Cavernous Hemangioma of the Cavernous Sinus: A Case Report

Kavernöz Sinüs Kavernöz Hemangiomu: Olgu Sunumu

ABSTRACT
A case with giant cavernous hemangioma in the cavernous sinus is reported. The patient had a 5-month history of headache and visual symptoms. Computerized tomography (CT) and magnetic resonance imaging (MRI) studies demonstrated a lesion of the cavernous sinus in the left middle fossa. Only a biopsy of the cavernous malformation of cavernous sinus was performed during the first surgery because of uncontrollable haemorrhage during surgery. After the operation, radiation therapy was performed. Five years later, the lesion was totally removed because of its gradual enlargement. The second surgery was uncomplicated except for partial third nerve palsy. Ten years after the operation, the patient was in good health and there was no recurrence of the cavernous angioma.

KEY WORDS: Cavernous malformation, Cavernous sinus, Magnetic resonance imaging, Radiation therapy, Surgery

ÖZ
Kavernöz sinüsde dev kavernöz hemangiom olan bir olgu bildirilmektedir. Hasta beş aydır baş ağrısi ve görme şikayetlerine sahipti. Bilgisayarlı tomografi ve manyetik rezonans görüntüleme incelemeleri sol orta fossada kavernöz sinüsde yer alan bir lezyonu gösterdi. İlk cerrahi girişim esnasında, kavernöz malformasyonla uyumlu lezyondan kaynaklanan ve kontrol edilememeyen bir kanama olması nedeniyle sadece biyopsi alındı. İlk operasyondan sonra hastaya radyoterapi yapıldı. Fakat lezyonun tedrici olarak büyümesi nedeniyle, ilk operasyondan beş yıl sonra tekrar cerrahi girişim yapılarak total eksizyon yapıldı. Ameliyattan sonra sadece üçüncü sinir parezisi vardı. Olgunun on yıl sonraki kontrolünde klinik olarak ek problem yoktu ve control MRG'de lezyon saptanmadı.

ANAHTAR SÖZCÜKLER: Cerrahi tedavi, Kavernöz malformasyon, Kavernöz sinus, Manyetik rezonans görüntüleme, Radyoterapi

Correspondence address
Nurullah YÜCEER
Dokuz Eylül Üniversitesi, Tip Fakültesi, Nöroflirürji Bölümü,
Balcova, 35340 Izmir, TURKEY
Phone: +90 (312) 412 3306
Fax: +90 (312) 278 8802
E-mail: nurullah.yuceer@deu.edu.tr
INTRODUCTION

Cerebral vascular malformations are classified as arteriovenous malformation, venous angioma, cavernous malformation and capillary telengiectasia (22). Cavernous malformations make up of 5 to 10% of all central nervous system vascular malformations (22,25). Intracranial cavernous malformations may be intracerebral or extracerebral. Cerebral cavernous malformations frequently occur in the sylvian fissure and brain stem (11,22,25,30).

Intracranial extracerebral cavernous malformations (ECCMs) are rare vascular malformations. ECCMs make up approximately 1% of intracranial cavernous malformations (2,4,34). The most common localization for ECCMs is the middle cranial fossa (8,13,14,17,19,24,33). Treatment of middle cranial fossa cavernous malformations is difficult when compared with cerebral cavernous malformations and has high morbidity and mortality due to surgical complications.

In this study, we report a case with cavernous malformation in the cavernous sinus, mimicking a meningioma. The case is interesting because giant cavernous malformations of the cavernous sinus are rare. Cavernous malformations may grow up progressively in spite of radiation therapy. Treatment of growing cavernous sinus cavernous malformations is surgical removal.

CASE REPORT

First admission

A 33-year-old man was admitted with a 5-month history of headache and double vision. His neurological examination confirmed ptosis of the left eye and oculomotor paresis. The left pupil was dilated. A CT scan with contrast enhancement showed a hyperdense lesion located in the cavernous sinus (Figures 1A,1B). MR scans demonstrated a 2x2x2 cm homogeneously enhancing tumour which filled the left cavernous sinus, mimicking a meningioma or a cavernous angioma (Figures 1C,1D).

First operation

The preoperative diagnosis was a meningioma or a cavernous malformation of the cavernous sinus. The patient underwent a left frontotemporal craniotomy in the supine position with the head elevated. The left cavernous sinus was explored. The lesion was covered with dura. An incision of the lesion from the lateral wall of cavernous sinus was performed. Profuse bleeding from the vascular lesion did not allow us to perform subtotal or total removal. The pathology result was a cavernous hemangioma. The postoperative course was uneventful. One month after the operation, cranial radiation therapy was performed at a dose of 5600 cGy. The patient was followed up with MR scans.

Second admission

Five years later, he was admitted again with increased complaints of headache, nausea, vomiting and double vision. His neurological examination confirmed a left oculomotor nerve palsy. MRI scans with and without gadolinium enhancement demonstrated a hyperintense, well-demarcated, dumbbell-shaped lesion measuring 6x6x7 cm which compressed and displaced the left carotid artery (Figures 2A,2B,2C). A left common carotid artery angiogram did not show a vascular stain.

Second operation

After the balloon occlusion test, the patient underwent a left frontotemporal recraniotomy. The sylvian fissure was widely dissected. After exposure of the optic nerves and the optic chiasm, the left internal carotid artery was traced to its exit from the cavernous sinus. The lesion was covered with dura. Swollen dura over the cavernous sinus was cut. The mass was seen to extend outward from the lateral
wall of the cavernous sinus, stretching the oculomotor nerve upward and medially and the first and second divisions of the 5th cranial nerve laterally. The cavernous sinus was entered through the lateral approach. The lateral wall was opened. An encapsulated, dark red, pulsating soft mass was exposed. The mass was extremely vascular. The lesion was reduced in size with bipolar coagulation, and removed piecemeal with tumour forceps. Despite bleeding from the mass, a complete excision was performed with preservation of vascular structures. However, the left oculomotor nerve could not be preserved and an interfasicular anastomosis of the left oculomotor nerve using a sural nerve graft was performed. The cavernous sinus was then packed with Gelfoam and Surgicell.

Follow-up
The patient’s neurological examination revealed only a paresis of the third cranial nerve on the left side. The histopathological examination confirmed a cavernous hemangioma. Control MRI demonstrated total excision (Figures 3A,3B). Three years later, the patient’s ptosis on the left eye was unchanged. However, medial movement of the left eye was present. Nine years later, there was no recurrence of the cavernous hemangioma on MRI images (Figures 4A,4B). Ten years after the operation, the patient’s neurological examination showed that the ptosis had partially recovered.

DISCUSSION
Cavernous malformations of the cavernous sinus are rare vascular lesions. Cavernous hemangiomas of the cavernous sinus have been reported mostly as case reports in the literature (3,5,6,7,8,9,10,13,14,16,17,18,19,20,24,28,29,31,33,36). Cavernous malformations are seen equally among male and female patients. Male patients represent the majority of patients in the first 2 decades of life. Female patients seem to be more predisposed to haemorrhage, and typically haemorrhage in the middle decades of life (8,15,27,33).

Cavernous malformations of the cavernous sinus are usually associated with acute or subacute onset of visual symptoms such as diplopia, exophthalmos and decreased visual acuity (5,13,14,16,28,31,33,36). They may grow in size and also cause symptoms and signs related to compression of neural structures. Our case had both a subacute onset of visual symptoms and chronic compression of neural structures due to the mass of the cavernous malformation of the cavernous sinus.

The primary diagnostic tool for the cavernous malformations is MRI. MRI demonstrates a well-
demarcated, round or dumbbell-shaped, low to isointense mass lesion on T1-weighted images and an extreme high-intensity lesion on T2-weighted images (7,12,23,25,33,36). Plain skull radiographs and CT scans typically show bony erosion involving the dorsum sellae, posterior clinoid, orbital fissure, and floor of the middle fossa. Angiography shows no vascular stain or reveals early staining in the area of the cavernous sinus (24,33).

Some cavernous malformations may gradually enlarge, demonstrated by MRI follow-ups, as in our case. Some authors (11,30) have suggested that enlargement of the lesion may be due to reendothelialization within the haematoma (30) or to stimulation by the haematoma of connective tissue proliferation with neovascularization (11). Some studies have pointed out trends of aggressive clinical behaviour including pregnancy, familial or multiple forms of the disease, previous whole brain or stereotactic radiotherapy, incomplete lesion removal, and associated venous malformation (15,21,26,35). Some authors (1,26,27) have proposed that female hormonal factors are responsible for enlargement of cavernous angiomas. Behaviour of cavernous malformation after radiation therapy remains controversial. Maraire and Awad (15) reported that cavernous malformations did not become quiescent after radiation. Some authors have reported effectiveness of gamma knife radiosurgery for those who are not suitable for surgical excision or have residual tumour after surgery of ECCMs (6,9,18,20).

Our case was a male, and MR scans demonstrated cavernous malformation measuring 2x2x2 cm in diameter in the cavernous sinus. He underwent a biopsy of the lesion. After the first operation, radiation therapy was performed. Five years later, the lesion reached a size of 6x6x7 cm on MR scan follow-ups, in spite of radiation therapy.

The surgical treatment of cavernous malformations in the cavernous sinus remains a challenge for the neurosurgeon, as there is a high mortality and morbidity associated with uncontrollable and massive haemorrhage during surgery (8,17,19,24,33,36). Total surgical removal of cavernous hemangiomas in the cavernous sinus is difficult because of profuse bleeding from the vascular lesion as in our case. Some authors (14,16,23,32) have attempted combined treatment with surgery and radiation therapy, as radical surgical treatment alone still presents a high risk of mortality and morbidity. In the presented case, only a biopsy could be taken from the intracavernous lesion because of massive haemorrhage during the operation. On follow-ups by MR scans, the lesion gradually grew and the patient’s complaints increased and we decided to operate. Five years after the first operation, total removal of cavernous malformation in the cavernous sinus was successfully performed. Massive intraoperative bleeding from the cavernous angioma was controlled with bipolar coagulation and surgicell. Ten years after the operation, the patient was in good health and there was no recurrence of the cavernous hemangioma.

Cavernous malformations of the cavernous sinus should be carefully differentiated from meningiomas by the high signal that characterizes these lesions on T2-weighted MRI scans. This is unique for meningiomas, which appear more isodense than brain parenchyma on T2-weighted MRI images. This is an important imaging characteristic that should alert the surgeon to these notoriously vascular lesions that can bleed profusely at the time of surgical excision.

In conclusion, cavernous hemangiomas may progressively enlarge, as has been demonstrated by MRI scan follow-ups in our case. Symptomatic cavernous hemangiomas of the cavernous sinus should be removed because of growth of these cavernous angiomas.

REFERENCES