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Cerebral arteriovenous malformations in children: report on 62 cases

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Abstract A series of 62 children with cerebral arteriovenous malformations admitted to our department in the course of 17 years (1975–1992) was reviewed in a retrospective study. In 54 cases hemorrhagic stroke was the first presenting symptom, followed by epilepsy in five cases. On admission 26 children presented with a neurological deficit, and 21 were admitted with a grade 3 status according to Botterell. Fifty-one malformations were supratentorial (41 hemispheric, 10 deep-seated) while 11 were infratentorial. According to Mori's criteria, 28 lesions were small, 19 medium, and 15 large. Fifty-two children were operated on, with total excision of the malformation achieved in 47 cases. In two children the malformation recurred. The evolution of neurological disorders has been studied with a mean follow-up of 8.5 years. Fifty patients had a satisfactory outcome on the Glasgow Outcome Scale. Four children died. These results were compared with those reported elsewhere in the pediatric literature.

Key words Cerebral arteriovenous malformation · Children · Surgery Embolization · Radiosurgery Recurrence

Introduction

Arteriovenous malformations (AVMs) are congenital but the clinical signs usually first appear in adulthood. Clinically manifest AVMs in children are rare and few pediatric series have been reported in the literature [4, 7, 10, 16, 23].

In children the most common initial symptom of AVM is hemorrhagic stroke [7, 10, 16, 36], with high mortality and morbidity rates [4, 10]. In the past, surgery was the only means of therapy. The developments of radiosurgery and embolization have considerably modified the therapeutic management of AVMs.

The purpose of this paper is to report our experience with 62 cases of AVMs in children with long-term follow-up. The therapeutic results were compared with those in other pediatric series in the literature.

Patients and methods

Between 1957 and 1992, 106 children with intracranial AVMs were admitted to our department, but only the 62 cases admitted after 1975 were reviewed for this study, to make up an homogeneous group. Aneurysms of the vein of Galen or dural AVMs were excluded.

Clinical data were reviewed. The diagnosis of intracranial AVM was made on the basis of cerebral angiography in 56 cases and during surgery for intracerebral hematoma in 6 cases. In all cases the diagnosis was documented by histopathological study.

Follup-up data were obtained from either a recent interview and physical examination or a questionnaire answered by the patient, the patient's parents, or the family physician. In some cases physical examination was carried out during a visit at the patient's home. The period of follow-up ranged from 1 year to 17 years, with an average of 8.5 years. The condition of each patient was rated according to the Glasgow Outcome Scale.

Results

Clinical material

The study group consisted of 29 male and 33 female patients. Ages ranged from 3 months to 14 years (mean 9.8 years; Fig. 1). No patient had a family history of AVM. Seven children had a previous history of headache, five had a previous history of epilepsy (partial seizures in three, generalized seizures in two).

Clinical symptoms

Intracranial hemorrhage was the most frequent symptom (54 cases), with intracerebral hematoma in all cases (Fig. 2). In three cases, seizures occurred at the time of hemorrhage. In three children head trauma had occurred before the intracranial hemorrhage. Epilepsy was the first symptom in 5 patients (8%). Of the remaining 3 patients, two presented with headaches and one with congestive heart failure.

Clinical examination on admission

Clinical data in the form of the Botterell score are summarized in Table 1. Fundoscopic examination was performed in 50 patients (3 with hemorrhage, 4 with edema). It revealed no abnormalities in 35 patients who presented with intracranial hemorrhage (81%).

Radiological findings

Skull plain films were performed in all cases. They were normal in all patients but one who presented with intracranial calcifications.

Computerized tomography (CT) was available in our institution from 1980. CT was carried out in 49 patients on admission (79%) and showed intracerebral hematoma in 42. In two cases, CT disclosed a hematoma and the AVM. In 5 other patients CT with infusion of contrast revealed the AVM. The size of the intracerebral hematoma is shown in relation to the size of the AVMs in Table 2. Magnetic resonance imaging (MRI) was only performed in 12 recent cases.

Cerebral angiography was performed in all cases. The site and size of the AVM were classified according to the criteria of Mori et al. [26]. When the AVM was not visible on angiography (6 cases), its site and size were defined according to the data collected during surgery. Most AVMs were supratentorial (82%); 41 were hemispheric (66%). The most frequent location was the parietal lobe (15 cases); 10 were deeply located (8 in the basal ganglia, 2 involving the corpus callosum). The remaining AVMs were located in the posterior fossa, 10 in the cerebellum, 1 in the brain stem (Fig. 3). There were 28 small lesions, 19 medium-sized and 15 large (Table 2). The main feeding vessels were the middle cerebral artery (34 cases), the anterior cerebral artery (21 cases), and the posterior cerebral artery (15 cases).

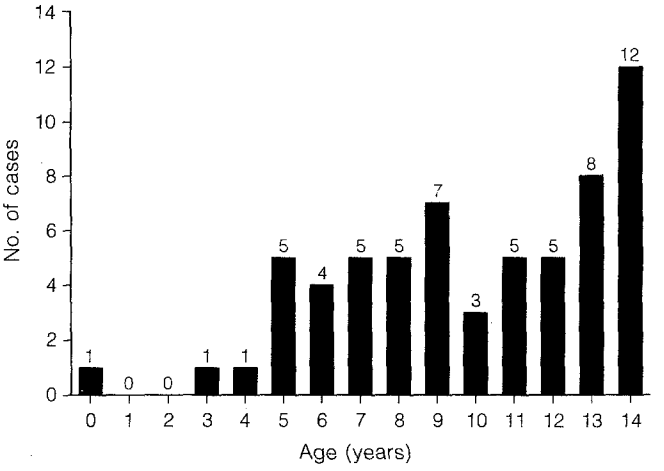

Fig. 1 Age distribution in the study group

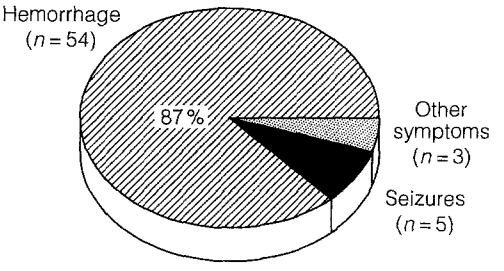

Fig. 2 Clinical symptoms at time of diagnosis

Table 1 Clinical symptoms on admission: Botterell score

Grade 0	Grade I	Grade II	Grade III	Grade IV	Grade V
$n=8$	$n=10$	$n=15$	$n=21$	$n=7$	$n=1$

Table 2 Size of arteriovenous malformations (AVMs) in relation to size of hematoma

Size of hematoma	Size of AVM			
	<2 cm	2–4 cm	>4 cm	Total
None	0	2	6	8
<2 cm	7	3	2	12
2–4 cm	10	10	2	22
>4 cm	11	4	5	20
Total	28	19	15	62

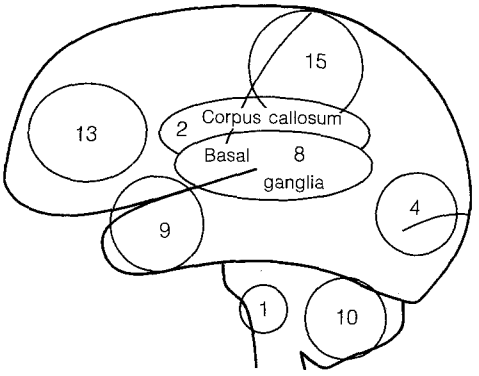

Fig. 3 Locations of arteriovenous malformations (AVMs)

Table 3 Therapeutic management of AVMs in 62 children

Treatment	No. of cases
Surgery	52 (+2 embolizations)
Embolization alone	2
Embolization + radiosurgery	4
Radiosurgery alone	3
Conservative treatment	1
Total	62

Table 4 Number of surgical operations and extent of AVM excision

	No. of operations				Total
	1	2	3	4	
Complete excision	40	5	1	1	47
Partial excision	1	2	2	0	5

Table 5 Clinical outcome as shown by Glasgow outcome scale

Good	Mild	Poor	Death	Total
$n=50$	$n=6$	$n=2$	$n=4$	$n=62$

Management

Conservative treatment was indicated in one case (a huge unrupted AVM). Fifty-two children were operated on (84%), in two cases with pre- or postoperative embolization (Table 3). Total surgical excision was performed in 47 cases (45 confirmed by early or late angiography), and partial excision in 5 cases. Table 4 shows the number of surgical procedures needed to obtain these results. Of the remaining nine children, two were treated with embolization only (partial obliteration in one case and total obliteration in the other), three underwent radiosurgery only (linear accelerator Saturne 18 MeV) with total disappearance of the AVM 18 months after irradiation in two cases and reduction in size of the AVM in one, and four underwent radiosurgery after reduction in size of the AVM by embolization.

Follow-up

Table 5 summarizes clinical outcomes according to the Glasgow Outcome Scale.

Mortality

Four children died (6.5%). Death was related to severe hemorrhage in one case, to the surgical procedure in two other cases (AVM located in the corpus callosum in one, infratentorial in the other). One child died after a recurrent hemorrhage 13 years after the initial hemorrhage (partial excision).

Recurrence of hemorrhage

In addition to the fatal hemorrhages, recurrent hemorrhage was seen in two children. One was a 6-year-old girl operated on for a cerebellar malformation revealed by hemorrhage, with total excision confirmed by early postoperative angiography. Two years later she was admitted for a recurrent hemorrhage (cerebellar hematoma) located in the same area. A preoperative angiogram was negative, and during surgery no malformation was identified. A postoperative angiogram was normal. The other case was in a 12-year-old boy admitted for an intracerebral hematoma related to a deep AVM. No surgical procedure was possible and this malformation was embolized with partial success (four procedures). A second and third hemorrhage occurred respectively 1 and 8 years later, with worsening of a residual hemiparesis.

Recurrence of AVM

Recurrence of the AVM was defined as resurgence of the AVM after treatment despite an early postoperative angiogram showing total excision after primary treatment. Two children presented with a recurrence located in the same area (AVM located in the corpus callosum in one case, in the parietal lobe in the other). These recurrences were revealed by late angiography. These patients underwent radiosurgery, with total disappearance of the recurrence, 15 and 18 months respectively after treatment.

Outcome in regard to aphasia

Ten children had aphasia before treatment. Of these, five are improved, four have mild residual aphasia, and one has severe residual aphasia. One child with no preoperative aphasia suffered impairment in this respect after surgery.

Outcome in regard to hemiparesis

Twenty-five children presented with hemiparesis on admission. Twelve were improved (48%) after treatment and at the present time have no residual deficit. Six (24%) have mild residual hemiparesis (four hematomas, two AVMs in a functional area), and seven (28%) have a severe residual deficit (three hematomas, four AVms in a functional area). Of the remaining patients who had no preoperative deficit, two (6%) developed a deficit after surgery; the AVMs were located in a functional area in both cases.

Outcome in regard to epilepsy

Five children had seizures before treatment; three were operated on and two are seizure-free without any treatment. Of three patients who experienced seizures during or just after intracranial hemorrhage, one is seizure-free and two have residual epilepsy (generalized seizures). Of the 47 surviving patients who were operated on with no preoperative seizures and who presented with intracranial hemorrhage only, seven patients (14.8%) have residual epilepsy, well controlled by the treatment in six cases (generalized seizures).

Discussion

Despite advances in new therapeutic techniques such as radiosurgery and embolization, we believe that surgery still remains the most reliable treatment for cerebral AVMs in children. Nevertheless, the management of large AVMs or of AVMs located in functional areas requires a multidisciplinary approach to combine the experience of the neurosurgery, embolization, and radiosurgery teams.

Brain AVMs are uncommon and few series in the literature give details of their real prevalence. McCormick [24], in a series of 5850 consecutive autopsies, found 30 individuals (0.52%) with a brain AVM. The incidence is seven times lower than that of aneurysms [31]. In different reports including children and adults, the incidence of AVMs in children usually ranges from 15% to 23% [11, 39, 46].

In children, cerebral hemorrhage is the most frequent initial symptom (70%–90%) [7, 10, 16, 36, 46]; in our series the figure was 87%. AVMs are the most frequent cause of intracranial hemorrhage in children after infancy [34, 35, 41]. The risk of hemorrhage is lower in adults, ranging from 50% to 71% [1, 7, 39]. Humphreys et al. [16] suggest that this phenomenon is related to "the progressive biological activity of the malformation in children". Moreover, the development of other symptoms (epilepsy, neurological deficit, or other signs of cerebral ischemia) needs a long evolution time.

The role played by head trauma in bleeding from AVMs has been discussed. Nishi et al. [27] postulated that the relationship between AVM bleeding and head trauma was due to intracerebral shearing forces in the anteroposterior or posteroanterior directions.

The size of the malformation is an important factor for the estimation of the risk of hemorrhage. As in our series, the smaller the AVM, the higher the risk [4, 16, 26, 41]. Spetzler et al. [37] explained this phenomenon by the differences in arterial feeding pressure between small and large AVMs. Furthermore, Spetzler et al. reported an inverse relationship between AVM size and hematoma size. We did not find such a correlation in our series (see Table 2). Infratentorial or deep-seated AVMs bleed more frequently than cortical AVMs [17, 18] and the mortality is higher [9, 15]. Anatomopathological [49] and hemodynamic studies [27] have shown that the origin of hemorrhage is probably venous.

Epilepsy is the second most frequent presenting symptom, ranging in incidence from 7.1% to 14.2% (8% in our series) [10, 16, 26, 46]. In adults the incidence of epilepsy is higher and ranges from 22% to 28% [9, 11, 31]. Celli et al. [4] reported identical incidences of epilepsy in children and adults. Large AVMs are more frequently revealed by seizures than small AVMs [16, 17, 26, 41]. All our epileptic patients but one were at least 10 years old at the time of diagnosis, but we never found any similar observation in other pediatric reports [2, 10, 12]. The risk of bleeding in children, in case of AVMs revealed by epilepsy, ranges in the literature from 20% to 28% [10, 23]. However, in our series no patient who had an AVM revealed by epilepsy (five cases) suffered intracranial hemorrhage later on.

Despite improvements in CT scanning and MRI for diagnosis of AVMs, angiography still remains the best investigation for the diagnosis and therapeutic management of AVMs. It defines all the characteristics of the malformation: location, size, feeding vessels, and the angioarchitecture of the nidus. Ninety percent of AVMs are located in the supratentorial area [31], mostly fed by the middle cerebral artery, followed in order of frequency by the anterior cerebral artery and the posterior cerebral artery [10, 14]. Willinsky and Lasjaunias et al. [43] have studied the relationship between the angioarchitecture of the AVM (venous anomalies, aneurysmal dilatation,

aneurysms of arterial feeding) and the risk of bleeding. Aneurysm with AVM is a classical association in adults (9% of cases) [11], but this association is very rare in children [30].

During recent years, new methods of therapeutic management of AVMs have been developed. Follow-up of patients treated by radiosurgery is now long enough for an assessment of the results of this technique [3, 5, 22, 29, 38]. The efficacy and safety of radiosurgical treatment of AVMs in children are confirmed, with a total obliteration rate ranging from 50% to 54.8% at 1 year and near 80% at 2 years [5, 38]. Nevertheless, results are variable depending on the size and blood flow of the malformation. When the nidus is small and the flow within the malformation is reduced, results are certainly better [3, 8].

Embolization also appears an efficient and safe technique in the treatment of brain AVMs, especially before or after surgery and before radiosurgery. Few authors have reported total obliteration of the malformation with exclusive embolization [32, 33, 45]. Nevertheless, in the patients with partial obliteration of the AVM, headache and clinical condition related to steal syndrome are improved, although the risk of rebleeding seems to remain unchanged [21, 34, 44]. Complications are infrequent and depend mainly on the experience of the interventional neuroradiologist [32, 42].

The aim of treatment should be complete disappearance of the malformation, which is the only guarantee of cure. Surgery still remains the most reliable form of treatment especially in children, possibly in association with radiosurgery or embolization, when AVMs are located in functional areas or are large. Table 6 summarizes our opinions about the management of AVMs in children. In the series reported by Humphreys et al. [16], 77% of children underwent surgery and total excision was performed in 72% of cases (84% and 90% respectively in our series). Postoperative mortality in children ranged from 8.5% to 11%, versus a mortality of 23% to 57% with conservative treatment [7, 10, 16, 36]. Postoperative mortality is less in children than in adults [4] In pediatric series it is difficult to estimate postoperative morbidity, but it appears that in children the capacity to recover from pre- or postoperative neurological deficit is better. Humphreys et al. [16] called this characteristic the "child's biologic plasticity."

Table 6 Recommendations for management of AVMs in children

Deep AVMs	Small →	Radiosurgery
	Large →	Embolization ± radiosurgery
Superficial	<4 cm, nonfunctional area →	Surgery
	<4 cm, functional area →	Embolization ± surgery ± radiosurgery
	>4 cm, nonfunctional area →	Surgery ± embolization
	>4 cm, functional area →	Embolization ± radiosurgery

Recurrence of AVMs has rarely been reported [19]. Humphreys et al. [16] reported the case of a child who suddenly died at home 6 years after total resection of his malformation. The cause of death was never established, but it may have been related to rebleeding from a recurrence of the AVM. Recently, Lizuka et al. [20], in a pediatric series of multiple cerebral AVMs emphasized angiogenesis as an explanation of the evolution of acquired AVMs. Many authors have reported changes in the size of intracranial AVMs, but these malformations have never been operated on [6, 13, 25]. These authors have emphasized the growth potential of AVMs. In our cases we think that microshunts might remain invisible on early postoperative angiograms, but might increase in size during the following months. We feel that it is essential to perform late postoperative angiography (from 18 months to 2 years after treatment) to assess for total excision of the AVM and the absence of recurrence.

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