Original Article

Intracranial cavernomas: Analysis of 37 cases and literature review

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Aims: Thirty-seven patients with intracranial cavernomas managed in our department are retrospectively analyzed. **Materials and Methods:** The data of 37 patients with cavernoma who were admitted to our department between 1995 and 2003 were reviewed retrospectively. There were 30 male and 7 female patients with a median age of 26 years (range, 9-57 years). Four cases were treated surgically, 13 were treated by stereotactic radiosurgery (SRS) and the remainder were managed conservatively. **Results:** New hemorrhage or additional neurological deficits were not observed in the surgically treated cases, 12 patients who underwent SRS and the other patients who were followed up. One of the 13 patients treated by SRS, underwent microsurgery due to increased seizure frequency. One of the patients treated surgically died on the 11th postoperative day. **Conclusion:** Clinical observation should be the choice of management for patients without new or progressive neurological deficits, without two or more hemorrhages and in patients where the seizures are controlled with drugs. Surgery is the first choice for the cavernomas located in the non-eloquent locations. Radiosurgery may be an alternative for patients having deep-seated and eloquent area located cavernomas and for patients not willing or suitable for surgery.

Key Words: Cavernoma, cerebral, radiosurgery.

Introduction

Cavernoma is a benign vascular hamartoma. Approximately 50% of the cases are multiple.[1] They rarely occur in the spinal cord.[2,3] The onset of symptoms is usually in the third or fourth decade of life, although some cavernomas have presented in childhood.[4] Retrospective surgical series show good results after surgery, but the outcome in such patients if they had not undergone surgery is unknown.[5] Recently, radiosurgery has been proposed for the treatment of cavernomas located in an eloquent region of the brain.[6-8]

We review our experience with 37 cases of cavernomas.

Materials and Methods

The data of 37 cavernoma cases (30 males, 7 females) admitted to our department between 1995 and 2003 were reviewed retrospectively. The median age was 26 years (range, 9-57 years). Clinical features are summarized in Table1 and Table 2. In all the patients, computed tomography (CT) and magnetic resonance imaging (MRI) were performed. Locations of the lesions were 57% supratentorial. Electroencephalogram (EEG) was performed in 19 patients presenting with seizures. Except for two cases, the EEG was abnormal and concurrent with the site of the lesion. Digital substraction angiogra-

Table 1: Symptoms and signs of the cavernous angiomas

Symptoms and Signs	No. of patients	%
Headache	19	51
Epilepsy	19	51
Focal Neurological Deficits		
Imbalance	7	19
hemiparesis	5	13
speech disorder	4	11
dismetri	3	8
impairment of vision	2	5
hearing loss	2	5
tremor	2	5

Table 2: Localizations of the lesions and management modalities in cavernous angiomas

Supratentorial (57%)	21	Follow-up	SRS*	MS†
Parietal	3	1	2	-
Occipital	2	2	-	-
Temporal	5	2	1	2
Frontal	2	-	(2-1=1)	1
Thalamic	4	2	2	-
Basal ganglia	5	2	3	-
Infratentorial (43%)	16			
Cerebellar	10	8	2	-
Brainstem	5	3	1	1
Inferior bulbus +upper cervical	1	1	-	-
Total	37	21	12	4

*=Stereotactic Radiosurgery, †=Microsurgery

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phy (DSA) was performed in 5 patients who had hemorrhage. The angiogram did not reveal any vascular abnormality. Four cases were treated surgically (Figure 1), 12 cases were treated by SRS and the rest of the patients were managed conservatively. The criteria for the conservative management of cavernoma were when the seizures were controlled with antiepileptics, there was no progressive neurological deficit and when there were less than two hemorrhages. The patients' preference was taken into consideration. Two patients with frontal cavernoma were treated by SRS as they did not accept the surgical treatment. In radiosurgical treatment, Linear Accelerator (Philips SL-25, UK), Isocentric subsystem (Philips K-X 200, UK), Brown-Roberts-Wells Head Frame (Radionics Co., USA) and X knife3 planning system (Radiosonics Software Applications Inc., USA) were used. 6 MeV X-ray was produced by Linear Accelerator. In all cases single isocenter, median 310 degree total arch angle (300-320) and median 6 arch (5-7) were used. Median 15 Gy (14-20) was applied to the peripheral zone of the lesion (80%) (Figure 2).

Results

A 40-year-old female who also had multiple sclerosis and was treated surgically for brainstem cavernoma died on the 11th postoperative day. Of the 13 patients treated by SRS, one patient with frontal cavernoma (9-year-old patient whose surgical treatment was not accepted by his parents) had an increase in seizure frequency refractory to medical treatment and underwent surgery in the 12th month after radiosurgery. The median follow-up period was 2.5 years (1-8 years). None of the patients in the series had any neurological or radiological deterioration.

Discussion

The etiology of cavernomas is unknown. Cranial radiation, coexistent vascular malformation, genetic, and hormonal factors all have been implicated for the cavernomas. The proportion of patients developing clinical symptoms is higher in the hereditary form than in the sporadic form of the cavernoma.[9-16] De novo formation of a cavernoma during immunosuppressive treatment has also been reported.[17] There are some radiation-induced cavernomas occurring in childhood and adolescence.[18]

Cavernoma was reported in 0.39% of 8131 patients evaluated with craniospinal MRI.[19] Our incidence was 3%, with 37

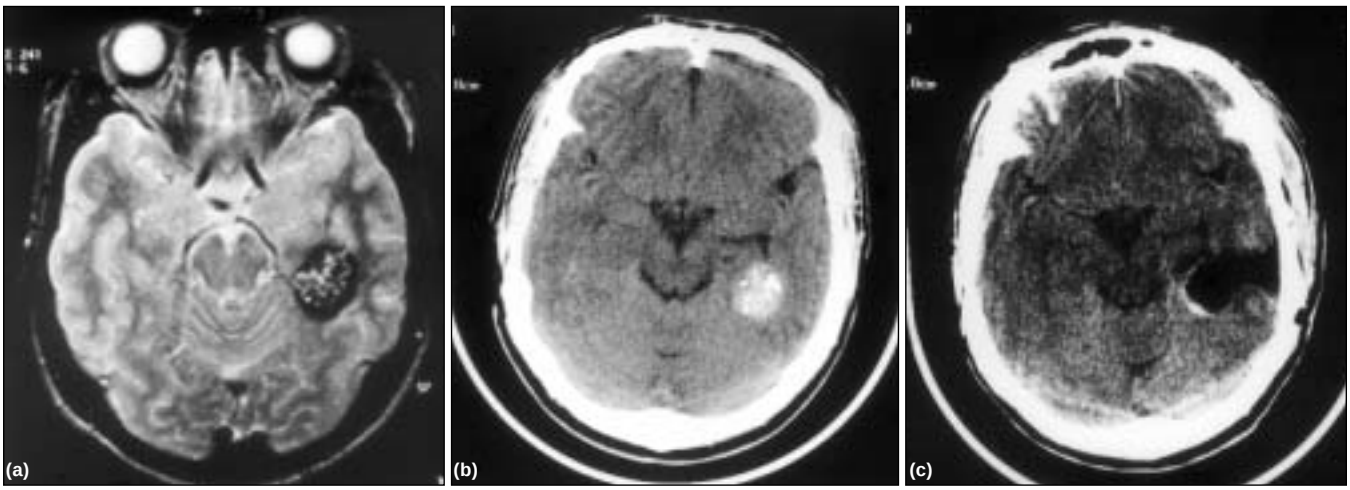

Figure 1: (a) Preoperative axial MRI of a calcified cavernous angioma is located left temporal lobe (b) Preoperative CT of the same patient (c) Postoperative CT of the same patient(pathologic diagnosis is same)

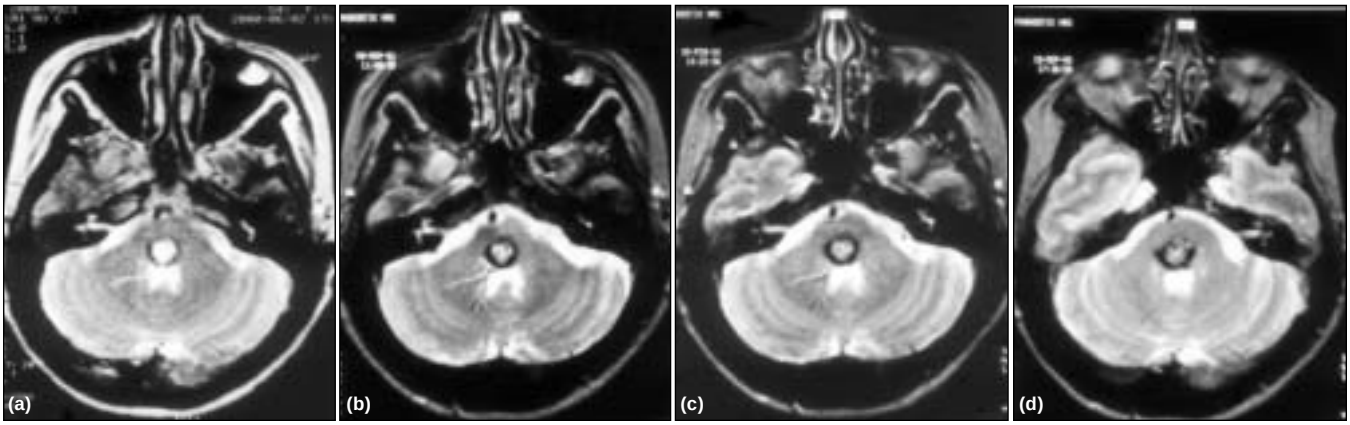

Figure 2: (a) T2 weighted axial MRI of a cavernous angioma is located brainstem. Before SRS (b) 6th month after SRS (c) 9th month after SRS (d) 16th month after SRS.There is no edema around the lesion, no increase in size of the lesion and there is no bleeding after radiosurgery

patients with cavernous angioma out of 1228 patients with intracranial mass lesion treated during the period. The majority of the cavernomas are supratentorial in location.[19] In our series 43% of the lesions were located infratentorially. Approximately 60% patients present with seizures, 50% patients have progressive neurological deficit and 20% patients have hemorrhage in intracranial cavernomas.[20,21] Despite the preponderance of cavernomas above the tentorium cerebelli, the general belief is that hemorrhage is more likely to occur from infratentorial lesions.[19,21] The location of the lesion is an important factor in predicting hemorrhage or the neurological course. In another series all hemorrhages occurred in the cavernomas located supratentorially.[22] In our series hemorrhage was observed in 5 patients (13.5%). Three of the hemorrhages (60%) were supratentorial.

MR features of cavernomas are characteristic and diagnostic of these lesions obviating the need for angiography.[23] Evidence of a recent or old hemorrhage is commonly present in cases of cavernomas, including hemosiderin-laden macrophages, cholesterol crystals and deposits of hemosiderin in the surrounding parenchyma.[24] We observed calcified cavernomas in two patients. Calcified cavernomas are called "hemangioma calcificans" or "brain stone".[25-27] Cavernomas have been located in a variety of cranial sites.[28-38] Pediatric cavernomas are still diagnostically and therapeutically challenging lesions. There is a higher risk of hemorrhage in children when compared to adults.[39]

Although some authors have stated that there was no difference in the hemorrhage rate before and after radiosurgery,[6] other authors recommend it especially for the brainstem, deep and eloquent located cavernomas, and a significant reduction has been observed in the annual hemorrhage rate after radiosurgery.[40-46]

X-knife radiosurgery was performed for 13 patients in our series and the results were uneventful except for one pediatric case. Hemorrhage, edema around the lesions and increase in the size of the lesions were not observed in our 13 patients treated radiosurgically up to date (Figure 2).

Although the dominant role of surgery in cavernomas is recently undergoing re-evaluation, an accessible causative cavernoma is an indication for surgical resection.[19]

Conclusion

Conservative treatment for cavernoma should be the choice for the patients without new or progressive neurological deficits and two or more documented hemorrhages and with seizure responsive to the medical treatment. For the others, the treatment is primarily surgical for the cases with non-eloquent locations and the results have been best where complete excision is achieved. Radiosurgery may be an alternative for treatment of deep and eloquent area located cavernomas and for the patients who do not accept surgical treatment.

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