Intracranial Cavernomas: Indications for and Results of Surgery

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Abstract

Between April 1991 and April 1997, 46 patients were treated in our department presenting with intracranial cavernomas. Initial symptoms were focal seizures, bleeding episodes, and/or headaches. Mean age was 41 year (range 9 to 68 years). There were 24 female and 22 male patients. Computed tomography and magnetic resonance imaging were performed in order to establish the diagnosis, angiography was only indicated when the hemorrhaged area was so close to the subarachnoid space in the vicinity of the basal cisterns that an aneurysm had to be ruled out. Aggressive indication for surgery also in brainstem cavernomas was based on the natural history of the lesion, since the majority of patients presenting with intracranial bleeding had suffered several (up to six) episodes of previous hemorrhages. Patients' clinical status upon admission and accessibility of the cavernoma were taken into account for planning the operation. The operative planning and approach were greatly facilitated by using a neuronavigational device and intraoperative electrophysiological monitoring particularly in cavernomas located in the brainstem, thalamus, and medulla oblongata. Surgical removal of the lesions resulted in a new permanent neurological deficit only in two patients (4%). These data show that patients benefit from modern neurosurgical techniques in contrast to conservative approach in this disease of rather prolonged natural course.

Key words: cavernoma, hemorrhage, seizures, neuronavigation

Introduction

Voigt and Yasargil\(^{19}\) in 1976 presented the first series of 164 cases of cerebral cavernous hemangiomas. Most of these were only detected and diagnosed at autopsy, and are described as brownish, mulberry-like lesions. During the last 20 years due to improved neuroradiological diagnostic methods this cerebral vascular malformation has been diagnosed with increased frequency. Magnetic resonance (MR) imaging is today the method of choice to prove these cavernomas.\(^{14}\) Also the possibility to remove these lesions with acceptable morbidity improved especially with the help of microsurgery and electrophysiological monitoring, particularly during removal of thalamic\(^{10}\) and brainstem cavernomas.\(^{7,17,18,24}\)

Although complete removal of these benign lesions is usually possible and recurrences are rare, Pozzati et al.\(^{13}\) described more aggressive forms in 18 out of 145 patients with recurrent hemorrhages and de novo appearance in another location after successful first removal. They recommended MR imaging follow-up to diagnose new or recurrent cavernomas.

Materials and Methods

Retrospectively, we analyzed a series of 46 patients with an intracranial cavernoma who were treated in our department between April 1991 and April 1997.

Computed tomography and MR imaging were performed in order to establish the diagnosis. Cerebral angiography was only performed in 29 patients, when the hemorrhaged area was so close to the subarachnoid space in the vicinity of the basal cisterns that an aneurysm had to be ruled out. Aggressive indication for surgery also in brainstem cavernomas was based on the natural history of the lesion, since the majority of patients presenting with intracranial bleeding had suffered up to six episodes of previous hemorrhages.

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Results

Among the 46 patients with an intracranial cavernoma, four patients had multiple cavernomas. There were 24 female and 22 male patients (1.1:1). The youngest patient was 9 years old, the oldest 68. Mean age was 41 years. Most often a cavernoma became symptomatic at the age between 31 and 40 years (Fig. 1).

Thirty cavernomas were located supratentorially. Eight were frontal, 10 temporal, eight parietal, and one occipital. There were three thalamic cavernous malformations (Fig. 2). Infratentorially 16 cavernous hemangiomas were found. Eleven were located in the brainstem and five in the cerebellum. Four patients had multiple cavernomas. Another patient with recurrent symptoms had to be operated 7 months after the first operation because of an incomplete removal at the first operation.

Clinical presentation were bleeding in 21 patients (46%), seizures in 18 (39%), and headache in five (11%). Two asymptomatic cavernomas (4%) were detected on MR images performed for other reasons. The symptoms of the 11 patients with brainstem cavernomas were hemiparesis in seven, cranial nerve deficits in seven, headaches in six, gait ataxia in five, and dizziness in three.

The average time between the first symptoms and the operation was 8 months (1 day–48 months) for the patients who presented with bleeding. The latter time interval represented the group of patients who had a cavernoma in a deep-seated location, where the treatment options were discussed with the patients and their relatives. Some patients refused surgery and preferred to wait. The average length of history of patients who presented with seizures was 4 months (2 days–17 months). Cavernomas, which were responsible for headache, were removed within a mean of 2.5 months (1–6 months) after onset of symptoms.

Angiography was performed in 29 patients. Twenty-three were negative and only six were suspicious for a vascular malformation. In recent cases, MR angiography was thought to be sufficient to rule out an aneurysm or an arteriovenous malformation (Fig. 3).

Eighteen patients presented with seizures (39%).
Fig. 3 Cavernoma in the brainstem. left: Magnetic resonance (MR) image, right: MR angiogram.

Fig. 4 Postoperative results (Karnofsky performance status [KPS] 40–100) as compared to preoperative clinical status (Glasgow Coma Scale [GCS] 12–15). ○: GCS, ▽: postoperative KPS, ▼: KPS at follow-up.

Eleven had generalized fits, three had a partial focal (Jackson) seizure, and four had psychomotoric epilepsy. These patients had the best benefit from an operation. In the group of patients presenting with generalized seizures, eight had no seizures postoperatively and the anticonvulsant medication could be reduced and subsequently stopped postoperatively. The frequency was less in one patient, and two patients with a single generalized seizure preoperatively had another fit after the operation. In the group of psychomotoric epilepsy, three patients had less frequent fits postoperatively and so had one in the focal group.

There was no surgical mortality. Postoperatively good results were achieved in 35 patients (76%). Neurological deficits were unchanged in seven patients (15%) and worse in four patients (9%). Additionally, postoperative deficits improved within one week. After one week four patients with brainstem cavernomas improved further, whereas six patients remained unchanged in the first week, but four of them had improved neurological function on later follow-up examinations. During follow-up examination in April 1997, 41 patients were in an excellent or good condition (89%). The neurological condition was unchanged in four patients (9%) and worse in one patient (2%) (Fig. 4).

Discussion

Although cavernomas are vascular lesions with benign character, their natural history remains unpredictable.²,⁶,¹⁰,¹⁵ Their tendency of recurrent hemorrhages leads to stepwise impairment of the patients' neurological function, not as dramatically as with hemorrhage due to arteriovenous malformations but still often reducing quality of life and warrants surgical removal. Zabramski et al.²² demonstrated that there is also a familial form of cavernous malformations, which is a dynamic disease. They described an increasing incidence in the last years.

Nowadays the diagnostic method of choice is the MR imaging. Typically, cavernomas present on T₂-weighted MR images with a hyperintense (cavernous) center and a hypointense hemosiderin rim. Because the MR imaging appearance is so classical for a hemorrhaged lesion—the differential diagnosis being only tumor bleeding or very small arteriovenous malformation—angiography is mostly thought to be unnecessary. In our department, angiography was only performed in patients in whom the MR imaging shows the lesion so closely related to the subarachnoid space that an aneurysm has to be ruled out.

The indications for an operation, apart from seizures, were neurological deficits and recurrent hemorrhages. Incidental cavernomas should be operated dependent on their localization. In 12 out of 21 patients (57%) who presented with bleeding, recurrent hemorrhages had occurred, the maximum being six times. Previous hemorrhages and the risk of further bleeding were the most convincing arguments for an operative removal of the lesion when discussing treatment options with the patients and his relatives.

Controversy exists how to treat incidentally detected intracranial cavernomas. Some authors would remove superficially located lesions and observe the deep-seated ones.³,²¹ We favor aggressive surgical approach even in these areas, if accessible, to prevent the patient from the risk of further hemorrhages. Lombardi et al.¹⁰ recommended also the removal of cavernomas in the pineal region. Two of their cases were associated with a venous malformation. This coexistence of cavernomas with venous malformations is well known.¹,⁸,¹²,²⁴ One of the most important
points in operations of especially deep-seated cavernous hemangiomas is to leave a possible associated venous malformation intact to reduce the risk of a hemorrhagic infarction.

While Fahlbusch et al.\(^2\) in 1991 reported on 20 brainstem cavernomas and described surgical accessibility in only 10, the number of especially deep-seated lesions who are removed surgically has increased in the last years.\(^8\)

Intraoperative electrophysiological monitoring and neuronavigational devices such as the Viewing Wand (ISG, Toronto, Canada), which are routinely used at our institution, are quite helpful to achieve easy access via a sulcus with minimal or no brain damage, to the lesion to allow for a safe and complete excision of the lesions. The last 24 patients were operated with the help of this device so far. Stereotactically guided removal of cavernomas also showed promising results.\(^4\)

Besides the gold standard therapy of surgical removal, radiosurgical therapy has also been discussed. Personal experience with patients treated by proton beam therapy elsewhere suggests against radiosurgery, similarly to Steiner's opinion (personal communication in 1988). Kondziolka et al.\(^9\) reported good results after stereotactic radiosurgery for cavernous malformations. They reported a reduced annual hemorrhage risk beginning 2 years after radiosurgical treatment. Weil et al.\(^20\) compared the results after radiosurgery and microsurgery in the treatment of brainstem cavernomas and found good results in both groups. Forty to sixty percent of the cavernomas were found as the cause of seizures by Zevgardidis et al.\(^23\) They also presented gratifying results after removal of supratentorial cavernomas who are removed surgically has increased in the last years.\(^3\)

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