breaking through into the surrounding soft tissue. These tumors grow asymptotically, and loosened teeth may draw attention to the lesions. Otherwise, they are usually diagnosed during routine dental examinations. Radiologically, the appearance of the lesions can range from a unilocal radiolucency to multilocal soap-bubble radiolucency, or they may have a honeycomb or tennis racquet appearance with a well-defined or diffuse margin. They may resemble a hemangioma, ameloblastoma, central giant cell tumor, or chondroma.⁸

According to some studies, root displacement rather than resorption is the rule for jaw myxofibromas. In our case, neither the premolar nor the molar was mobile. In some cases, resorption of tooth roots can be observed to varying extents,⁹,¹⁰ but no resorption was seen in our case. There was only a painless swelling for 5 months.

Radical surgery, excision, and enucleation followed by curettage of the surrounding bony tissue have all been recommended.¹⁰,¹¹ The recommended therapy is local conservative enucleation with adequate margins, the extent of which depends on the size and location of the tumor, because the tumor is not encapsulated and its myxomatous tissue infiltrates the surrounding bony tissue without causing its immediate destruction. Consequently, conservative treatment may result in recurrence. To avoid recurrence, after removing the tumor, we performed curettage, extracted the premolar and the molar, and smoothed off the surrounding bone, which had a burr. There has been no recurrence for approximately 12 months. A conservative approach may be used for smaller lesions to preserve function, reserving more radical surgery for recurrences and larger lesions.

It remains unclear whether the origin of the lesion is odontogenic. However, the presence of odontogenic epithelium suggests that this tumor has an odontogenic origin.⁹ We observed odontogenic epithelial islands scattered in the tumor, and they were thought to be truly odontogenic.

The differential diagnosis involves any lesion with myxoid change. Depending on the location, soft-tissue myxofibromas must be distinguished from nerve sheath tumors, oral focal mucinosis, and pleomorphic adenomas. Nerve sheath tumors are positive for S-100 protein, while myxomas are negative for this marker. Whether oral focal mucinosis and myxofibromas are separate and distinct lesions is debatable. Those that separate the two indicated that oral focal mucinosis, in contrast to myxofibromas, is usually approximately 1 cm in size, contains few if any reticulin fibers, and is a noninfiltrative, circumscribed
lesion. Pleomorphic adenomas may at times be myxoid. The presence of an epithelial ductal component and myxochondroid stroma separate them from true myxofibromas.\textsuperscript{12}

In conclusion, a myxofibroma is not an immediate threat to life, but it is infiltrative, locally invasive, and tends to recur if excision or curettage is incomplete. Complete removal of the tumor, leaving no remnants attached to the soft tissue or bone, should be considered. Whichever surgical approach is chosen, the patient should be observed over a long term, because of the well-known potential of myxofibromas to recur.

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