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CASE REPORT

Odontogenic myxoma of the maxilla: a case report

Maksillanın odontojenik miksoması: Olgu sunumu

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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.

Otuz sekiz yaşında erkek hasta, sağ çenesinde yavaş büyüyen, ağrılı şişlik nedeniyle başvurdu. Palpasyonda, sağ maksillada, üzeri normal deriyle örtülü, sert, fikse 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 aşındırdığı ve nazal kaviteye yayılım gösterdiği görüldü. Manyetik rezonans görüntülemede benzer yayılım bulguları gözlendi. Yapılan transnazal biyopsi 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.

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.[1-3] 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.[3] The maxilla and mandible are equally involved in most series even though some authors found a higher incidence in the mandible.[1,4] It is most often located centrally in the maxilla and mandible and represents 3% to 6% of all odontogenic tumors.[1,4] Mostly it is diagnosed between the ages of 10 and 40 years, and is slightly more common in females.[1,4,5]

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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.[2]

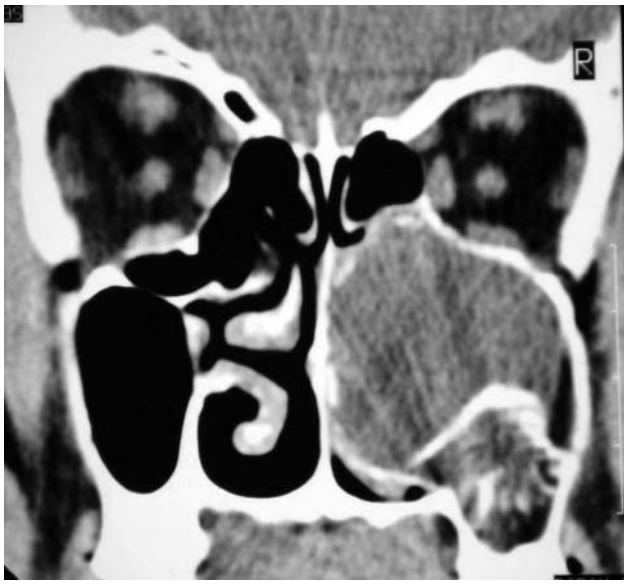

Fig. 1 - *Contrast computed tomographic coronal scan of the paranasal sinuses showed a well-defined, 4x4 cm mass with cystic compartments, obliterating the right maxillary sinus, with destruction to the medial wall of the sinus and extension into the right nasal cavity.*

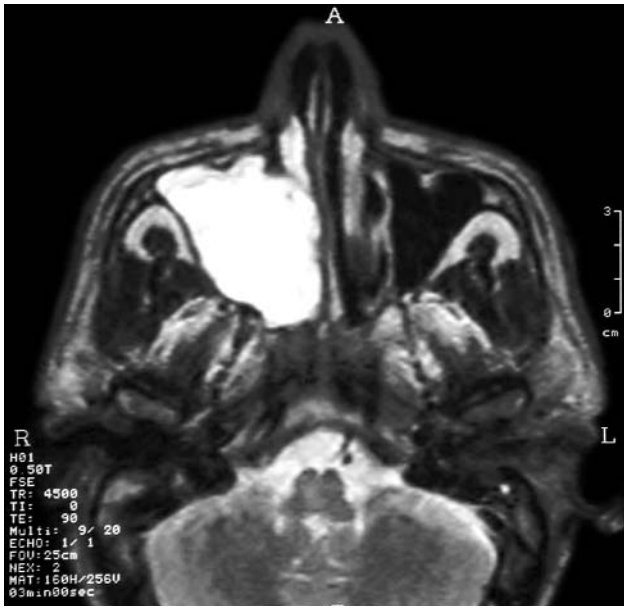

Fig. 2 - *Magnetic resonance imaging showed a polypoid mass obliterating the right maxillary and ethmoidal sinuses and infiltrating into the nasal cavity. The sphenoid sinus was filled by secretion.*

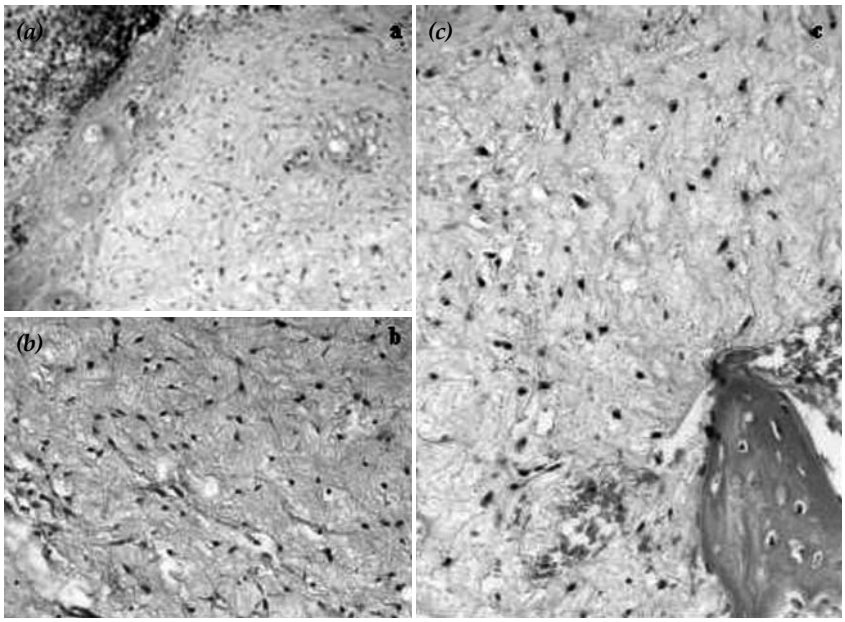

Fig. 3 - ***(a)*** *Uniform spindle cells lying in a myxoid background with a modest number of blood vessels (H-E x 40).* ***(b)*** *Tumor has a scant cellular proliferation of spindle-shaped cells embedded in a myxoid stroma (H-E x 100).* ***(c)*** *The periphery of the tumor presents local infiltration with replacement of bone (H-E x 100).*

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,[1,2,4,9] 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.[1-4,7,8,10] 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.[1,3,4,9]

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.[1,4,8]

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.[1,9] 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.[6] Immunohistochemically vimentin and muscle actin may be positive as well as S100 in few cases.[1,5,11] 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] Recurrence rates have been reported as 25% to 43% after curettage and local surgical excision.[5] High recurrence rates and local aggressive behavior of myxomas have led some surgeons to perform disfiguring radical surgery at the initial stages of therapy.[1,2,9,11,12]

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.[1,6] Chemotherapy is not recommended, as well.[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.[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.

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