

# Primary Intracranial Myxoma of the Lateral Skull Base: A Rare Entity in Clinical Practice

## Lateral Kafa Kaidesi Yerleşimli Primer Miksoma: Nadir Bir Olgu Sunumu

### ABSTRACT

Myxomas are rare benign tumors arising from mesenchymal tissues throughout the body. These tumors are usually seen in the atrium of the heart and the jawbone. Involvement of the skull base with intracranial extension is extremely rare, and only a few cases of primary intracranial myxomas have been described in the literature. A rare case of primary myxoma of the temporal bone is presented in this article. The patient underwent a skull base surgery with a pre-diagnosis of possible chondrosarcoma. The tumor pathology revealed a diagnosis of myxoma with bone and meningeal involvement. Despite the radical surgery, the tumor showed a local recurrence in three years. A second surgery with subtotal petrosectomy was required. In the article, the etiology, histological and radiological findings as well as treatment options of this rare entity were briefly discussed under the highlights of the relevant literature. Such a localization and intracranial extension of myxomas is extremely unusual in clinical practice; the diagnosis therefore requires a high degree of suspicion and detailed histopathological examination. The differential diagnosis frequently includes chondrosarcomas, chordoma, metastatic tumors of the skull, hemangiopericytoma, meningioma and other neoplasms of the dura and skull base in this location.

**KEYWORDS:** Myxoma, Skull base, Temporal bone, Neoplasm

### ÖZ

Miksomalarda nadir olmakla birlikte en sık kalbin atriyumunda ve çene kemiğinde yerleşen benign davranışlı mezenkimal doku tümörleridir. Kafa kaidesi tutulumu ve özellikle intrakranyal uzanım son derece nadir olup, literatürde bugüne kadar sadece birkaç vaka bildirilmiştir. Bu makalede, temporal kemikten köken alan ve intrakranyal uzanım gösteren bir primer miksoma vakası sunulmuştur. Hasta kondrosarkom ön tanısı ile opere edilmiş ancak patolojik inceleme bulguları kemik ve meningeal tutulum gösteren miksoma ile uyumlu bulunmuştur. Uygulanan radikal cerrahi girişime rağmen izleminde 3 yıl sonra lokal nüks gözlenmesi üzerine hastaya ikinci bir cerrahi girişim ve subtotal petrozektomi yapılması gerekmiştir. Bu makalede miksomaların etiyojisi, radyolojik ve histolojik özelliklerinin yanı sıra tedavi seçenekleri ilgili literatür eşliğinde kısaca tartışılmıştır. Miksomalarda bu tip yerleşim ve intrakranyal uzanım çok nadir olup, tanı için sıklıkla kişisel öngörü ve detaylı patolojik inceleme yapılması gereklidir. Kondrosarkomlar, kordoma, metastatik tümörler, hemanjiyoperisitoma ve meninjiomların yanısıra dura ve kafa kaidesinin diğer neoplazmları ayırıcı tanıda düşünülmesi gereken başlıca lezyonlardır.

**ANAHTAR SÖZCÜKLER:** Miksoma, Kafa kaidesi, Temporal kemik, Neoplazm

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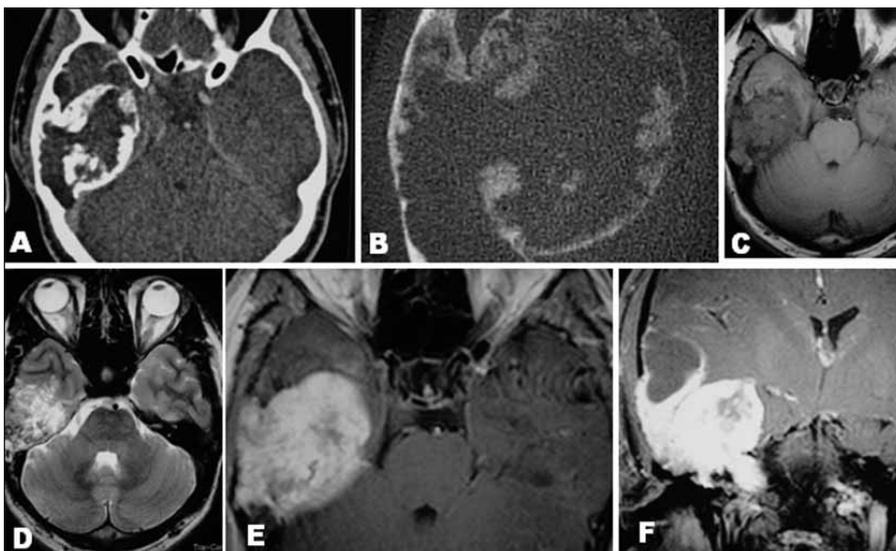
## INTRODUCTION

Myxomas are benign tumors of primitive mesenchymal tissue, and are usually found in the heart, skin, certain bones and genitalia (4). These tumors may be primary or embolic from an underlying cardiac myxoma. Primary myxomas of the head and neck are rare lesions in clinical practice, and usually involve the maxillofacial skeleton (1). There are only a few cases concerning primary intracranial myxomas that have been published in the literature (5,6). Intracerebral lesions that are metastatic or embolic from cardiac myxomas are more frequent but also uncommon (3).

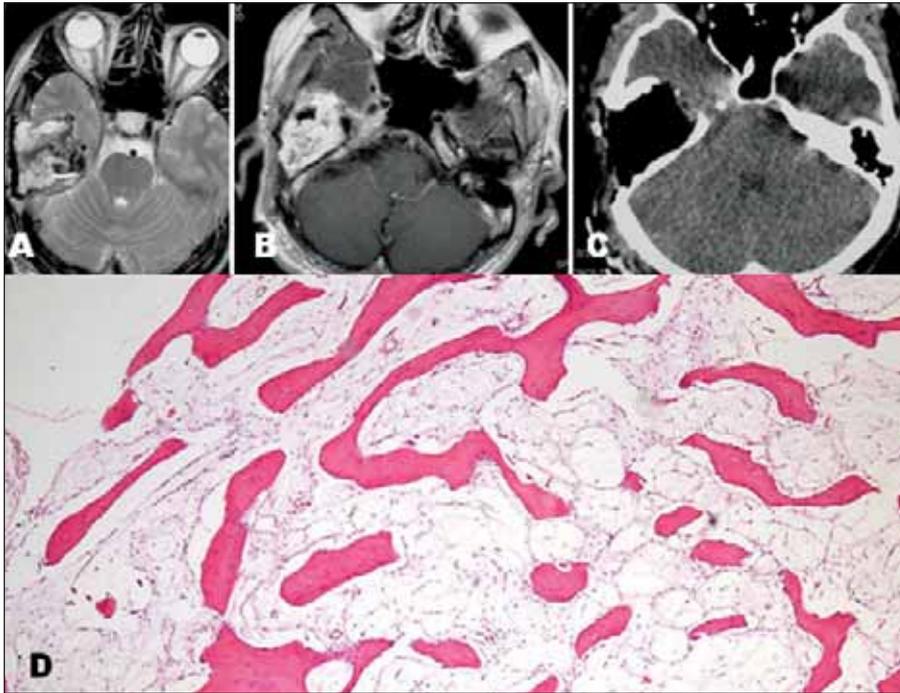
## CASE REPORT

A 34-year-old healthy male patient initially presented to an otolaryngologist with a complaint of right-sided facial weakness. His audiological and neurological examination revealed a conductive hearing loss and peripheral facial paresis on the right side. He referred to our hospital with a pre-diagnosis of intracranial neoplasm documented with a Magnetic Resonance Imaging (MRI) study. His MRI study revealed an extraaxial mass lesion of possible bony origin measuring 64x48x57 mm in diameter. The lesion compressing the temporal parenchyma with no associated edema was hypointense on T1-weighted (WI) and hyperintense on T2-WI images (Figure 1 C,D). Significant enhancement with a superiorly located cystic component was also remarkable (Figure 1 E,F). His Computed Tomography (CT) studies revealed an

enhancing calcified lesion invading the temporal bone and extending to the middle fossa (Figure 1 A,B). Its expanding nature in the petrous bone with decreased thickness of the inner and outer table was noteworthy. The tumor had poor vascularity on MRI angiography. He underwent combined surgery by neurosurgeons and otolaryngologists with a pre-diagnosis of chondrosarcoma. A skull base approach with right-sided temporal craniotomy and partial petrosectomy was used. The tumor was composed of solid, fibrous stroma and soft, mucoid islands scattered throughout the eroded bony cavities resembling a honeycomb pattern. It was firmly attached to the duramater and invaded it at the tentorial surface. The tumor was removed with adjacent invaded dura by a piecemeal fashion. Histopathological examination revealed a diagnosis of myxoma. He was advised to attend regular follow-up after the surgery. At the 3rd-year follow-up, his radiological screening showed a local recurrence in the lateral skull base (Figure 2 A,B). He underwent a second surgery through the previous route, and a radical resection with subtotal petrosectomy was done (Figure 2C). The findings of the histopathological examination were quite similar to those of the first one. The tumor was composed of satellite cells scattered within a mucoid stroma (Figure 2D). The absence of cellularity, cellular pleomorphism and mitosis was remarkable. Immunohistochemistry findings supported the diagnosis of myxoma (positive staining with vimentin and negative with pan-cytokeratin, keratin and S-100).



*Figure 1: Axial CT scans of the brain and temporal bone (A,B) show an enhancing mass lesion arising from the temporal bone. The tumor expands the petrous bone and extends to the middle fossa. MRI study shows an expansile bony lesion with hypointense in T1-WI (C) and hyperintense in T2-WI (D) images. Significant gadolinium enhancement and right-to-left brain shift due to mass effect are noteworthy in axial and coronal images (E and F).*



**Figure 2:** T2-WI and Gd-enhanced axial MR images (A and B) show local recurrence 3 years after the surgery. Early CT scan after the second surgery (C) shows the extent of resection.

Histopathological sections (D) demonstrate characteristic hypocellular areas of satellite and spindle cells scattered within a myxoid matrix and few bone spicules (x100). The absence of polymorphism and mitotic activity is typical.

## DISCUSSION

Myxomas are slow growing, benign neoplasms of mesenchymal origin that may arise in soft tissues and bone throughout the body (4). Histopathological features are usually benign; however they have infrequently been noted as local aggressive tumors in the literature (1). Craniofacial myxomas usually tend to arise at the maxilla and mandible, and manifest usually in the 2nd to 4th decades of life with no sexual predilection (6). Facial soft tissues, parotid gland, nasopharynx and the orbit are less frequent but possible cranial sites for these tumors (4). Myxomas typically manifest with painless, slowly growing masses in soft tissues or bones. However; they frequently cause the signs and symptoms of cranial nerve palsies at the time of diagnosis if they involve the skull base (8).

Radiological examination plays a key role in the diagnosis. Myxomas show an expansile growing pattern frequently limited in the inner and outer tables of the bone. They may be hypodense to isodense in CT scans, and show variable enhancement pattern (6). The bone window imaging better demonstrates the degree of bony erosion and expansile pattern of the mass as seen in fig.1B of the present case. The main determinant of MRI characteristics is the heterogeneous composition of the lesion. The MRI signal characteristics of

myxomas are low signal intensity in T1-WI, and hyperintensity in T2-WI images (1). Heterogeneous enhancement is a frequent finding (1,4). Clinical presentation and demographic data of myxomas may resemble many other neoplasms and vascular lesions at this site. Chondrosarcoma, chordoma, metastatic tumors of the skull, hemangiopericytoma, meningioma and other neoplasms of the dura and skull base are the most frequently encountered lesions in differential diagnosis (4,5,7). Chordomas originate from embryonic notochord remnants in the clivus, thereby showing a midline-growing pattern. By contrast, myxomas and chondrogenic tumors usually arise from cartilaginous synchondroses at the skull base, and tend to involve a more lateral location. Similar to myxomas, chondrosarcomas are expansile, lobulated, soft tissue masses with endosteal bone resorption. These lesions have high signal intensity on T2-WI images. Heterogeneous enhancement gives a typical appearance of a honeycomb pattern on MRI studies (2). Pure intra-osseous meningiomas and intradiploic epidermoid cysts are rare but possible lesions that should be kept in mind in differential diagnosis.

Histological examination of myxomas shows characteristic hypocellular areas of satellite and spindle cells suspended in a myxoid matrix (7,8). The absence of nuclear pleomorphism, hyperchromasia and mitotic activity is typical. Identification of the

immunohistochemical pattern is frequently necessary to distinguish these lesions from the other neoplasms with areas of myxomatous degeneration. Immunohistochemical staining characteristically shows positivity for vimentin and negativity for S-100 protein, neuron specific enolase, keratin, desmin and glial fibrillary acid protein (1,6,7). Myxomas are insensitive to radiotherapy, and the goal of treatment must be radical en bloc resection with safe margins (6,7). However; close proximity to the dura and venous sinuses at the tentorial area is the major concern that limits the extent of surgery at this location. Reoperation and close follow-up must be the treatment of choice in cases with recurrence. Radiotherapy should only be considered in inoperable cases due to its limited efficacy.

In conclusion, diagnosis of a skull base myxoma requires a high degree of suspicion because of its diverse radiological characteristics. The suggestive criteria of myxomas include higher signal intensity in T2-W images, heterogeneous signal pattern in all sequences and preservation of the pial barrier as well as the absence of edema and dural tail sign. Histological examination and immunohistochemical staining are mandatory to distinguish these rare lesions from other myxoid neoplasms of the skull base.

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