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ARTICLE

Intracranial cavernous malformation in children: a single-centered experience with 30 consecutive cases

Angiomas cavernosos intracranianos em crianças: experiência de um único centro em 30 casos consecutivos

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ABSTRACT

Objectives: To determine the clinical presentation and treatment outcome of pediatric intracranial cavernous malformation (CM) in a single-centered institution. **Methods:** Clinical data review of 30 patients under 18 years-old who had undergone surgery for cavernous malformation from January 1993 to December 2011. **Results:** The Study Group included 18 males and 12 females (mean age: 8.7 years-old). Symptoms at presentation were seizures (16/30, 53.3%), headache (15/30, 50.0%), and focal neurological deficits (11/30, 36.6%). Multiple cavernous malformations were found in 5/30 (16.6%). According to location, patients were classified in groups: (G1) brain-steam in 5/30 (16.6%), (G2) cerebellum in 2/30 (6.6%), (G3) supratentorial associated with seizures in 16/30 (53.3%), and (G4) supratentorial without seizures in 7/30 (23.3%). Surgical resection was performed in 26 out of 30 (86.6%) patients. The mean follow-up period was 4.1 years. Of 15 children followed-up with preoperative seizures, all were rendered seizure-free after surgery. **Conclusions:** For symptomatic solitary cavernous malformation, the treatment of choice is complete microsurgical excision preceded by careful anatomical and functional evaluation. For multiple cavernous malformation or asymptomatic patients, the treatment modalities must be cautiously considered.

Key words: hemangioma, cavernous, seizure, brain tumor.

RESUMO

Objetivos: Determinar a apresentação clínica e o acompanhamento do tratamento em crianças com angioma cavernoso intracraniano numa única instituição. **Métodos:** Revisão de dados clínicos de 30 pacientes menores de 18 anos com que passaram por uma cirurgia de angioma cavernoso intracraniano entre janeiro de 1993 a dezembro de 2011. **Resultados:** O grupo de estudo incluiu 18 sujeitos masculinos e 12 femininos (idade média: 8,7 anos). Os sintomas iniciais eram convulsões (16/30, 53,3%), cefaleia (15/30, 50,0%) e déficits neurológicos focais (11/30, 36,6%). Havia angiomas cavernosos intracranianos múltiplos em 5 de 30 (16.6%). A classificação foi feita em grupos de acordo com a localização: (G1) tronco cerebral em 5/30 (16,6%); (G2) cerebelo em 2/30 (6,6%); (G3) supratentoriais associados a convulsões em 16/30 (53,3%) e (G4) supratentoriais sem convulsões em 7/30 (23,3%). Ressecção cirúrgica foi realizada em 26 de 30 (86,6%) pacientes, com seguimento médio de 4,1 anos. De 15 crianças com convulsões pré-operatórias, todas ficaram livres das crises após a cirurgia. **Conclusões:** Para angioma cavernoso intracraniano solitário e sintomático, o tratamento de escolha é excisão microcirúrgica total precedida de avaliação funcional e anatômica meticulosa. Para angiomas cavernosos intracranianos múltiplos ou pacientes assintomáticos, as modalidades terapêuticas devem ser consideradas cautelosamente.

Palavras-Chave: angioma cavernoso, convulsão, tumor cerebral.

Intracranial cavernous malformations (CMs) are common vascular anomalies in the brain and have an incidence of 0.1 to 0.5% in the general population[1].

These malformations represent 10 to 20% of all vascular lesions in the brain and can cause symptoms such as seizures, hemorrhages, headache, and focal neurological deficits[2,3]. The lesions are characterized by a "blackberry-like" aggregation of grossly enlarged capillary cavities consisting of a single layer of endothelium, without intervening neuronal tissue[4]. Since the introduction of sensitive diagnostic tools like the magnetic resonance imaging (MRI), CMs have become increasingly reported in the pertinent literature because asymptomatic lesions have also become detectable[2,5].

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Pediatric CMs may have characteristics that differ from those of adults in clinical presentation and treatment, such as higher rates of hemorrhage[6-10] and larger dimensions[9]. Even though there are not many reports about exclusive pediatric CMs, only a few large series have yet been reported, and ideal group classification has not yet been defined.

This report is an attempt to evaluate clinical and surgical data in a single-centered pediatric series of CMs in the context of published literature. Special emphasis was placed on management of different groups and on outcome of patients suffering from seizure prior to surgery.

METHODS

Patient population

We reviewed files of patients younger than 18 years harboring a CM, over an 18-year period at the Division of Pediatric Neurosurgery, University Hospital of Ribeirão Preto Medical School, from January 1993 to December 2011. Thirty CM cases were identified and enrolled in this study.

The variables analyzed included age, sex, clinical presentation, radiological features, extent of resection, and histopathology.

All patients had all been preoperatively evaluated by structural brain imaging with computed tomography (CT) and/or MRI. Shortly after surgical excision, all patients were put on regular follow-up.

The subjects were classified according to the main lesion location: brain-steam (G1), cerebellar (G2), supratentorial associated with seizures (G3), and supratentorial not associated with seizures (G4).

Postoperative seizure outcome was classified according to Engel's classification[11]: class I, free of disabling seizures; II, rare disabling seizures; III, worthwhile improvement; and IV, no worthwhile improvement.

A PubMed database search was also performed in order to retrieve articles published in the last 20 years, which combined the subject headings: cavernoma; cavernous angioma; pediatric; intracranial; and children. Papers with less than ten patients were excluded as well as series with patients above 20 years-old. Only articles in the English language containing all types of clinical manifestation of the CMs were considered, those concerning only seizures, brainstem, radiation-induced cavernomas or restricted types of cavernomas' classification were excluded from this review.

This study was approved by the Research Ethics Committee of the University Hospital of Ribeirão Preto Medical School, at University of São Paulo (Protocol 6591/2007).

RESULTS

Clinical and surgical data

The Study Group included 18 males and 12 females with a male-female ratio of 1.5:1. The mean age at onset ranged from 6 months to 17 years-old (mean 8.7 years-old). Patient's age and gender, CM location, clinical aspects, and outcome are summarized in Table 1.

The most common presenting symptoms were seizures in 16/30 (53.3%), headaches in 15/30 (50%), followed by focal neurological deficits in 11/30 (36.6%), and behavior disturbance in 1/30 (3.3%). Acute intracranial hemorrhage was presented in 16/30 (53.3%).

Single CMs were found in 25/30 (83%), while five patients (17%) had multiple ones. Regarding only symptomatic lesions, 23 (76.6%) supratentorial and 7 (23.3%) infratentorial, location was as follows: frontal lobe (7/30, 23.3%), temporal lobe (6/30, 20.0%), occipital lobe (3/30, 10.0%), parietal lobe (1/30, 3.3%), insula (1, 3.3%), thalamus and basal nuclei (4/30, 13.3%), brainstem (5/30, 16.6%), and cerebellar hemisphere (2/30, 6.6%).

A total of 26 CMs undergone surgery and four patients were managed conservatively. There were no postoperative deaths or significant complications in this series. No child was asymptomatic prior to the resection.

In the G1 (5/30), three patients were submitted to surgery (Fig 1). Two patients that showed spontaneous total improvement did not undergone surgery. In the G2 (2/30), microsurgical resection was performed after cerebellar hemorrhage in both patients (Fig 2).

In G3 (16/30), 11 patients underwent simple lesionectomy without further neurophisiological evaluation. Five patients had long-lasting symptoms (more than one year) or high frequency of seizures and were therefore deeply investigated and treated (Table 1). In cases of epilepsy, the surrounding hemosiderin-stained tissue had also been removed. Of 15 children followed-up with preoperative seizures, all of them were rendered seizure-free (Engel's class 1) after the CM removal.

Seven patients composed G4. Five cases among them were submitted to surgery (Fig 3), and two were managed conservatively (thalamic and basal ganglia CM).

Outcome/follow-up

The mean follow-up period was 4.1 years (range 6 months to 13 years). The overall post-treatment results were positive. Major morbidity and mortality from surgical procedures were absent. One patient (case 10) showed a permanent monoparesis after surgery.

Complete recovery of the neurological condition was achieved in 15 out of 17 (88.2%) children with neurological impairment prior to surgery (headache, focal deficits, and behavior disturbance).

Table 1. Summary of clinical-radiological characteristics and outcome of patients.

Pt	Age (years)	Gender	FH	ML	Localization of main lesion	Main Symptoms	Clinical Hemorrhage	Group	Surgery	Technique	Follow-up	Outcome
1	6	M	No	Yes	Pons	Coma	Yes	G1	No		2	No deficit
2	14	M	No	No	L cerebellar	IH, ataxia	Yes	G2	Yes	Lesionectomy	6	No deficit
3	0.6	F	No	No	L thalamus	Seizures, R hemiparesis	Yes	G3	Yes	Lesionectomy	3	Seizure-free (Engel1a)
4	6	M	No	No	R parietal	Seizures	No	G3	Yes	Lesionectomy	1	Seizure-free (Engel1a)
5	16	F	No	No	Pons	CN, hemiparesis	Yes	G1	Yes	Lesionectomy	2	Transitory VI CN palsy
6	15	F	No	No	L basal nuclei	R hemiparesis	Yes	G4	No		6	No deficit
7	17	M	Yes	Yes	L parietal	Seizures	No	G3	Yes	Lesionectomy + hemosiderin removal	13	Seizure-free (Engel1b)
8	5	M	No	No	R frontal	Seizures	No	G3	Yes	Lesionectomy	5	Seizure-free (Engel1a)
9	4	M	No	No	L frontal	Seizures	No	G3	Yes	Lesionectomy	6	Seizure-free (Engel1a)
10	12	F	Yes	Yes	R Insula (multiple)	Headache	No	G4	Yes	Lesionectomy	2	Monoparesis
11	13	M	No	No	R frontal	Seizures	No	G3*	Yes	Lesionectomy + hemosiderin removal	1	Seizure-free (Engel1a)
12	1	M	No	Yes	R occipital	Seizures	No	G3	Yes	Mesial occipital pole resection	2	Seizure-free (Engel1c)
13	1.25	M	Yes	No	L temporal	Seizures, hemiparesis	Yes	G3	Yes	Left temporal pole resection	4	Seizure-free (Engel1b)
14	11	M	No	No	L thalamus	R hemiparesis	Yes	G4	No		2	No deficit
15	12	M	No	No	L thalamus	Headache	Yes	G4	Yes	Lesionectomy	5	Epilepsy
16	14	F	No	No	R frontal	Seizures	No	G3	Yes	Lesionectomy	2.33	Seizure-free (Engel1b)
17	16	M	No	No	L frontal	R hemiparesis	Yes	G4	Yes	Lesionectomy	1	No deficit
18	12	M	No	No	Medula oblongata	Coma	Yes	G1	Yes	Lesionectomy	5	No deficit
19	2	F	No	No	Pineal	Headache	No	G4	Yes	Lesionectomy	12	No deficit
20	7	M	No	No	Pons	CN, hemiparesis	Yes	G1	Yes	Lesionectomy	8	No deficit
21	14	F	No	No	R frontal	Seizures	No	G3*	Yes	Lesionectomy + hemosiderin removal	7	Seizure-free (Engel1a)
22	11	F	No	No	Cerebellum	Coma	Yes	G2	Yes	Lesionectomy	9	No deficit
23	4	M	Yes	No	R frontal	Seizures	No	G3	Yes	Lesionectomy	1	Seizure-free (Engel1a)
24	6	F	Yes	Yes	L temporal	Seizures	Yes	G3*	Yes	Lesionectomy + hemosiderin removal	3	Seizure-free (Engel1a)
25	6	F	No	No	R occipital	Headache	Yes	G4	Yes	Lesionectomy	0.5	No deficit
26	2	F	No	No	Pons	Headache	Yes	G1	No		1	No deficit
27	11	F	No	No	L temporal	Seizures	No	G3*	Yes	Lesionectomy + amigdalo-hippocampectomy	6.5	Seizure-free (Engel1a)
28	11	M	No	No	R temporal	Seizures	No	G3*	Yes	Electrocorticography + lesionectomy	4	Seizure-free (Engel1b)
29	5	M	No	No	L occipital	Seizures	Yes	G3	Yes	Lesionectomy	2	No deficit
30	8	M	No	No	L temporal	Seizures	No	G3	Yes	Lesionectomy	1	No deficit

Pt: patient; ML: multiple lesions; FH: family history; M: male; F: female; R: right; L: left; CN: cranial nerve deficit; IH: intracranial hypertension; G1: brain-steam; G2: cerebellum; G3: supratentorial associated; G4: supratentorial; G3* patients with epilepsy that required further neurophysiologic assessment.

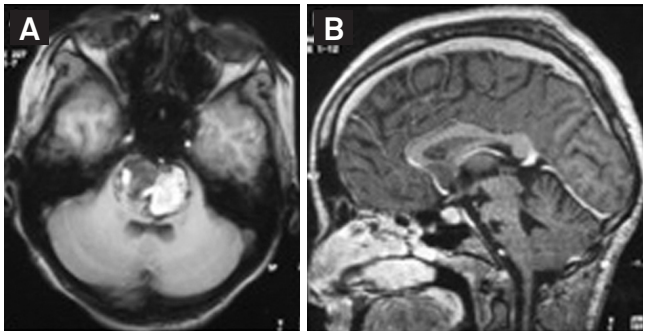


Fig 1. Pre (A) and postoperative (B) images of a brainstem cavernous malformation. This patient presented with tetraparesis and 11 points at the Glasgow Coma Scale. Three years after surgery she was asymptomatic. (A): axial T1-weighted magnetic resonance imaging revealed a typical cavernous malformation, characterized by heterogenic mass and hypointense halo signal around it; (B): sagittal T1-weighted gadolinium-enhanced magnetic resonance imaging revealed total removal of the cavernous angioma and decompression of brainstem (Patient 5).

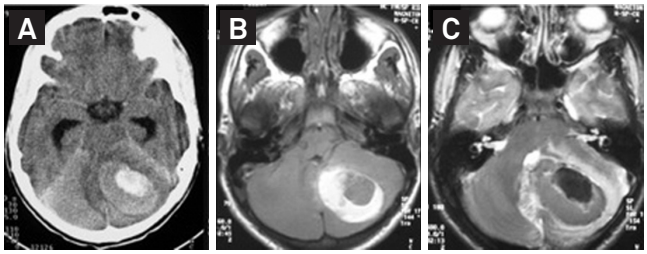


Fig 2. (A) brain computed tomography showed a hemorrhagic lesion on the left cerebellum hemisphere and incipient dilatation of ventricular temporal horns. Axial T1 (B) and T2-weighted (C) magnetic resonance imaging revealed heterogenic lesion with mass effect and fourth ventricle compression. Histopathology confirmed the diagnosis of a cavernous angioma (Patient 2).

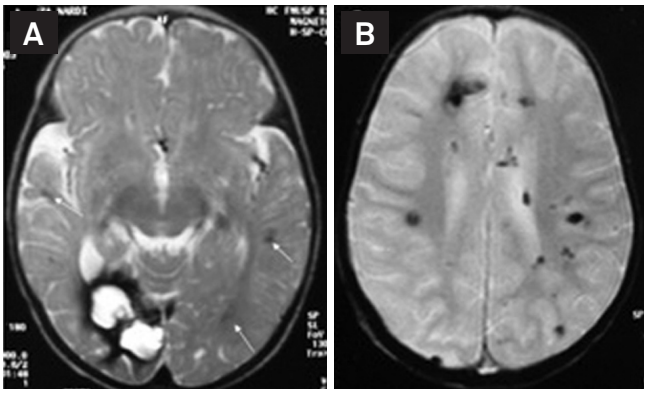


Fig 3. Axial T2-weighted magnetic resonance imaging (A) revealed a right occipital lesion with a hypointense halo around it, and other lesions can also be seen (arrows). The gradient-echo sequence (B) confirmed multiple cavernous malformations (Patient 12).

Among the 15 patients with seizures, eight are now seizure — and drug-free; in seven patients, however, antiepileptic medications were not discontinued, although these children also scored Engel's class 1.

Four patients that were not operated improved spontaneously and remained asymptomatic in the follow-up.

During the follow-up period, no new lesions or recurrence on MRI images were seen in all children except for a boy (case 15) with a CM in the thalamus that had to be re-operated. He presented seizures due to severe hyponatremy after surgery and remained with antiepileptic drugs afterwards. The remaining lesions were stable in children with multiple CMs.

DISCUSSION

CM is one of the congenital vascular malformations affecting the central nervous system (CNS) that may remain asymptomatic for a long time or may produce clinical manifestations[12]. The pathogenesis of CM remains unclear, and little is known regarding the molecular pathogenesis of the disease[3].

There are two types of CMs: sporadic and familial. The familial type is characterized by a multiplicity of lesions[13-15] and the possibility of *de novo* CM formation[16]. In this series, five patients (16.6%) had multiple lesions, three of them had also positive familial history. One subject with familial history (total of 4–13.3%) did not have multiple lesions. Among the literature review, incidence of familial history and multiple lesions both varies from 0.0 to 26.3%[2,3,6-10,17-19].

Table 2 summarizes previous studies on pediatric CM. Note that only ten series were according to the selection criteria and therefore included in this review[2,3,6-10,12,17,19].

In our series, the mean age at clinical manifestation was 8.7 years-old, which is similar to the literature. The ratio of male to female was 1.5:1. In addition to the present study, there seems to be a slightly predominance of boys in most analyzed studies (Table 2). Since these are patient-based studies, the definitive predominance is hard to define.

The majority of CMs come to medical attention after children experience any number of clinical symptoms, and up to 20% are discovered incidentally[19-21]. In pediatric patients, CM symptoms in the CNS are mainly present as hemorrhage, seizure, or focal neurological deficits[3]. Xia et al. also observed acute intracranial hemorrhage in 20% of children with CMs, according to CT and MRI studies. However, a higher proportion of hemorrhage was confirmed histopathologically in 52.5% (32/61) of operated children with CMs in their series[19]. Acute intracranial hemorrhage was observed in 53.3% (16/30) in our series.

In the current study, 56.6% of patients suffered from seizures, which is significantly higher than in earlier studies[2,6-8,10,17,19], although Acciarri et al.[9] reported 70% of seizures in a series of 42 children with CMs.

Presentation of symptoms has some correlation with lesion locations[19]. Patients with lesions located superficially in the cerebral hemispheres more frequently are present with seizures. On the other hand, patients with lesions

Table 2. Summary of literature review.

Referência	n	Age	Male:Female	FH	ML	Site					Main symptoms at diagnosis					Post-surgical results					
						Supra:infra-tentorial	Thalamus and Basal Gnaglia	Brainstem	Cerebellum	Spine	Seizures	Headache	Focal neurological deficits	none	Hemorrhage	Surgery	Follow-up	Improved	Unchanged	Worsened	Mortality
6	17	18 m to 16 y	1.12:1	4/17 (23.5%)	2/17 (11.7%)	15:2	2	2/17 (11.7%)	0	0	4 (23.5%)	NA	NA	0	12 (70.5%)	15 (88.2%)	NA	11/15 (73.3%)	1/15 (6.6%)	3/15 (20%)	0
14	19	7 m to 17 y (mean=9,1)	1.11:1	5/19 (26.3%)	5/19 (26.3%)	12:6	2	4/19 (21%)	2/19 (10.5%)	1	5/18 (27.7%)	7/18 (38.8%)	13/18 (72.2%)	0	12/18 (66.6%)	all	6 m to 9 y	17/19 (89.4%)	0	2/19 (10.5%)	0
18	18	10 m to 17 y	1:1.25	0	0	15:2	NA	0	2/18 (11.1%)	1	11/17 (64.7%)	1/17 (5.8%)	5/17 (29.4%)	0	NA	all	1 to 16 y	14/17 (82.3%)	3/17 (17.6%)	0	0
7	24	6 m to 15 y (mean=8 y)	NA	0	2/24 (8.3%)	20:4	NA	NA	NA	0	13 (54.1%)	NA (9 IH)	10 (41.6%)	0	19 (79.1%)	all	6 m to 14 y (mean=4 y)	23/24 (95.8%)	0	0	1/24 (4.1%)
8	36	9 m to 17 y (mean=8.6)	1.4:1	4/36 (11.1%)	NA	23:12	6	7/36 (19.4%)	5/36 (13.8%)	1	16/35 (45.7%)	1/35 (2.8%)	10/35 (28.5%)	5/36 (13.8%)	19/36 (52.7%)	all	NA	30/35 (85.7%)	3/35 (8.5%)	2/35 (5.7%)	0
3	33	1 to 20 y (mean=11.6 y)	1.2:1	0	0	27:6	3	5/33 (15.1%)	1/33 (3%)	0	19 (57.6%)	NA	NA	0	25 (75.7%)	25 and RS on 8	2 to 17 y (mean=5.8 y)	29/33 (87.9%)	4/33 (12.1%)	0	1/33 (3%)
9	42	10 m to 17 y	1:1	1/42 (2.3%)	5/42 (11.9%)	35:5	0	2/42 (4.7%)	3/42 (7.1%)	2	28/40 (70%)	11/40 (27.5%)	16/40 (40%)	0	17/40 (42.5%)	all	1 to 16 y	29/42 (69%)	10/42 (23.8%)	3/42 (7.14%)	0
19	66	15 m to 17.8 y (mean=11.6 y)	1.53:1	1/66 (1.5%)	7/66 (10.6%)	55:6 (both:4)	1	2/66 (3%)	4/66 (6%)	1	31/65 (47,7%)	30/65 (46,2%)	8/65 (12,3%)	2/65 (3,1%)	13/65 (20%)	62/66 (93.9%)	5 m to 9.3 y (mean=3.2 y)	43/46 (93.5%)	1/46 (2.2%)	2/46 (4.3%)	0
10	32	2 d to 17 y (mean=7.1)	1.13:1	3/32 (9.3%)	8/32 (25%)	24:08	1	6/32 (18.7%)	2/32 (6.2%)	0	13 (40.6%)	6 (18.8%)	7 (21.8%)	0	21 (65.6%)	28 (87.5%)	mean=4.43 y	27/28 (96.4%)	1/28 (3.5%)	0	0
2	79	4 m to 17 y (mean=9.7 y)	1.07:1	1/79 (1.2%)	3/79 (3.8%)	>74:NA	NA	NA	NA	0	41 (51%)	8 (10%)	14 (18%)	0	18 (23%)	all	1 m to 16 y (mean=3 y)	39/54 (72%)	12/54 (22.2%)	1/54 (1.8%)	0

n: number of patients; ML: multiple lesions; FH: family history; NA: not available; IH: intracranial hypertension; RS: radiosurgery; >: at least. Data were taken from the text and tables presented in the articles. Symptoms and post-surgical results excluded sometimes the spinal cord cavernomas and/or the patients without enough follow-up, correct amounts of patients are shown.

located in deep structures such as the brainstem generally present with focal neurological deficits. In this series, focal neurological deficits (36.6%, 11/30) happened mostly in deep-seated and infratentorial lesions. Only two cases with focal neurological deficits did not result from deep-seated lesions, an acute hemorrhage of a temporal (case 13) and a frontal CM (case 17).

CMs have been observed throughout the CNS, and their locations in children are comparable to those in adults[8,19,22]. Supratentorial location accounts for about 80.0%, while the other 20.0% are located in the posterior cerebral fossa[3,8,19,22]. The proportion of supratentorial CMs in the descending order is frontal, temporal, occipital, and parietal lobe. A deep location in the basal ganglia, hypothalamus, or ventricular system is rare[8,19,22]. In this series, regarding only the symptomatic lesions, 23 patients (76.6%) presented supratentorial CM and seven (23.3%) infratentorial.

Management

Presence or absence of hemorrhage is important to decide how to manage the patients. The risk of bleeding is higher after the first event, 22.9% against 0.25–3.8%

without prior hemorrhage[4]. However, without a documented episode of hemorrhage, treatment should be based on the precise location of the cavernoma, patient's age, symptoms, and outcome expectation[23,24].

Additional data of natural history also favor surgical treatment. Seizures seem to happen precociously when CMs are located in the frontal or temporal lobes[25], and apparently their risk of development varies from 1.5 to 4.8% per year[4,21]. Focal deficits and intracranial hypertension may be caused not only by acute macro-hemorrhage, but also due to progressive growing secondary to recurrent micro-hemorrhage[24].

Figs 4 and 5 summarize management strategies for each following group.

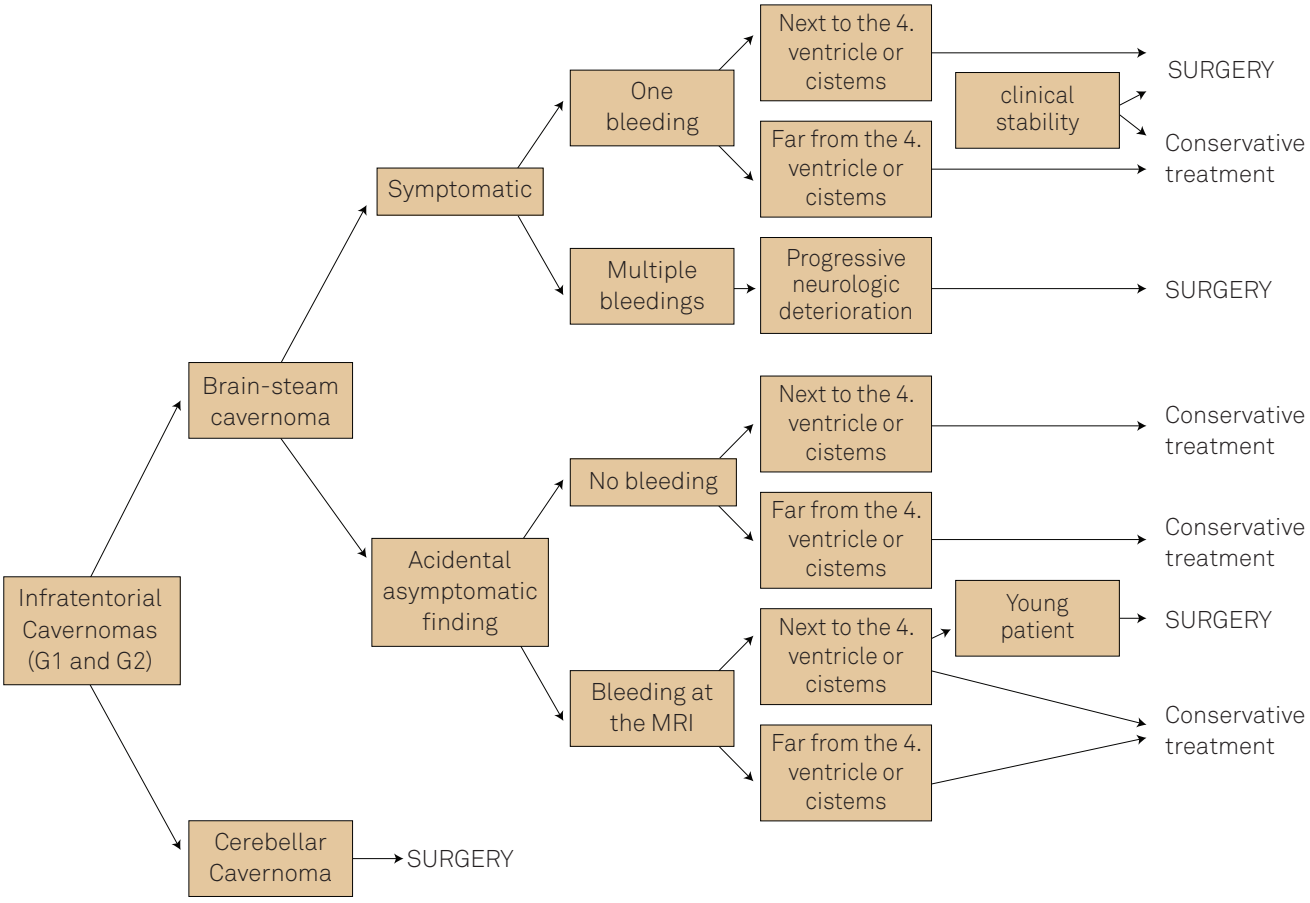
Brain steam cavernomas (G1)

Brainstem CMs constitute 0 to 21% of all cavernomas in different pediatric series[3,6,8-10,17-19]. Clinical presentation is generally acute and severe due to peculiar localization, despite frequent small amount of hemorrhage. Neurological improvement often happens a few days later. This apparently benign course is due to slow flow and pressure inside the CMs, consequently hemorrhage usually displaces the neural tissue instead of its destruction[7]. Two of our patients were not submitted to surgery and improved totally spontaneously.

Surgical resection is indicated for those brainstem lesions that appear in the pial surface[23]. Asymptomatic patients or with good neurological recover should be observed, mainly if the CMs are located deeply in the brain steam[4,17]. Some authors believe that the risk of not operating on a patient that had already suffered hemorrhage is very high, especially if he/she is young with high life expectancy, independent of the lesions depth, because of the possible catastrophic evaluation if it rebleeds or grows[3,26]. Neurosurgical team experience and skills, as well as neurophysiologic monitoring, should be considered to treat such patients[19,23].

Cerebellar cavernomas (G2)

Cerebellar location is less frequent, and clinical presentation is generally due to great hemorrhage inside the cerebellar parenchyma that may cause mass effect and occlude



G1: brain-steam; G2: cerebellum; MRI: magnetic resonance imaging.

Fig 4. Therapeutic guideline for G1 and G2 (modified from Samii et al.[23]).

the fourth ventricle resulting in cerebellar dysfunction and hydrocephalus[27].

Patients with cerebellar cavernoma (G2) should be treated with surgery even in the absence of hemorrhage. Surgery is usually safe and successful while a first or recurrent hemorrhage in the posterior fossa represents great threat. Exception is made for multiple asymptomatic cerebellar CMs[4]. Cerebellar cavernomas represent 0.0 to 13.8% in earlier studies[3,6,8-10,17-19], in our series there were two patients in this group (6.6%).

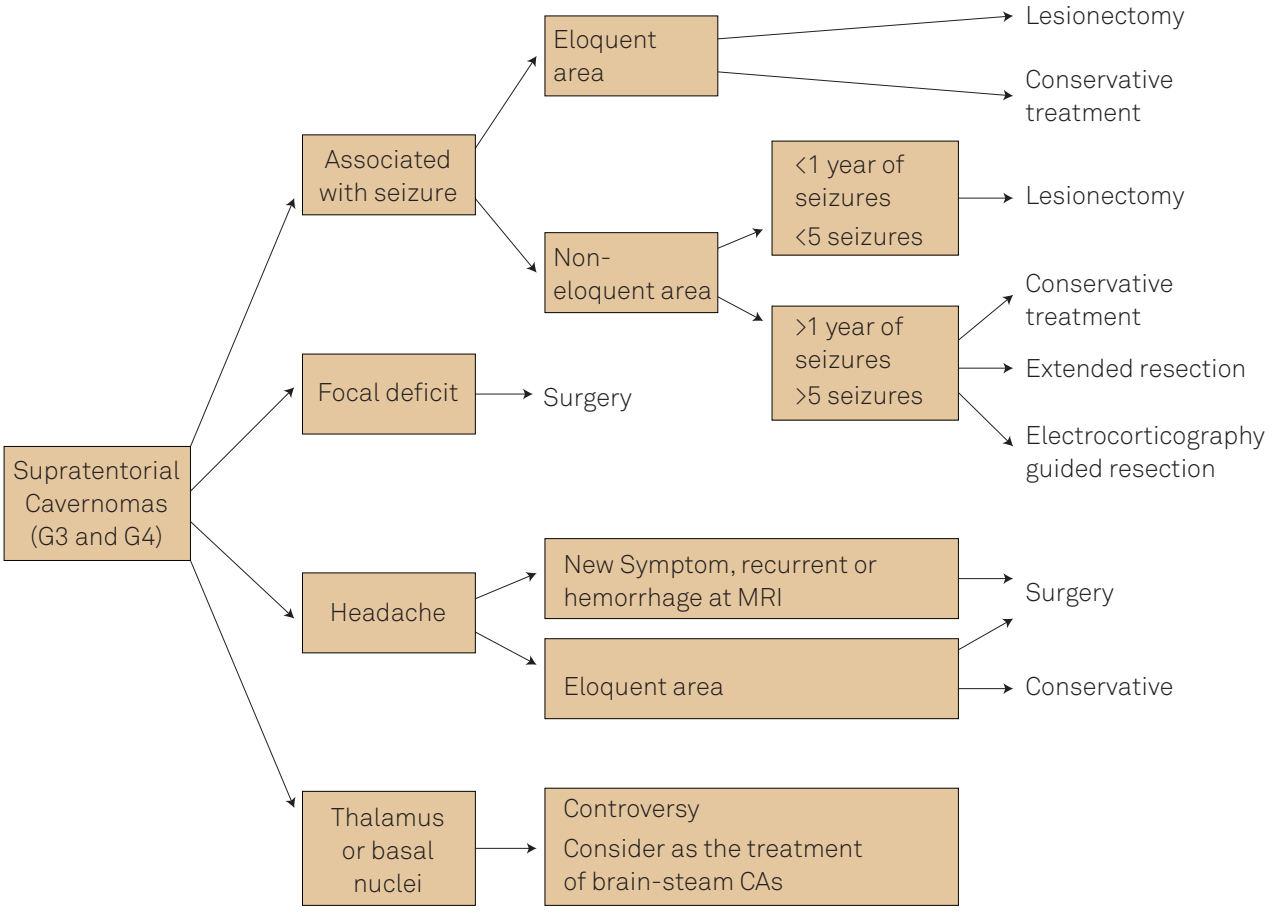
Supratentorial cavernomas associated with seizure (G3)

Surgical treatment should be indicated to the majority of patients that suffered hemorrhage and to those which surgical risk was considered low[4,9].

Lesionectomy alone showed favorable results[28]. In a series of patients submitted to lesionectomy of supratentorial CMs, a strict relationship was found between the duration of seizure history before the surgical treatment and the cure of epilepsy with the surgery. Cohen et al.[28] showed that all patients that presented one preoperative seizure or less than two months of history did not have any seizures after surgery; 75.0 to 80.0% of patients with two to five preoperative seizures or 2 to 12 months of history also had not any seizure after surgery. In addition, 50% of patients with more than five seizures or more than 12 months of history were postoperatively seizure free[28]. Although resection of the hemosiderin stained tissue that surrounds the lesion was not proved to be of better effectiveness[4,8-10,29], it was adopted by several studies when CMs are placed in noncritical areas[2,3,9], including the present one. Eleven patients of this study underwent simple lesionectomy.

Longer seizure history and increased number of preoperative seizures are consistent with more difficulty in treating epilepsy with simple lesionectomy surgery[28]. We believe these patients should have a complete epilepsy evaluation to a possible extended surgery guided by electrocorticography. In fact, five patients with refractory seizures were further evaluated, and two of them needed an extended resection.

This group represented 53.3% of patients (16/30) and surgical techniques chosen for each case are showed in Table 1.



G3: supratentorial associated; G4: supratentorial; CAs: cavernous angiomas; MRI: magnetic resonance imaging.

Fig 5. Therapeutic guideline for G3 and G4.

Supratentorial cavernomas not associated with seizure (G4)

This group represented 23.3% of patients (7/30). The management of thalamic and basal ganglia CMs is controversial[4]. In selected cases, surgery is considered when the CM is next to the ventricle surface or in the insular lobe through to the Sylvian fissure approach[10]. Indeed, among three patients, two were conservatively treated with a remarkable neurological outcome.

Recurrent headache seems to be attributed to the CM. Surgery is indicated to prevent future neurologic deficit, except for patients with unacceptable risk due to critical lesion location[4,9]. In this series, one patient (case 19) with a pineal lesion had insidious onset of headache and somnolence. Another patient (case 10) with multiple lesions had history of recurrent headache and behavior abnormality, the insular lesion was operated through a pterional approach and a monoparesis persisted in the follow-up.

If a focal neurologic deficit is present, there is usually an associated hematoma, and the surgical removal should be performed in order to avoid further deficit and to help clinical recovery. The management of CM in eloquent brain areas (motor, sensitive, speech, or visual) should be tailored for each case[4].

In conclusion, we found a favorable outcome for surgically treated children with symptomatic CMs, and we suggest that resection should be the golden-standard therapy for patients with lesions that do not cause an excessive surgical risk, such as localization in a highly eloquent area.

Natural history is particularly important in children and should be individualized, considering besides bleeding also CM location, symptoms, surgical treatability, and life expectancy. Surgical removal of CMs in eloquent areas, such as brainstem and thalamus/basal nuclei, can prevent neurological deterioration secondary to re-bleeding or more seldom lesion growth, but these are still challenging and conservative treatment is still an option, particularly in those cases with considerable clinical improvement. Cerebellar cavernoma should be generally treated with surgery. Recent record of seizures is associated with high rate of success after lesionectomy. Extended surgery associated with CM lesionectomy for long-term epilepsy or high frequency of seizures should be considered after proper investigation and failure of drug therapy.

Conservative treatment may be considered for asymptomatic patients, multiple CMs, critical lesion location (brainstem, thalamus, and basal nuclei) and in cases of small lesions that are not associated with hemorrhage.

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