Child's Nerv Syst (1992) 8:49–52

Child's Nervous System CNS

Intracranial cavernous angioma in the 1st year of life and a review of the literature

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Received January 4, 1991

Abstract. Three cases of symptomatic cavernous angioma occurring in the 1st year of life are presented. All three patients were treated with radical extirpation. Postoperatively, two of them improved, but the third died due to cerebral herniation secondary to recurrence of bleeding of the lesion. The clinical characteristics of cavernous angioma in the 1st year of life are briefly discussed and a review of the literature presented, including 16 previous cases operated on directly.

Key words: Cavernous angioma – Surgery – Literature review

In the present study, we report our experience with three cases of intracranial cavernous angioma in the 1st year of life. The clinical aspects of the lesion are discussed and a review of the literature is presented.

Case reports

Case 1

A 6-month-old boy was admitted on 2 August 1990 because of abrupt convulsive attacks. On admission, there were no abnormal findings on neurological examination except for motor weakness of the extremities on the right side. Computed tomography (CT) scan showed, in the left parietal lobe adjacent to the lateral ventricle, a well-defined, inhomogeneous hyperdense mass, 1.5 cm in diameter, partly with calcifications (Fig. 1, top left); the effect of contrast enhancement on the lesion was scant (Fig. 1, top right). Magnetic resonance imaging (MRI) revealed a homogenous, hyperintense mass, 1.5 cm in diameter, on T_1- and T_2-weighted images (Fig. 1, center left and center). In addition, proton imaging of the lesion clearly showed an inhomogeneous hyperintensity and an obvious hemosiderin rim (Fig. 1, center right).

A transcortical approach with parietal craniotomy was chosen, and microscopically a dark-red lesion, partially composed of hematoma, was totally removed. Histological examination of the specimen verified a hemorrhagic cavernous angioma. The patient's postoperative course was uneventful. After a follow-up period of 4 months, the patient is symptom-free, with no residual lesion except for a very small high-density spot on CT (Fig. 1, bottom).

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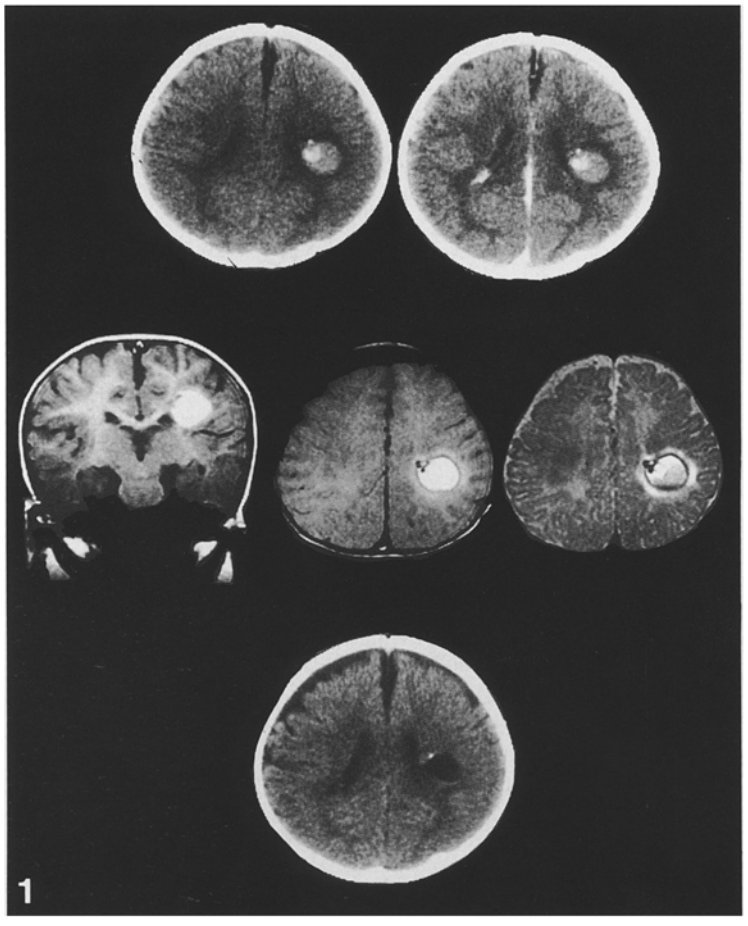
Case 2

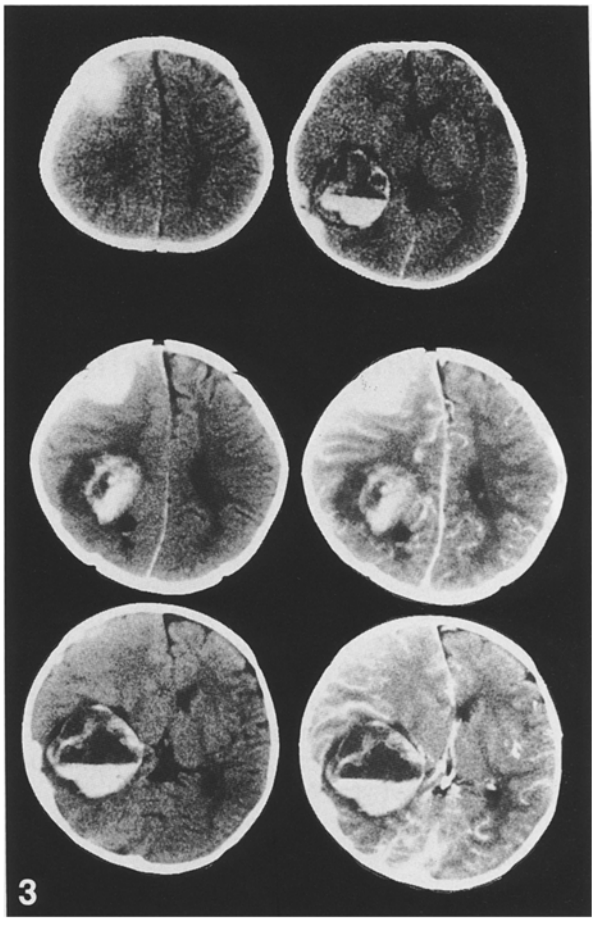
A 1-month-old boy was admitted on 14 July 1990 because of sudden inactivity and an increase of body temperature (39 °C). There were no abnormalities relating to his delivery. CT scan showed, in the right frontal lobe, a large inhomogeneous hyperdense mass, 6 cm in diameter (Fig. 2, left), with slight contrast enhancement, suggesting a cystic lesion with bleeding (Fig. 2, center). A frontal craniotomy was immediately carried out, and disclosed a large cortical mass with clearly defined limits; a large cyst containing brownish fluid was evacuated, and then the mass was completely removed with a small amount of bleeding. Histological diagnosis was a typical cavernous angioma. The postoperative course was uneventful. The patient is now symptom-free with no residual lesion on follow-up CT (Fig. 2, right).

Case 3

An 8-month-old boy was admitted on 26 May 1990 because of abrupt, repeated convulsive attacks originating in the upper extremity on the right side, and fever (38 °C). The patient was alert upon admission, although the anterior fontanel was tense.

CT scan showed two lesions: one was located in the right frontal region with a homogeneously high density, and was 2.5 cm in diameter (Fig. 3, top left); the other was located in the right parietal region with an inhomogeneously high density, and was 4 cm in diameter (Fig. 3, top right). CT was repeated 4 h after admission because the patient became drowsy and showed that both lesions had not noticeably changed in size, although the edematous changes around the lesion of the parietal lobe were increasing (Fig. 3, center). Furthermore, 2 h later, a fronto-parieto-temporal craniotomy had to be performed urgently due to the appearance of signs of uncal herniation on the right side. The preoperative CT scan demonstrated more clearly enhancement of the parietal lesion, 6 cm in diameter, and displacement of cerebral midline structure (Fig. 3, bottom). The two lesions were successfully extirpated in one stage. Histopathologically, cavernous angioma was verified in the two lesions. No improvement in the level of consciousness was seen postoperatively; the postoperative CT scan showed markedly edematous changes of the whole brain, following cerebral infarction in the territory of the right posterior cerebral artery. The patient died 1 month after the operation.





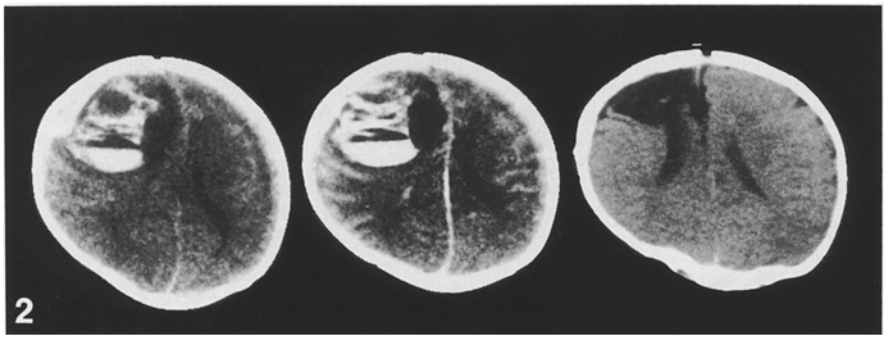


Fig. 1. Case 1. *Top:* Preoperative CT scans before (*left*) and after (*right*) contrast enhancement: in the parietal region, inhomogeneous hyperdense lesion 1.5 cm in diameter. *Center:* Preoperative MR images: homogeneous hyperintensity and hemosiderin rim. *Bottom:* Postoperative CT scan

Fig. 2. Case 2. Preoperative CT scan before (*left*) and after (*center*) contrast enhancement: in the frontal region, inhomogenous hyperdense mass with cystic component. *Right*: Postoperative CT scan

Fig. 3. Case 3. *Top:* CT scans before contrast enhancement at admission: in the frontal and parietal lobes, homogeneous and inhomogeneous hyperdense lesions respectively. *Center:* CT scans after contrast enhancement 4 h after admission: increase of the perilesional edema, although both lesion sizes were not changed. *Bottom:* Preoperative CT scans before (*left*) and after (*right*) contrast enhancement more than 2 h later: the size of the parietal lesion increased 1.5 times

Discussion

Recently, intracranial cavernous angioma has been reported frequently, its detection having been made easier by CT scan and MRI. It comprises 5–13% of CNS vascular anomalies [14]. Although this lesion has been found in all age groups [8, 16], it is exceedingly rare in the 1st year of life. In the literature available to us, we have been able to review only 16 cases that were histopathologically verified and directly operated on (Table 1). We have now added 3 of 26 cases from our own series. The age ranged widely from neonates to infants of 12 months, with a mean age of 5 months. There was no significant predilection as regards sex. Onset of symptoms consisted of convulsive attacks in 47.4% of the cases, an enlarged head in 31.6%, and various other symptoms. The lesion sites

Table 1. Literature cases histopathologically verified and directly operated cavernous angiomas

Author	Age	Sex	Onset symptoms	Location (size, cm)	Surgery	Outcome
McGuire et al. (1954) [9]	3 months	F	Head enlargement	Both lateral ventricles (1.8 × 0.8 × 0.5)	Partial removal	Improved
Sonntag et al. (1981) [17]	4 weeks	F	Head enlargement	Pineal region (1.5)	Total removal	Improved
Sugiyama et al. (1981) [18]	8 months	M	Generalized seizures	Left temporal (7 × 4 × 4)	Total removal	Cured
Iwasa et al. (1983) [5]	8 days	M	Head enlargement	Left lateral ventricle (3)	Total removal	Improved
Jooma et al. (1984) [6]	–[a]	M	Head enlargement	Cerebellar hemisphere	Total removal	Cured
Chadduck et al. (1985) [1]	4 months	F	Generalized seizures	Right lateral ventricle (4)	Total removal	Cured
Moritake et al. (1985) [11]	Neonate	F	Prenatal diagnosis	Right cerebellar tentorium (4 × 4.5 × 5)	Total removal	Cured
Yamasaki et al. (1986) [20]	2 months	F	Generalized seizures	Right frontal (3 × 3 × 3.5)	Total removal	Cured
Miyagi et al. (1987) [10]	2 months	M	Head enlargement	Right frontal (1.5 × 2 × 2)	Total removal	Good (hemiparesis)
Yasargil (1988) [21]	4 months	F	Swelling (left parietal)	Left parietal (6)	Total removal	Cured
Gangemi et al. (1989) [3]	9 months	F	Generalized seizures	Right frontal (large)	Total removal	Cured
	6 months	F	Head enlargement	Right frontal (large)	Total removal	Cured
Okada et al. (1989) [12]	4 months	F	Generalized seizures	Right frontal (6 × 4 × 2.5)	Total removal	Cured
Fortuna et al. (1989) [2]	12 months	M	Generalized seizures	Right ventricle (walnut size)	Total removal	Cured
Hubert et al. (1989) [4]	9 months	F	Hemorrhage	Left frontoparietal	Yes	–[a]
	10 months	M	Epilepsy	Right frontoparietal	Yes	–[a]

[a] Not described

were supratentorial in 85.7% and infratentorial in 14.3%. The most frequent sites in a total of 21 lesions, including 2 cases with multiple lesions (10.5%) were: frontal lobe (38.1%), lateral ventricle (28.6%), parietal lobe (14.3%), and cerebellum (10.5%). Of the 2 cases with multiple lesions, one was located in biventricular [9] and the other in frontal and parietal lobes of the right side (our present case 3). No significant laterality of the lesions was seen.

In general, if onset of symptoms and lesion sites of cavernous angioma in the 1st year of life are compared with those in other age groups [2, 8, 16], an enlarged head and lateral ventricle were most frequent, respectively. CT scans, performed in 14 cases with 15 lesions, showed a round hyperdensity mass with low contrast enhancement in the majority. Of all the lesions, 10 were homogeneous. Three of 5 lesions with inhomogeneous hyperdensity were accompanied by a large cyst [3, 12]. Such cyst formation seems to be characteristic of cavernous angioma in the 1st year of life. MRI was only performed in 2 cases. One is a case of Hubert et al. [4], in which the lesion showed an inhomogeneous high intensity, and the other is case 1 in our present series, in which the lesion demonstrated a homogeneous high intensity and a thin rim of hypointensity (hemosiderin rim). In general, the findings of CT and MRI of cavernous angiomas in the 1st year of life were similar to those in other age groups [7, 13, 19]. Surgical removal was total in the majority of cases, and yielded favorable results in all but 1 case, whereas a case of cavernous angioma with recurrence of bleeding has been reported, in which only the hematoma was removed [15]. However, our present case 3 had worsened within about 6 h after admission, and CT scans revealed an acute enlargement of the lesion, probably inducing cerebral infarction and edema. Such an abrupt hemorrhage of the lesion involving a cerebral vascular disorder should be noticed in the 1st year of life.

We can conclude that CT scan and/or MRI have made it easy to diagnose cavernous angioma easy, and radical extirpation of cavernous angiomas, even during the 1st year of life, is strongly recommended at an early stage especially in symptomatic cases, to avoid the risk of hemorrhage and of secondary enlargement or recurrence.

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