A 38-year-old male presented with a slowly progressive and painful swelling in the right cheek. On palpation, a firm, fixed and tender mass was detected over the right maxilla, with normal overlying skin. Endoscopic examination revealed a submucosal mass protruding into the right nasal cavity along the lateral wall, extending from the vestibule to the sphenoid sinus. Computed tomography showed a well-defined, 4x4 cm mass with cystic compartments, obliterating the right maxillary sinus, with destruction to the medial wall and extension into the nasal cavity. Magnetic resonance imaging showed similar findings of extension. A transnasal biopsy yielded a diagnosis of myxoma. En bloc resection of the mass was performed through a medial maxillectomy. No clinical or radiological recurrences were noted during a follow-up period of 24 months.

Key Words: Maxillary neoplasms/pathology/surgery; myxoma/surgery; odontogenic tumors/pathology/surgery.

Myxomas are relatively rare tumors of mesenchymal origin that can be found in numerous sites throughout the body including the heart, skin, subcutaneous tissue, and bones. In the head and neck region, two forms can be identified: “bone” type derived from facial skeleton and “soft tissue” type derived from perioral soft tissue, parotid gland, ear, and larynx. The maxilla and mandible are equally involved in most series even though some authors found a higher incidence in the mandible. It is most often located centrally in the maxilla and mandible and represents 3% to 6% of all odontogenic tumors. Mostly it is diagnosed between the ages of 10 and 40 years, and is slightly more common in females.

Otuz sekiz yaşında erkek hasta, sağ çenesinde yavaş büyüyen, ağrılı şişlik nedeniyle başvurdu. Palpasyonda, sağ maksillada, üzerinde normal deriyle örtülü, sert, fıkse ve ağrılı kitle vardı. Endoskopik incelemede, lateral duvardan sağ nazal kaviteye taşma gösteren, vestibülden sfenoid sinüse uzanan submukozal kitle saptandı. Bilgisayarlı tomografide, kitlenin 4x4 cm boyutlarında ve iyi sınırlı olduğu, kistik komponentler içerdiği, sağ maksiller sinüsü oblitere ettiği, medial duvarı aşındırıldığı ve nazal kaviteye yayılım gösterdiği görüldü. Üyelleşen transnasal biyopsis sonucu miksoma olarak bildirildi. Kitleye medial maksillektomi ile en bloc rezeksiyon uygulandı. Hastanın 24 aylık klinik ve radyolojik takibinde nüks bulgusuna rastlanmadı.

Anahtar Sözcükler: Maksilla neoplazileri/patoloji/cerrahi; miksoma/cerrahi; odontojenik tümör/patoloji/cerrahi.
In this article, we present a male patient with maxillary odontogenic myxoma.

**CASE REPORT**

A 38-year-old male presented with a slowly progressive and painful swelling of five-month duration in the right cheek. There was no history of tooth extraction or trauma. On palpation, a firm, fixed and tender mass was detected over the right maxilla, with normal overlying skin. Endoscopic examination revealed a submucosal mass protruding into the right nasal cavity along the lateral wall, extending from the vestibule to the sphenoid sinus. Other findings of head and neck examination were unremarkable.

Contrast computed tomographic (CT) scans of the paranasal sinuses showed a well-defined, 4x4 cm mass with cystic compartments, obliterating the right maxillary sinus, with destruction to the medial wall and extension into the nasal cavity (Fig. 1). To differentiate the cystic and solid compartments of the mass, magnetic resonance imaging (MRI) was performed, which showed similar findings of extension (Fig. 2).

The result of a transnasal biopsy was reported as myxoma. A medial maxillectomy with en bloc resection of the mass through a lateral rhinotomy incision was performed. The mass was extending to the sphenoid region without invasion. Laterally, on the floor of the maxillary sinus, extension to tooth roots was observed.

On histopathologic examination, the tumor had a scant, loosely cellular proliferation consisting of spindle-shaped or stellate cells, embedded in an abundant mucinous stroma. There were small and hyperchromatic nuclei. Mitotic figures and necrosis were absent (Fig. 3a-c). Immunohistochemical staining showed positivity for vimentin.

No clinical or radiological recurrences were noted during a follow-up period of 24 months.

**DISCUSSION**

Myxoma of the jaws has been classified as a benign odontogenic tumor composed of odontogenic ectomesenchyme with or without odontogenic epithelium. The evidence for its odontogenic origin stems from its almost exclusive location in the tooth-bearing areas of the jaws, its occasional association with missing or unerupted teeth, and the presence of odontogenic epithelium in a minority of cases.\[5,6\] Although myxomas of the jaws are benign, slow-growing, expansile tumors, they can be locally aggressive.\[2,4\] Maxillary myxomas spread rapidly through the cancellous bone and behave more aggressively than mandibular and oral soft tissue myxomas.\[3,1\]
Odontogenic myxoma of the maxilla: a case report

Myxomas have various non-specific clinical and radiological features and can resemble many other intraosseous jaw lesions.[7,8] The symptoms depend on the localization and extension,[1,2,3,4,5] and the average time between the first symptoms and treatment ranges from 1 to 5 years.[1] Radiographically, the lesions appear as multilocular radiolucencies, giving the bone a honeycomb or soap bubble appearance.[1-4,5,6,7] Sometimes they have a radiopaque appearance, particularly in the maxillary sinus.[8] In mandibular lesions, the cortex is usually sclerotic and intact. However, in the maxilla, antral involvement may occur as a soft tissue mass with occasional destruction to the antral walls.[1] The radiologic differential diagnosis includes cysts, fibroma, ameloblastoma, giant-cell reparative granuloma, fibrous dysplasia, hemangioma, and sarcoma.[1,2,3,4]

On gross examination, the tumor appears as a smooth, glistening, gelatinous, lobulated mass. Its consistency varies from soft to firm depending on the fibrous tissue content. Its color varies from gray-white or milk-white to yellow or amber. Most authors stated that the tumor was not encapsulated, but a few described a pseudocapsule.[1,2,3,4,5]

Histologic features of odontogenic myxoma are well-documented and resemble the primitive dental pulp, dental papilla, and tooth follicle.[2] These neoplasms exhibit a loose arrangement of mesenchymal stellate-shaped cells lying in a myxoid stroma.[1,2] Due to large amount of myxoid stroma, the tumor appears very hypocellular.[1] Enhanced mitotic activity and atypical mitoses indicate biologic aggressiveness and have been described only in reports of the rare variant of malignant odontogenic myxoma. Overproduction of mucoid ground substances are thought to be the cause of rapid growth as seen with malignant transformation.[9] Immunohistochemically vimentin and muscle actin may be positive as well as S100 in few cases.[1,5,6] Histopathological differential diagnosis includes hyperplastic myxoid dental follicle, benign or malignant nerve sheath tumors with myxoid degeneration, myxoid chondrosarcoma, and sometimes sinonasal hyperplastic inflammatory polyps.[6]

The treatment for odontogenic myxoma remains controversial. These tumors are benign and locally invasive, and have a propensity for recurrence if incompletely resected. The tumor is not encapsulated, and its apparent clinical and radiographic margins may not correspond with the true margins. Many authors favor simple enucleation with curettage, whereas others follow this with electrical or chemical cautery.[1,5,6] Recurrence rates have been reported as 25% to 43% after curettage and local surgical excision.[5,6] High recurrence rates and local aggressive behavior of myxomas have led some surgeons to perform disfiguring radical surgery at the initial stages of therapy.[12,4,6,11,12]

Although some authors advocate preoperative radiotherapy in order to achieve shrinkage of the
tumor, it should not be considered a standard therapy. These tumors are benign, occur in young patients, and are easily excised; therefore, the risk for radiation-induced tumors should be avoided.\textsuperscript{[5,6]} Chemotherapy is not recommended, as well.\textsuperscript{[12]} Recurrences most likely occur within two years; hence, close clinical and radiologic follow-up would be required. Early detection of recurrence would allow additional local excisions.\textsuperscript{[1-4,6,9,10]}

In conclusion, odontogenic myxoma, albeit less frequent among odontogenic tumors, should be considered in the differential diagnosis of patients presenting with a jaw mass.

\textbf{REFERENCES}

1. Chiodo AA, Strumas N, Gilbert RW, Birt BD. Management of odontogenic myxoma of the maxilla. Otolaryngol Head Neck Surg 1997;117:S73-6.
2. Kumar N, Jain S, Gupta S. Maxillary odontogenic myxoma: a diagnostic pitfall on aspiration cytology. Diagn Cytopathol 2002;27:111-4.
3. Landa LE, Hedrick MH, Nepomuceno-Perez MC, Sotereanos GC. Recurrent myxoma of the zygoma: a case report. J Oral Maxillofac Surg 2002;60:704-8.
4. Fenton S, Slootweg PJ, Dunnebier EA, Mourits MP. Odontogenic myxoma in a 17-month-old child: a case report. J Oral Maxillofac Surg 2003;61:734-6.
5. Lo Muzio L, Nocini P, Favia G, Procaccini M, Mignogna MD. Odontogenic myxoma of the jaws: a clinical, radiologic, immunohistochemical, and ultrastructural study. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1996;82:426-33.
6. Pahl S, Henn W, Binger T, Stein U, Remberger K. Malignant odontogenic myxoma of the maxilla: case with cytogenetic confirmation. J Laryngol Otol 2000;114:533-5.
7. Belet Ü, Sakan BB, Akan H, Yakubov K. Maksiller mik soma. Türk Tanısal ve Girişimci Radyoloji Dergisi 2003;9:54-6.
8. Kaffe I, Naor H, Buchner A. Clinical and radiological features of odontogenic myxoma of the jaws. Dentomaxillofac Radiol 1997;26:299-303.
9. Batti JS, Zahtz G, O’Reilly B. Quiz case 4. Myxoma of the maxillary sinus. Arch Otolaryngol Head Neck Surg 2000;126:679-83.
10. Allphin AL, Manigilia AJ, Gregor RT, Sawyer R. Myxomas of the mandible and maxilla. Ear Nose Throat J 1993;72:280-4.
11. Slootweg PJ, Wittkampf AR. Myxoma of the jaws. An analysis of 15 cases. J Maxillofac Surg 1986;14:46-52.
12. Chatterji P, Soni NK, Agarwal BK. Myxoma of the jaw. J Laryngol Otol 1985;99:391-6.