

Original papers

Cerebral cavernous angioma in children

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Abstract. Cerebral cavernous angioma is a rare vascular malformation at any age and is very rare in childhood. In the literature available to us, we have been able to trace only 50 cases, to which we have added the 6 cases from our own series. The incidence in pediatric group is higher at 0–2 years (26.8%) and at 13–16 years (35.7%). The clinical onset shows epilepsy in 45.4% of cases, hemorrhagic syndrome in 27.3%, intracranial hypertension in 16.4%, and focal neurological deficits in 10.9%. Furthermore, we discuss the neuroradiological features (CT, angiography, and MRI) and the therapy of pediatric cavernous angioma.

Key words: Cavernous angioma – Cerebral angiography – Computed tomography – Epilepsy – MRI – Surgery.

Cerebral cavernous angioma is a relatively uncommon vascular malformation, accounting for 5%–13% of all vascular malformations [20, 25, 28, 34]. As a rule, it presents clinically in the third, fourth, and fifth decades of life and very rarely in childhood [29, 39, 49]. We have been able to trace only 50 cases of cerebral cavernous angioma diagnosed in the first 16 years of life (Table 1). Of the 205 angiomas operated on in the Neurosurgery Section of the department (formerly Institute of Neurosurgery) between 1952 and 1987, 14 were cavernous (6.8%) and of these 6 (42.8%) were in patients under the age of 16. We analyze here the clinical features, diagnosis, and treatment of cerebral cavernous angiomas in children.

Case reports

Case 1

This 1-year-old boy was admitted because of generalized epileptic seizures over the previous 9 months and loss of strength in the left side of the body lasting 2 months. Neurological examination re-

vealed mild left hemiparesis. EEG showed signs of irritation in the right temporal and parietal lobes, with slight slowing in the right hemisphere. Plain X-rays of the skull were normal. Right carotidography showed a vascular blush at the ventricular trigone on the same side.

Right temporoparietal craniotomy was followed by exposure at the ventricular trigone of a well-demarcated reddish mass, the size of a walnut, which was gradually isolated and completely removed. On histological examination, it proved to be a cavernous angioma. The postoperative course was uneventful, and after 17 years the boy is symptom-free on a low dosage of antiepileptic therapy.

Case 2

A 7-year-old girl was admitted because of a 3-year history of left-sided Jacksonian seizures, which in the past few months had become an almost daily occurrence despite anticonvulsant therapy. Neurological examination was normal; EEG revealed signs of irritation in the right parietal lobe; a CT brain scan (Fig. 1) showed a slightly hyperdense area at the level of the right parietal cortex, with poor contrast enhancement. Right carotidography revealed an avascular mass at the same site.

Right temporoparietal craniotomy was performed and a small, well-demarcated, dark red mass was removed totally. On histological examination, the lesion proved to be a cavernous angioma. Three years after operation the patient is seizure-free and requires no drugs.

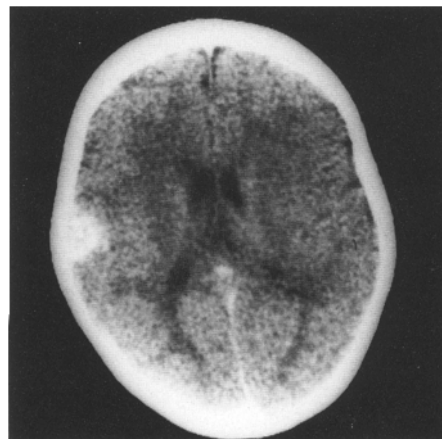


Fig. 1. Case 2: cerebral CT scan. Slightly hyperdense lesion in the right parietal lobe

Table 1. Reported cases of pediatric cerebral cavernous angioma. ICH, Intracranial hypertension; H, hemorrhage; FND, focal neurological deficits; S, seizure; LCE, low contrast enhancement; ME, mass effect; HE, hematoma; SI, signal intensity; y, years, m, months

Author	Year	Sex	Age	Clinical onset	Location	CT	MRI	Angiography	Surgery	Follow-up
Finkelnburg [9]	1905	M	2.5 y	ICH	IV ventricle	-	-	-	Partial removal	Postoperative death
Merritt [26]	1940	F	16 y	H (SAH)	Left, ventricle	-	-	-	Total removal	Worsened
Grotts [14]	1950	F	2 y	H (SAH)	Brain stem	-	-	-	Not done	Death
Aristein et al. [2]	1951	M	3 days	H	Right, ventricle	-	-	-	Partial removal	Postoperative death
McGuire et al. [26]	1954	M	3 m	ICH	Biventricular	-	-	-	Partial removal	Improved
Schneider and Liss [36]	1958	M	2 y	FND	Right, temporal	-	-	-	Total removal	Improved
Jain [19]	1966	F	15 y	ICH	Right ventricle	-	-	-	Total removal	Symptom free (6 m)
Genlach [11]	1969	F	6 y	H	Left, parietal-occipital	-	-	-	-	-
Aoki et al. [1]	1971	F	14 y	S	Left, frontal-parietal	-	-	-	-	-
Bartlett and Kishore [3]	1977	M	15 y	S	Left, temporal	Hyperdense	-	-	-	-
Giombini and Morello [12]	1978	M	14 y	S	Right, parietal-occipital	-	-	-	-	-
Savoirdo [35]	1978	M	8 y	FND	Left, frontal-parietal	-	-	-	-	-
	1978	F	10 y	S	Left, temporal	Hyperdense; edema	-	-	-	-
	1978	M	15 y	S	Left, parietal	Inhomogeneous; LCE	-	-	-	-
	1978	M	14 y	S	Left, frontal	Hyperdense; LCE	-	-	-	-
	1978	M	12 y	S	Left, parietal-occipital	Hyperdense	-	-	-	-
	1978	F	13 y	H	Right, parietal	Hyperdense	-	-	-	Symptom free (1 y)
	1978	F	4 y	H	Left, parietal	Hyperdense, ME	-	-	-	Improved (1 y)
	1978	F	13 y	S	Right, parietal	Hyperdense, ME	-	-	-	Improved (2 y)
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
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	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-
	1978	F	13 y	S	Right, parietal	Hyperdense	-	-	-	-
	1978	F	4 y	S	Left, parietal	Hyperdense	-	-	-	-

Table 1 (continued)

Author	Year	Sex	Age	Clinical onset	Location	CT	MRI	Angiography	Surgery	Follow-up
Present report	1988	M	14 y	FND	Multiple Brain stem	Hyperdense	-	Normal	Total removal	Symptom free
		F	7 y	FND		Hyperdense	Mixed SI; hypointensity rim	-	Total removal	Improved
	M	1 y	S	Right, ventricle	-	Hyperdense; LCE	-	Vascular blush	Total removal	Symptom free (17 y)
	F	7 y	S	Right, parietal		Hyperdense; LCE	-	Avascular mass	Total removal	Symptom free (3 y)
	M	5 y	ICH	Left, frontal		Hyperdense; LCE; calcified; HE	-	Avascular mass	Total removal	Symptom free (3 y)
	F	1.5 y	S	Left, temporal		Inhomogeneous; ICE; edema; HE	-	Avascular mass	Total removal	Symptom free (3 y)
M	16 y	S	Right, parietal		Hyperdense; LCE; calcified	-	Blush; efferent vessel	Total removal	Symptom free (2 y)	
M	3 y	S	Left, frontal		Inhomogeneous; LCE	-	-	Total removal	Symptom free (6 m)	

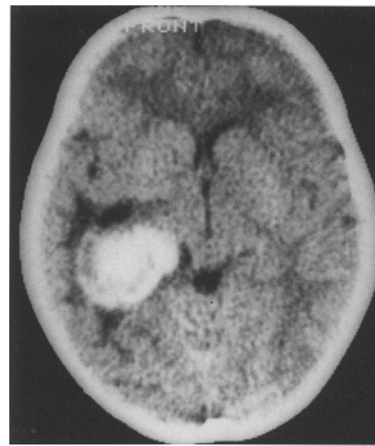


Fig. 2. Case 4: cerebral CT scan. Inhomogeneous lesion with central hematoma and peripheral edema in the left temporal lobe

Case 3

A 5-year-old left-handed boy had had an episode of acute intracranial hypertension with severe headache and vomiting 3 months before admission. Since then, he had had generalized epileptic seizures of increasing frequency. Neurological examination revealed bilateral papilledema. EEG showed signs of irritation in the left frontal lobe. A CT brain scan imaged a hyperdense lesion in the left frontal lobe, which enhanced poorly, with calcified areas and a collection of blood within the lesion. Left carotidography showed an avascular mass at this site.

Left frontal craniotomy, followed by prefrontal corticotomy, revealed pathological brown tissue, containing hemorrhagic and calcified areas, which were completely removed. On histological examination, the mass proved to be a cavernous angioma. The post-operative course was uneventful and 3 years later the child is completely free of symptoms and needs no drugs.

Case 4

This 1.5-year-old girl was hospitalized because of a 3-month history of right-sided Jacksonian seizures, poorly controlled by antiepileptic therapy. Neurological examination was negative. EEG showed signs of irritation in the left parietal lobe with slight left hemisphere slowing. A CT brain scan (Fig. 2) imaged a lesion of inhomogeneous density in the left temporoparietal region, surrounded by a hypodense zone. The lesion contained a hematoma and enhanced poorly. Carotidography showed a mass effect at the same site.

Left temporoparietal craniotomy, followed by incision of the first temporal gyrus, exposed a dark, encapsulated mass that extended as far as the tentorium and the choroid plexuses of the left temporal horn. The lesion, which was completely removed, proved on histological examination to be cavernous angioma. Two years after the operation, the patient is seizure-free and on low-dose anticonvulsants.

Case 5

A 16-year-old boy was admitted because of a 5-month history of almost daily, generalized epileptic seizures despite anticonvulsant therapy. Neurological examination was negative. EEG showed signs of irritation in the right parietal lobe. A CT brain scan visualized a small hyperdense area in the right parietal cortex, with calcified zones and slight contrast enhancement. Right carotidography showed a vascular blush with late venous drainage at the same level.

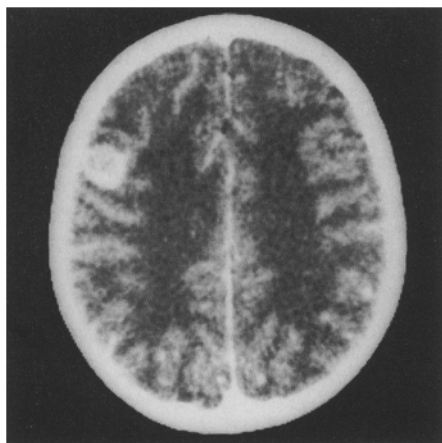


Fig. 3. Case 6: cerebral CT scan. Small lesion of inhomogeneous density in the left frontal cortex

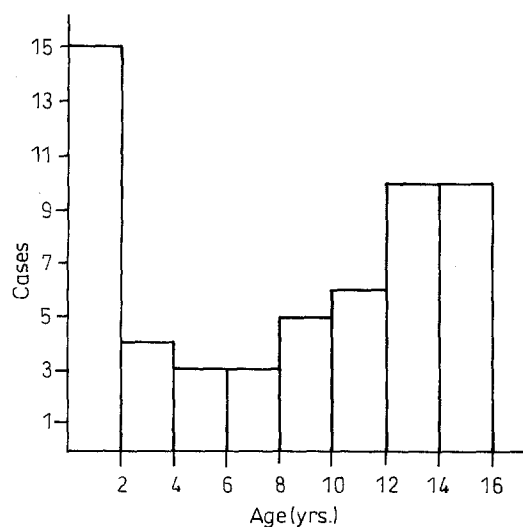


Fig. 4. Distribution of the 56 pediatric patients with cerebral cavernous angioma

After right parietal craniotomy, a walnut-sized reddish mass supplied by two small feeders was isolated. Closure of the feeders reduced the volume of the mass, which was totally removed. On histological examination, the lesion proved to be a cavernous angioma. The postoperative course was uneventful, and after 2 years the patient is seizure-free and on low-dose anticonvulsant therapy.

Case 6

This 3-year-old boy had a 2-year history of generalized epileptic seizures that occurred almost daily despite anticonvulsant therapy. The neurological examination at admission was normal. EEG showed signs of irritation in the left frontal lobe. A CT brain scan (Fig. 3) imaged a small lesion of inhomogeneous density with slight contrast enhancement in the left frontal cortex.

Left frontal craniotomy revealed a small, well-defined, bluish cortical mass, which was removed completely. The histological diagnosis was cavernous angioma. Six months after the operation, the patient is free of epileptic seizures and takes anticonvulsant drugs.

Discussion

Table 1 lists 56 cases of childhood cavernous angioma. The mean age is 8.3 years with two peak frequencies (Fig. 4): at 0–2 years (26.8%) and at 13–16 years (35.7%). There is no significant sex preference. The mean length of the clinical history is 27 months. The lesions presented clinically with epilepsy in 45.4% of the cases, with a hemorrhagic syndrome in 27.3%, intracranial hypertension in 16.4%, and focal neurological deficits in 10.9%. The most frequent sites were: parietal lobe (22.6%), ventricular system (18.9%), frontal lobe (17%), and temporal lobe (15.1%). There was 1 case of multiple cavernoma (frontal, cerebellar and temporal) [49]. Angiography, performed in 45 cases, visualized an avascular mass in 60%, a vascular blush or venous drainage in 13.3%, and was negative in 26.7%. CT scanning, done in 41 cases, imaged a slightly or moderately hyperdense lesion in 85.4% of cases, slight contrast enhancement in 46.3%, mass effect in 19.5%, calcifications in 19.5%, hematoma in 12.2%, an inhomogeneous area in 9.8%, edema in 4.9%, and a hypodense (cystic) lesion in 2.4% [21]. In 1 case [32], CT was normal. MRI, done in 4 cases [32, 49] showed an area of mixed signal intensity (SI), with decreased SI rim in all cases. Surgical removal was total in 87.2% and partial in 12.8%. In 1 case the diagnosis was established post mortem [14]. The perioperative mortality was 4.6%. After a mean follow-up of 2.4 years, the symptoms had cleared up altogether in 72.1% of the patients and improved in 18.6%; the deficits were stable in 2.3% and had worsened in 2.3%. The epileptic syndrome was relieved in all cases, totally and without antiepileptic therapy in 65% and with therapy, and/or reduced seizure frequency in 35%. There were no recurrences in patients who underwent total removal (Table 2).

Epilepsy, the most frequent presenting symptom in patients with cavernous angioma, is probably the outcome of the interaction of several epileptogenic factors: (a) frequently the cortical site (67.9% of pediatric cases); (b) the frequent presence of calcification within the surrounding parenchyma, which is marked by gliosis [29]; (c) accumulation of iron-containing substances produced by silent microhemorrhages [24] or by diffusion of pigment derived from the destruction of sequestered erythrocytes [31].

On the other hand, it is known that in childhood, especially during the second year of life, the likelihood of seizures is distinctly greater than at other ages. This probably explains why the epileptic syndrome (45.4%) is the most frequent symptom of onset of cavernous angioma in children, more so than in adults. Indeed, in grown-ups, according to Giombini and Morello [12], cavernous angioma presents with epilepsy in 38% of cases, with headache in 28%, with hemorrhagic syndrome in 23% and with focal neurological deficits in 12%. However, when considering the 56 pediatric patients by age group (0–2; 3–12; 13–16 years), we found no significant differences in the frequency of epilepsy.

Epilepsy may also be the clinical manifestation of hemorrhage. Bleeding is probably more frequent [12, 39, 42, 44, 47, 48] than is clinically apparent (27.3% of the pediatric

Table 2. Clinical features of 56 cases of pediatric cavernous angioma

Age	
Mean	8.3 y
Sex	
Female	55.4%
Male	44.6%
Length of history	
Mean	27 m
Presenting symptoms	
Seizures	45.4%
Hemorrhagic syndrome	27.3%
Headache	16.4%
Focal neurological deficits	10.9%
Location	
Parietal	22.6%
Ventricular	18.9%
Frontal	17.0%
Temporal	15.1%
Cerebellar/brain stem	9.4%
Parietal-occipital	7.5%
Frontal-parietal	3.8%
Occipital	1.9%
Cavernous sinus	1.9%
Multiple	1.9%
Angiography	
Avascular mass	60.0%
Normal	26.7%
Vascular blush or pooling	13.3%
Computed tomography	
Hyperdense lesion	85.4%
Low contrast enhancement	46.3%
Mass effect	19.5%
Calcifications	19.5%
Hematoma	12.2%
Inhomogeneous area	9.8%
Edema	4.9%
Hypodense lesion	2.4%
Normal	2.4%
Magnetic resonance imaging	
Mixed SI + hypointense rim	100.0%
Surgical removal	
Total	87.2%
Partial	12.8%
Results	
Symptoms free	72.1%
Improved	18.6%
Unchanged	2.3%
Worsened	2.3%
Operative mortality	4.6%

cases), since it is often masked by an epileptic syndrome. Simard [39], reviewing 138 published cases of adults and children, and Yasargil ([49]: 22 cases) state that over 50% present with bleeding, macroscopic or microscopic, within the excised malformation. Epilepsy masked a hemorrhagic onset in our case 3, in which the patient presented with generalized epileptic syndrome, after initial intracranial hypertension: on the CT scan and at operation, an intralésional hematoma was present. A hemorrhagic onset is also probable in case 4, in which a collection of blood within the

mass was evident both on the CT scan and at operation. Furthermore, headache may have been the expression of recurrent episodes of subarachnoid or intracerebral hemorrhage [30].

The diagnosis of cavernous angioma, before the introduction of CT, in a child whose only symptom was epilepsy was often established late, as the seizures were well controlled by medical therapy for a long time. For this reason, neuroradiological investigations are unfortunately often ordered only when the seizures become more frequent and/or no longer respond to drugs and/or change their clinical features [29].

CT scanning is a reliable procedure in the diagnosis of cavernous angiomas, both in adults and in children and often show indicative, even if not always specific features [32, 39, 48]. The typical CT features of cavernous angioma are [22, 29, 43]: (a) a well-defined roundish lesion, slightly and inconstantly hyperdense (85.4% of pediatric cases) and, more rarely, the lesion may be inhomogeneous (9.8%), hypodense (2.4%) or isodense; (b) slight contrast enhancement (46.3%); (c) inconstant mass effect (19.5%); (d) rare perilesional edema (4.9%). Angiographic diagnosis is more laborious, being negative in 26.7% of the pediatric cases and showing an avascular mass of doubtful nature in 60%. Only in 13.3% of pediatric cases did the angiogram show venous pooling or a vascular blush suggestive of cavernous angioma [7, 35, 42]. The introduction of MRI has made the preoperative diagnosis of cavernous angioma surer. The vascular malformation, clearer in the T2-weighted images, appears as a roundish area of mixed SI ("lattice pattern") with a hypointense rim [13, 32]. Only 4 of the published cases of pediatric cavernous angioma have been studied with MRI [32, 49], which provided the preoperative diagnosis in all.

The treatment of choice of cavernous angioma is surgical, with total removal of the lesion if the site allows [12, 29, 39, 43, 45, 48]. This is the right course for patients with a long life expectancy, especially children who show definite improvement of epilepsy after removal of the lesion, and because of the high risk of bleeding. The upshot of our review of the literature is that 65% of patients with epilepsy who had undergone surgery no longer had seizures, as happened in our cases 2 and 3. In the remaining 35% of the cases, the seizures were controlled with antiepileptic drugs and/or were reduced in frequency. Our patients 1, 4, 5 and 6 are seizure-free but are on antiepileptic therapy, even though at low doses. However, the follow-up in the last three cases (2 years in cases 4 and 5, and 6 months in case 6) is too short for us to be able to say whether or not the drugs may be discontinued later on. The risk of the lesion bleeding is high, far higher [39, 49] than the 27.3% of cases in which it presents clinically. Of the 15 pediatric patients in whom cavernous angioma had a hemorrhagic onset, 2 (13.3%) died [2, 14]. Furthermore, in 4 cases (26.6%) hemorrhage left the patients with more or less disabling neurological deficits. These factors, and the proven autonomous growth potential of cavernous angioma [46], apparent also from sequential CT scans [4, 8, 17, 48, 49], make early diagnosis and removal of the lesion essential.

At sites carrying a high surgical risk, the therapeutic trend is different if cavernous angioma is symptomatic or asymptomatic (chance finding). In the first case surgical removal of the lesion by a skilled surgeon is advisable because cavernous angioma is generally well circumscribed and surrounded by a gliotic plane that makes surgical removal relatively easy, with little damage to contiguous parenchyma (even if the lesion is not very large) [49]. Moreover, cavernous angioma has few arterial input and a slow rate of blood flow, with good control of bleeding during the operation relatively easy to accomplish [48]. In the second case, it is prudent to exam children with sequential CT and MRI. If these examinations show that the lesion is growing or there are signs of a previous hemorrhage, or if the cavernous angioma reveals itself clinically, the appropriate course is surgery.

Some authors [12, 36, 47] suggest stereotaxic radiotherapy of cavernous angioma, which may produce a reduction in volume of the lesion, thereby facilitating subsequent surgical excision [37]. However, the effectiveness of radiotherapy is by no means sure [12, 48], and its results, if any, do not occur for some months [37], depending on a mechanism of intravascular obliteration [48], during which time the child is at risk of hemorrhaging.

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