# Extracranial Meningioma in Maxillary Sinus: Case Report

## Maksiller Sinüste Ekstrakranial Meningiom: Olgu Sunumu

*** Sinan BAŞOĞLU, MD, * Hale ASLAN, MD, * Ali Ekber İLKNUR, MD, * Yılmaz ÖZKUL, MD, ** Mine TUNAKAN, MD, * Sedat ÖZTÜRKCAN, MD, * Hüseyin KATILMIŞ, MD**

* Atatürk Research and Training Hospital, Department of Otorhinolaryngology and Head and Neck Surgery,
** Atatürk Research and Training Hospital, Ministry of Health, Department of Pathology, İzmir

**ABSTRACT**

Paranasal sinus meningiomas are seen very rarely and called extracranial or ectopic meningiomas, although meningiomas are the second most common tumors of the central nervous system. Most of them are located in the head and neck, whereas paranasal meningiomas are very rare. Total surgical excision is the only treatment method. However, the location and spread of the mass may lead to surgical difficulties. This report presents a 24-year old woman admitted with the complaint of severe pain in the upper teeth and underwent a total excision after being diagnosed with a right maxillary meningioma. The clinical features, diagnosis, differential diagnosis, and therapy of paranasal sinus extracranial meningiomas were discussed with a literature review.

Keywords
*Meningioma; maxillary sinus*

**ÖZET**

Paranasal sinus meningiomaları etiyolojisi bilinmeyen ve çok nadir görülen tümörlerdir. Total cerrahi eksizyon, şu anda bilinen tek tedavi yöntemdir. Ancak kitlenin yerleşimi ve yayılımı cerrahi olarak zorluklara yol açabilir. Bu bildiride üst dişlerde şiddetli ağrı yakınması ile başvuran ve yapılan incelemeler sonucunda sağ maksiller ekstrakranial meningioma tanısı koyularak total eksizyon uygulanan 24 yaşında bayan olgu sunuldu. Bu olgu nedeniyle paranasal sinus ekstrakranial meningiomaları kliniği, tanısı, ayırıcı tanısı ve tedavisi literatür eşliğinde tartışıldı.

Anahtar S zc kler
*Menenjiyom; maksiller sinüs*

**Presented at the 7th Turkish Academy Congress, Antalya, Turkey, April 9-12, 2009.**

Çalışmanın Dergiye Ulaştığı Tarih: **11.05.2010** Çalışmanın Basıma Kabul Edildiği Tarih: **18.10.2010**

≈

Correspondence
Sinan BA O LU, MD
Atatürk Research and Training Hospital,
Department of Otorhinolaryngology and Head and Neck Surgery,
İzmir, Turkey
E-mail: basoglusinan@gmail.com

## INTRODUCTION

Meningiomas are second most common benign tumors of the central nervous system, mainly affecting the intracranial area.[1] The ones detected in the extracranial area are referred to as extracranial meningiomas, which occur very rarely. One to two percent of all meningiomas affect the extracranial area and they mostly reside in the head and neck. On the contrary, paranasal sinus meningiomas are extremely rare.[2,3] Despite certain theories, their etiology remains unknown. In the diagnosis, the histopathological examinations are necessary and the only accepted treatment method is the surgical removal of the mass.

## CASE REPORT

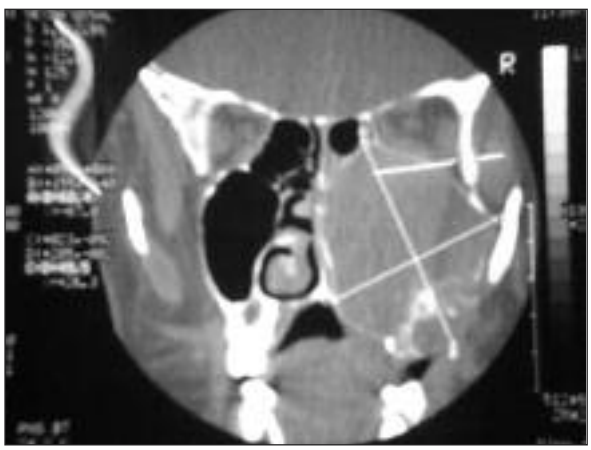
Our case is a 24-year-old woman admitted to our clinic with the complaints of swelling on the right side of her face, pain on her upper teeth and nasal blockage in the right nostril. Her signs and symptoms had started 10 months ago and gradually increased. It was found that there was a swelling in the right maxillary area, which led to obvious facial asymmetry as well as right exophthalmos. The examination revealed a significant thinning in the anterior wall and floor of the maxillary sinus. In the endoscopic nasal examination, it was observed that the anterior wall of the maxillary sinus was pushed medially, narrowing the nasal cavity in the absence of any vegetative mass. The computerized tomograpy (CT) scan of the paranasal sinuses revealed a 60 x 50 mm iso-hyperdense mass filling the right maxillary sinus and obliterating the right nasal cavity. The tumor contained calcifications in its internal structure, indicating contrast medium retention and destroyed bone anteriorly and inferiorly (Figure 1a, 1b). A transnasal endoscopic biopsy showed that the mass was a meningothelial meningioma indicating osseous metaplasia (Figures 2a, 2b, 3). In the operation performed through the canine fossa using the endoscopic instruments, the mass was freed from the anterior, posterior and lateral walls and excised totally with its capsule. The CT scan showed no recurrence on the post-operative 12$^{th}$ month follow-up (Figure 4).

## DISCUSSION

Meningiomas are the most frequent benign tumors detected in the central nervous system, accounting for 13-26% of primary intracranial tumors.[4] However, extracranial meningiomas are very rare and only 1-2% of all meningiomas are extracranial. They are clinically classified into two types as primary and secondary, based on the anatomical relation between the intracranial and extracranial areas.[5] Our case's tumor was considered as a primary meningioma.[6,7]

Four theories are proposed relatied to the origin of paranasal sinus meningiomas:

1) Arachnoid cell residues, getting out of the central nervous system during closure of neural tubes in the embryological period, 2) Ectopic arachnoid granulations and arachnoid cell clumps observed across the cranial nerves I, II, V, IX, and XI and at dural penetration points, 3) Metaplastic transformations of Schwann cells,



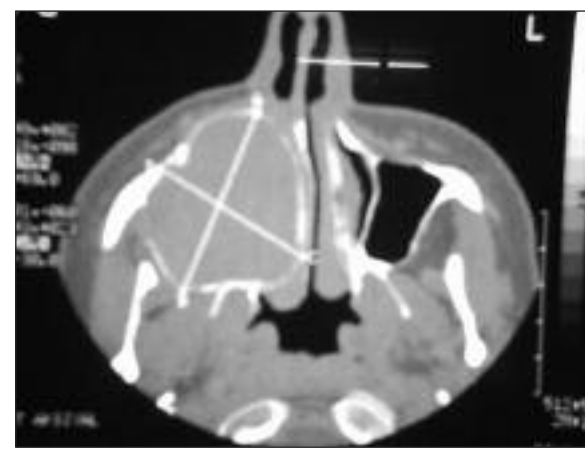




**Figure 1a, b.** The CT scan shows the iso-hyperdense mass, which occupies the right maxillary sinus and obliterates the right nasal cavity, containing calcifications in its internal structure, indicating contrast medium retention. The mass causes destruction on the anterior and inferior bony structures.

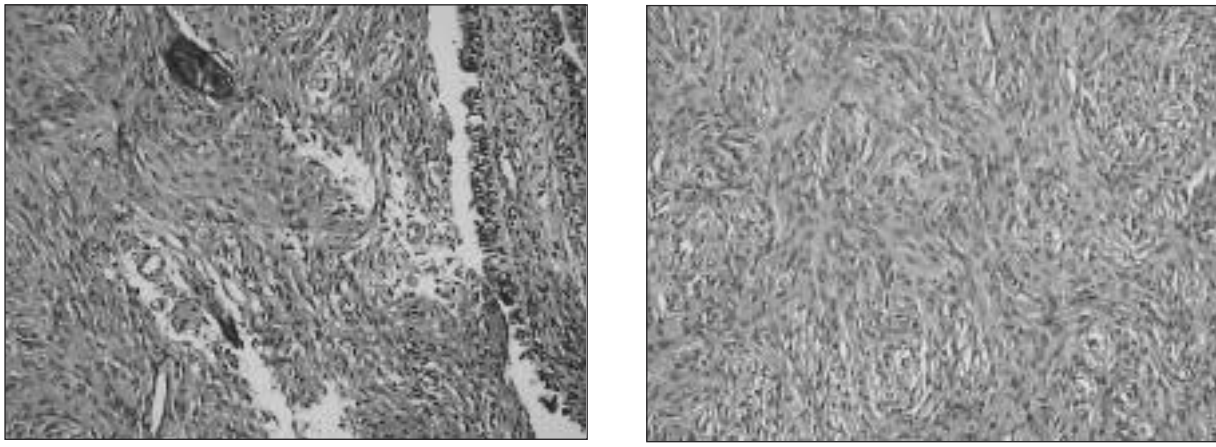


**Figure 2. a)** Meningothelial meningioma on the left and respiratory epithelium on the right side, **b)** Tumor cells are largely uniform with oval nuclei forming whorls (Hematoxylin and eosin, X200)

and 4) Differentiations of multi-potent mesenchymal cells.[2,3,6,7]

Extracranial meningiomas are diagnosed with histopathological examinations display four patterns as meningothelial, fibroblastic, transitional, and angioblastic.[8,9] Most paranasal sinus meningiomas are meningothelial.[10] The histopathological examination of our case also identified a meningothelial meningioma.

The meningiomas are usually solid, circular, and well-defined lobular tumors. The symptoms appear depending on its pressure on the retained sinus and anatomic formations around the sinus. These are slow-growing tumors and the time between detection of the first symptom and diagnosis is rather long and can extend to as long as 15 years.[6] In the present case, a solid, well-defined, lobular, and hemorrhagic tumor was detected. The patient was admitted with the swelling on the right side of her face, facial asymmetry and right exophthalmos. Besides, her signs and symptoms had started and gradually increased 10 months before the diagnosis.

The differential diagnosis of paranasal sinus meningiomas should include fibrous dysplasia, sinus mucoceles or mucopyoceles, and sinus carcinomas.[10] Furthermore, it is a well known fact that neurofibromatosis (NF), a genetic disease, occurs together with the meningiomas in some cases. Neurofibromatosis is classified into two subtypes and NF-2 typically occurs with extracranial meningiomas, acoustic neuroma, and other central nervous system tumors.[11] There were no signs and symptoms related to NF in our case.

The only treatment of the extracranial meningioma is total surgical excision of the tumor. The surgical approach must be determined taking into account the location of the tumor, the involved sinus, and the extent of the tumor. Extracranial meningiomas are reported to be radio-resistant; therefore, radiotherapy should be consi-

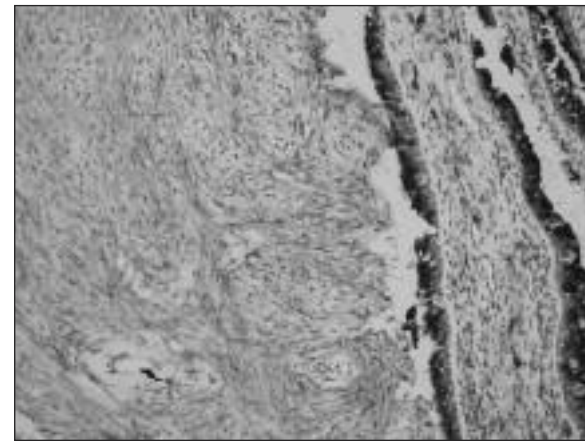


**Figure 3.** Immunoreactivity for epithelial membrane antigen (EMA) in tumor cells and respiratory epithelium (Immunohistochemical staining for EMA, X200).

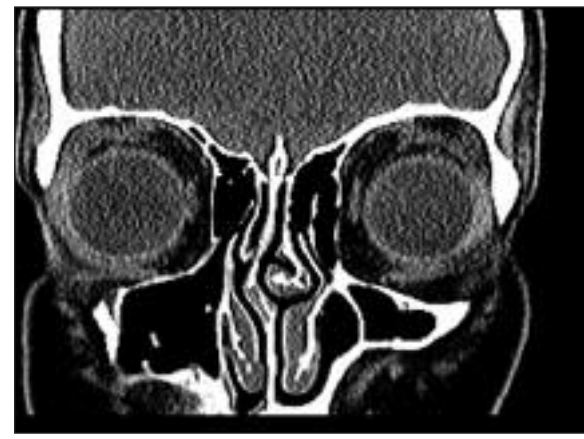


**Figure 4.** CT scan on post-operative 12th month shows no recurrence.

dered only for the palliative treatment in non-totally excised and recurrent tumors. It has been shown that the incidence of recurrence in meningiomas varies depending on the histopathological type of the tumor. In the angioblastic types, the recurrence rate is higher because of the difficulty of total resection of the tumor.[8] The prevention of the recurrence depends on the resection of the tumor totally. The prognosis of extracranial meningiomas is very good. Although recurrence is not reported after total surgical excision, the cases must be kept under close observation. In the present case, surgical excision could be performed totally and there was no recurrence at the 12th month follow-up examination.

## REFERENCES


1. Longstreth Jr WT, Dennis LK, McGuire VM, Drangsholt MT, Koepsell TD. Epidemiology of intracranial meningioma. Cancer 1993;72(3):639-48.
2. Kershisnik M, Callender DL, Batsakis JG. Extracranial, extraspinal meningiomas of the head and neck. Ann Otol Rhinol Laryngol 1993;102(12):967-70.
3. Shuangshoti S, Panyathania R. Ectopic meningiomas. Arc Otolaryngol 1973;98(2):102-5.
4. Kleihues P, Louis DN, Scheithauer BW, Rorke LB, Reifenberger G, Burger PC et al. The WHO classification of tumors of the nervous system. J Neuropathol Exp Neurol 2002:61(3): 215-25.
5. Lingen MW, Rao SM, Hutten MC, Pelzer HJ. Primary ectopic meningioma of the maxillary sinus: case report and review of the literature. Head Neck 1995;17(3):258-62.
6. New GB, Devine KD. Neurogenic tumors of the nose and throat. Arch Otolaryngol 1947;46(2):163-79.
7. Friedman CD, Costantino PD, Teitelbaum B, Berktold RE, Sisson Sr GA. Primary extracranial meningiomas of the head and neck. Laryngoscope 1990;100(1):41-8.
8. Swain RE Jr, Kingdom TT, DelGaudio JM, Muller S, Grist WJ. Meningiomas of the paranasal sinuses. Am J Rhinol 2001;15(1):27-30.
9. Ho KL. Primary meningioma of the nasal cavity and paranasal sinuses. Cancer 1980;46(6):1442–7.
10. Daneshi A, Asghari A, Bahramy E. Primary meningioma of the ethmoid sinus: A case report. ENT Journal 2003;82(4): 310-1.
11. Holland K, Kaye AH. Spinal tumors in neurofibromatosis-2: management considerations-a review. J Clin Neurosci 2009;16(2):169-77.