Cerebellar cavernous malformation in pediatric patients: defining clinical, neuroimaging, and therapeutic characteristics

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OBJECT Cerebellar cavernous malformations (CCMs) have not been specifically described in the pediatric age group. Authors of this study, after considering the published literature, describe the characteristic clinical, radiological, and surgical features of CCM in children.

METHODS Patients younger than 18 years of age who were known to have CCM and had undergone surgery between 1992 and 2014 at the authors' institution were reviewed. Pediatric CCM cases reported in the literature (case reports and cases included in series on CMs in the pediatric age group) were also analyzed for specific features of this entity.

RESULTS Four male patients and 1 female patient (2.5–14 years of age) with CCM presented acutely with severe headache followed by cerebellar dysfunction. In all patients, neuroimaging (cranial CT and MRI) demonstrated hemorrhagic cerebellar lesions with heterogeneous T1 and T2 signal intensities and hyperintense blooming on susceptibility-weighted imaging. The lesions reached large sizes exhibiting spherical, cystic, and often "pseudotumoral" morphology. In 3 patients, developmental venous anomalies (DVAs) were found. In 4 of the 5 patients, the CCMs and hematomas were totally removed. All patients had a clinically excellent functional outcome without surgical complication and with complete resolution of their presenting symptoms.

CONCLUSIONS Cerebellar CMs occur in all pediatric age groups and display characteristic clinical and imaging features. In children, CCMs reach large sizes and can result in massive hemorrhage, often leading to a possible diagnosis of hemorrhage into a tumor. An associated DVA is quite common. Surgery is a safe and efficient treatment option with excellent outcomes in patients.

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KEY WORDS cavernous malformation; cavernoma; cerebellum; pediatric age group; surgical treatment; neuroimaging; outcome; vascular disorders

CAVERNOUS malformations (CMs), also known as "cavernous angiomas," "cavernous hemangiomas," or "cavernomas," are common vascular abnormalities in the brain, with an estimated prevalence of 0.4% to 0.8% in the general population.^{4,12,21,28,33,34} In the pediatric age group, their prevalence is not known because of small numbers of patients in age-mixed reports.⁴ In pediatric series, CMs represent 1.7%–18% of all vascular growths.^{23,25}

In children, these low-flow vascular malformations are

one of the main causes of cerebral hemorrhage besides arteriovenous malformations.^{13,24,27,29,35} Cavernous malformations occurring in children differ from those in adults as they commonly have atypical neuroimaging at diagnosis, show more rapid rates of growth, and have larger dimensions and a greater likelihood of hemorrhage at presentation.^{2,14,26,27} Cavernous malformations are located throughout the CNS in the pediatric age group.^{24,27} Most are supratentorial in location,^{2,23,24,26,27} followed by those

ABBREVIATIONS BCCH = British Columbia Children's Hospital; CCM = cerebellar cavernous malformation; CM = cavernous malformation; CN = cranial nerve; CTA = CT angiography; DVA = developmental venous anomaly; MRA = MR angiography; SWI = susceptibility-weighted imaging.

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in the brainstem and less frequently in the cerebellum.^{10,31} de Oliveira et al.¹⁰ reported that CCMs in adults exhibit specific clinical characteristics. Except for a few case reports, however, CCMs in pediatric patients have not been described.^{11,16,19} In the present study, we describe the clinical and radiological presentation and surgical treatment of CCMs in patients presenting at an age less than 18 years.

Methods

Ethics approval for this study was obtained from the Clinical Research Ethics Board of British Columbia Children's Hospital (BCCH).

Patient Population and Clinical Data

The prospectively maintained pediatric neurosurgery electronic database of BCCH was queried for patients younger than 18 years of age who had been treated for cranial CMs from 1992, when MRI became available at BCCH, to 2014. For the purposes of our study, we then selected patients who had undergone surgery for CCMs and had a histologically confirmed diagnosis. Their charts were retrospectively reviewed for clinical presentation, imaging and surgical features, histopathology, and outcome. All patients had undergone cranial CT and MRI before surgery and MRI after surgery.

Literature Review

PubMed was searched for the following terms: "cavernoma" or "cavernous angioma" and "pediatric" or "children" and "cerebellar" or "cerebellum." Only articles in English (and 1 in French) were considered. We excluded studies in which pediatric and adult age ranges were mixed and in which cerebellar location was mentioned without providing any patient details, clinical symptoms, or imaging features.^{4,6}

Results

From 1992 to 2014, 27 patients under the age of 18 years were managed by the neurosurgical service at BCCH for intracranial CMs; 23 were surgically treated and 4 were managed conservatively. Five of these patients had CCMs and were surgically treated. Diagnosis was pathologically confirmed in all 5 patients.

Patient Age, Sex, and Clinical Presentation

There were 4 male patients and 1 female patient whose ages ranged from 2.5 to 14 years, with a mean age of 7 ± 4.6 years at presentation (Table 1). All patients were asymptomatic prior to presenting with acute or rapidly progressing headaches.

Case 1

This 11-year-old boy presented with a 2-week history of the sudden onset of occipital headaches and vomiting. He had intermittent fever and was treated for gastritis in an outpatient clinic. When the symptoms did not resolve and he developed gait ataxia and double vision, he was transferred to BCCH. On arrival, he was cachectic as a result of vomiting, was complaining of severe occipital headaches,

and had a left sixth cranial nerve (CN) palsy, ataxia, and left-sided dysmetria.

Case 2

This 5-year-old boy presented with a 14-day history of increasing headaches. During transport in our hospital, he experienced one syncopal episode. He did not have papilledema, dysmetria, or ataxia. His CN examination was normal.

Case 3

This 14-year-old girl had a 4-day history of occipital headaches. Her neurological examination was normal.

Case 4

This 3-year-old boy was admitted for increasing headaches and unsteadiness of gait, which started a few days before admission. He was neurologically intact except for gait ataxia. There was a history of familial CM, as his father was known to have multiple intracranial CMs.

Case 5

This 2.5-year-old girl presented with the acute onset of severe headaches and a normal neurological examination.

Neuroimaging Lesion Characteristics

Because of acute presentations, all patients underwent initial cranial CT, which demonstrated mixed-intensity, round or spherical hemorrhagic cerebellar lesions (Figs. 1A, 2A, 3A, 4A, and 5A). In Case 2, calcification was seen on the inferior portions of the lesion (Fig. 2B). Hemorrhage into a tumor was the commonest referring diagnosis.

In all cases, CT findings prompted MRI sequences, including T1-weighted, T2-weighted, 3D T1-weighted post-Gd injection, susceptibility-weighted, and diffusion-weighted (Table 1). In Cases 1, 2, and 4, MR angiography (MRA) was also performed with the time-of-flight technique. Preoperative MRI exhibited spheroid, cystic mass lesions with considerable variations in size. All images showed compression of adjacent crucial anatomical structures, especially in Case 1 (Fig. 1B–F). All lesions exhibited heterogeneous T1 and T2 signal intensities and characteristic hypointense blooming on susceptibility-weighted imaging (SWI) sequences with surrounding edema (Figs. 1D, 2E and F, 3C, 4D and E, and 5B). Case 3 differed morphologically, with a compact area (Fig. 3C and E) and diffuse multifocal small lesions scattered through adjacent cerebellar cortex (Fig. 3D) creating a more diffuse blooming pattern on SWI (Fig. 3F).

With the benefit of MRI, cavernous malformation was considered the most likely preoperative diagnosis, followed by hemorrhage into a tumor. The presence of developmental venous anomalies (DVAs; Cases 1, 2, and 5; Figs. 1F, 2H, and 5E and F), multifocal supra- and infratentorial lesions, and/or a family history of CM (Case 4; Fig. 4F) supported this diagnosis.

Formal vascular imaging was performed preoperatively (CT angiography [CTA] in Case 3, and MRA in Cases 1, 2, and 4). These studies excluded abnormal arterial feeders but did not reveal the DVA in Cases 1 and 2.

TABLE 1. Patients with surgically treated CCM at BCCH, 2008–2014

Case No.	Age (yrs)	Sex	Family Hx	Clinical Symptoms	MRI Findings			Duration of Illness		FU	Clinical Outcome
					Morphology*	Signal Intensity	DVA				
1	11	M	No	Acute onset of HAs, nausea, vomiting; 6th CN palsy & ataxia approximately 1 day before admission	Hemorrhagic, fluid level exhibiting cystic mass centered in lt cerebellar hemisphere; whole lesion: 4.0 × 4.1 × 4.7 cm; anteromedial nodule: 2.4 × 2.4 × 3.2 cm; beginning hydrocephalus	Mixed T1 & T2; predominant ↑T1 ↑T2 ↓T2 rim; blooming on SWI	Yes	14–16 days	4 wks after op no deficit (10 mos FU)	Excellent	
2	5	M	No	Progressive HAs; 1 syncopal episode on day of admission	Oval mass in lt cerebellar hemisphere w/ calcification (2.8 × 2.4 × 1.9 cm); associated superior (3.3 × 2.7 × 1.7 cm) & inferior (1.3 × 1.3 × 0.8 cm) hematomas; no hydrocephalus	Reticulated w/ mixed T1 & T2; ↓T2 rim; blooming in SWI	Yes	14 days	No deficit (1 yr FU)	Excellent	
3	14	F	No	Progressive occipital HAs	Large hemorrhagic lesion (4.76 × 3.34 × 2.9 cm) in rt cerebellar hemisphere; multiple foci in adjacent superior cerebellar hemisphere; mild dilation of ventricles	Mixed T1 & T2; ↑T1 ↑T2 hematoma ↓T2 reticular part SWI: compact & diffuse blooming	No	4 days	Growing remnant on MRI (6 yrs FU)	Good; no clinical complications	
4	3	M	Yes	Increasingly severe HAs & unsteadiness, ataxia	Cystic hemorrhagic lesion in lt cerebellar hemisphere w/ almost bell clapper–like hypodense encapsulation; lesion: 1.8 × 1.7 × 1.8 cm; hematoma: 3.4 × 3.5 × 3.15 cm; multiple supratentorial & pontine lesions; no hydrocephalus	Mixed T1 & T2 SWI: lesion blooming & multifocal supratentorial hypointense blooming	No	“Few days”	Ataxia resolved completely after 3 mos; clinically well (2 yrs FU); no seizures	Excellent	
5	2.5	M	No	Acute onset of severe HAs	Cystic rt-sided cerebellar mass (3.4 × 4.0 × 2.8 cm); no hydrocephalus	Mixed T1 & T2 SWI: lesion blooming	Yes	4–5 days	No deficits (2 yrs + 4 mos FU)	Excellent	

HAs = headaches; Hx = history; FU = follow-up.

* Lesion measurements: transverse × anterior/posterior × cranio/caudal.

FIG. 1. Case 1. **A:** Coronal CT scan exhibiting a hemorrhagic lesion centered within the left cerebellar hemisphere and vermis with surrounding edema. There was beginning hydrocephalus with mildly enlarged ventricles and transependymal extravasation. **B:** Sagittal T1-weighted MRI sequence showing a cystic lesion with heterogeneous, predominantly high T1 signal extending into the left cerebellar tonsil, which appeared caudally displaced. **C and D:** Sagittal and axial T2-weighted MRI sequences showing the lesion with heterogeneous signal with surrounding low T2 signal rim. There was a well-defined large nodule located anteromedially ($2.4 \times 2.4 \times 3.2$ cm) containing a fluid level of different densities. Furthermore, there was edema of adjacent cerebellar parenchyma including the middle cerebellar peduncle and dorsal pontine tegmentum. The mass resulted in distortion of the cerebellar peduncles and medulla at the foramen magnum level. **E:** Susceptibility-weighted image with signal drop in the lesion lateral to the anteromedial cyst. **F:** Post-Gd-enhanced sequence did not reveal abnormal enhancing tissue. One vertically oriented vessel exhibited features suspicious for an associated developmental venous anomaly (DVA; arrow), draining into the left inferior petrosal sinus.

All patients underwent whole spinal cord MRI before (Cases 1, 2, 4, and 5) or after (Case 3) surgery. No additional CMs were found.

Surgical Findings

All patients underwent suboccipital craniotomy and excision of the CM and its associated hematoma. In Cases 2, 3, and 5, the lesions seen at surgery exhibited typical berry-like appearances; in Case 4, the lesion was firm and calcified. In Case 2 a venous structure medial and anterior to the lesion, corresponding to MRI findings, was recognized and kept intact. In Case 1 the potential venous structure was not identifiable during resection and appeared to be removed on postoperative MRI. As confirmed with postsurgery MRI, complete removal of the lesion was achieved in all but 1 patient (Case 3). Although the lesion and hematoma in this patient were resected with clean resection margins, the superiorly located diffuse components remained.

Histopathological Lesion Features

In all cases, the suspected diagnosis of CM was confirmed pathologically. The microscopic specimens showed

numerous abnormal blood vessels ranging in size from small to large and were often closely packed together. Focal calcification was found in Cases 2, 4, and 5, and occasional thrombosed vessels were found in Cases 2, 3, and 5. No arterial components were seen with Movat's pentachrome stain (Fig. 6). The cerebellar parenchyma, when included in the resected tissue, exhibited acute to subacute remote damage, including acute neuronal necrosis (Case 5), gliosis (Cases 3 and 4), and atrophy. All lesions were associated with an inflammatory reaction, most prominent between the vessels and in the gliotic brain tissue. There were significant numbers of CD68-positive macrophages and occasional perivascular lymphocytes (Cases 1–3).

Surgical Outcome

In all patients, initial symptoms and clinical manifestations resolved shortly after lesion and clot removal. In Cases 3 and 4, clinically silent imaging changes were noted during follow-up. In Case 3, residual CCM was visualized on postsurgical MRI (Fig. 3). This residual lesion evolved over time with increasing T2 hypointensity (Fig. 3K and L) and hemosiderin blooming on SWI sequences. In addition, a new area of T1 hyperintensity (1 cm in diameter) in

FIG. 2. Case 2. Coronal CT scans obtained on admission, exhibiting a hyperintense, roundish hemorrhaging lesion with surrounding edema (A) and inferior calcifications (B) located in the left cerebellar hemisphere. There was no associated hydrocephalus. Sagittal T1-weighted MRI sequences demonstrating a mixed-density lesion with a hyperintense nodule (C) and superior cystic component (D). Axial T2-weighted MRI sequence (E) showing the reticulated lesion (inset picks up more basal proportions) with mixed, predominately low T2 density, surrounded by a hyperintense edematous rim and associated anteromedial high T2 signal nodule. Another large cystic component was located superior to the lesion with isointense T2 signal intensity (F). On an SWI sequence (G), the lesion was associated with marked blooming artifacts. With contrast enhancement, a vascular structure exhibiting typical features of an associated DVA (arrow) became apparent medially and anteriorly to the lesion (H), presumably draining into a venous sinus in the region of the left tentorium.

the right cerebellar hemisphere (Fig. 3H) appeared following surgery. The patient remained asymptomatic and her scan was unchanged at the 4-year follow-up. The patient in Case 4, who had had numerous bilateral supratentorial and pontine foci of SWI blooming on initial MRI, showed interval enlargement of several of these supratentorial foci and development of one single new lesion within the right temporal lobe. These lesions remained clinically silent at the last follow-up.

Literature Review

We found 3 case reports and 15 case series providing relevant information on CCM in the pediatric age group.^{2,5,7–11,13,14,16,19,20,23,24,27,32,35,36,39} These cases are summarized in Table 2.

Discussion

Incidence and Sex Distribution of CCM in Children

The occurrence of CCM in children has been reported to range from 3% to 13.5% (mean 8.7%) among all cranial CMs.^{13,20,24,27,40} In our experience, CCM accounted for 18% of CMs. In our surgical series, the occurrence was 21.7% of cranial CMs.

Cerebellar CMs occur in patients of all ages, with children 11 years and older appearing to be more frequently

affected. This contrasts with the bimodal age distribution of CMs in other sites, which shows an infant/early childhood peak at 3 years of age and younger and an older child/teenager peak between 13 and 16 years.¹⁷

Clinical Presentation

All CCM cases in our series and those cases reported in the literature presented with clinical signs related to acute cerebellar hemorrhage in previously asymptomatic patients. Headaches often localized in the occipital region had an acute onset and progressive course. Other clinical findings were related to cerebellar dysfunction. One patient in our series had sixth CN palsy. Epstein et al. described a case presenting with ear and facial pain.¹⁶ Two CCM cases presented with “coma,”^{5,11} and we are aware of 1 case presenting with a lethal hemorrhage (C. Dunham, personal communication, 2014).

Cerebellar CMs differ from supratentorial CMs that present most often with seizures^{2,5,23,24,27,39} and from brain-stem CMs, which become obvious with focal neurological deficits.^{31,39}

The time from symptom onset to treatment in our series was 3–16 days. Headaches and vomiting were often treated symptomatically until the patient developed cerebellar dysfunction or CN palsy, which prompted neuroimaging. Patients exhibiting short clinical courses have also

FIG. 3. Case 3. Coronal CT (A) demonstrating a right cerebellar hemisphere hemorrhagic lesion of mixed signal intensity with a hyperintense roundish component and surrounding edema. Sagittal T1-weighted MRI sequence (B) with mixed, partially hyperintense signal lesion. Axial T2-weighted MRI sequence (C) showing heterogeneous signal intensities of cystic and reticulated lesion components with surrounding edematous changes. Superiorly, the lesion proportions appeared to be more diffusely scattered throughout the adjacent parenchyma (D). Susceptibility-weighted images (E and F) picking up the cystic lesion area corresponding to that in panel C and a more diffuse blooming pattern scattered through the adjacent superior right cerebellar hemisphere (corresponding to that in panel D). Sagittal T1-weighted images (G and H) obtained directly after surgery, showing residual lesion (arrows) of mixed T1 intensity with one hyperintense nodule located under the tentorium. Postsurgical axial T2-weighted MRI sequences (I and J) demonstrating multiple foci of residual mixed T2 signal exhibiting lesion components extending from below the tentorium to its inferior margin. Arrow in panel J depicts residual lesion corresponding to its T1-weighted appearance in panels G, (K), and L. These foci showed dynamic changes over time with an overall increase of T2 hypointensity of hemosiderin deposition suggestive of interval hemorrhage. Moreover, the lesion changed its T2 signaling behavior over time as the posterior infratentorial nodule (J, arrow) disappeared over time, while 2 years later a new hyperintense foci became apparent more anteriorly (L, arrow).

been reported by Acciarri et al.² In contrast, the patient described by Epstein et al. was first treated conservatively but underwent an operation after a second hemorrhage of the CCM after 4 years.¹⁶

Imaging Features of Pediatric CCM

Computed tomography scanning shows cerebellar hemorrhage with significant mass effect.^{11,16} On MRI, these lesions exhibited a spherical or cystic appearance with mixed signal on T1- and T2-weighted sequences, characteristic hypointense T2 lesion rim, and blooming on SWI sequenc-

es. This appearance was in keeping with several episodes of bleeding. These hemorrhagic lesions reached significant sizes (Table 1) with distortion of the brainstem or other neighboring structures and were accompanied by perilesional edema.^{27,37} We did not routinely perform CTA preoperatively. Three patients had preoperative MRA. These studies did not add material information to our clinical decision making. The DVAs, when present, were detected on postcontrast T1-weighted sequences in our patients.

Our imaging findings were consistent with those described by Mottolese et al.²⁷ In their study, the most com-

FIG. 4. Case 4. A coronal CT (**A**) showing a large, almost isointense, partially hyperintense spherical hemorrhagic lesion in the left cerebellar hemisphere. Sagittal (**B**) and axial (**C**) T1-weighted MR images showing the different cystic lesion components with mixed signal intensity in keeping with hemorrhages of different ages and acute peripheral bleeding. The lesion resulted in mass effect with compression of the fourth ventricle. Axial T2-weighted MRI sequence (**D**) demonstrating cystic lesion portions with mixed signal intensities and one exhibiting an almost bell clapper-like hypodense encapsulation. Corresponding SWI sequence (**E**) with blooming of the lesion. Representative MR image (**F**) showing multiple supratentorial lesions with characteristic blooming dispersed throughout both cerebral hemispheres due to underlying familial CM.

mon MRI presentation in symptomatic patients was a large, spherical, well-delineated acute or subacute hematoma with sharp margins that contained or were associated with structures of different density signal.

The main differential diagnosis was hemorrhage into a preexisting cerebellar tumor. Because cerebellar tumors are much more common in children than CCMs, the diagnosis of tumor is often the first consideration. The spherical and “cystic” appearance may mimic that of other cystic tumors, for example, pilocytic astrocytomas.²⁵ However, features such as an associated DVA, multilocular lesions on SWI, and several hemorrhage episodes (bleeding of different ages) favor CM as the preoperative diagnosis.

Cerebellar CM—Associated DVA

The co-occurrence of DVA and CM is recognized in the literature.^{30,40} In the pediatric age group, however, the CM and DVA association has been reported less often.²⁷ This contrasts with our observations.

It has been suggested that CMs associated with a DVA have a high propensity to hemorrhage.³⁰ Wurm et al. reported that 93.3% of those harboring an associated DVA had symptomatic hemorrhage.³⁸ Abdulrauf et al. compared

CMs with and without an associated venous anomaly and showed that CMs associated with DVAs occur predominantly in the posterior fossa (with a slightly higher proportion of cerebellar [$n = 6$] compared with brainstem [$n = 4$] CMs) with an associated significant higher bleeding and rebleeding risk compared with CMs without associated DVAs.¹ As such, CCMs in the pediatric age group may be more prone to bleeding than CMs elsewhere in the nervous system.

The clinical importance of a DVA remains unclear. Some authors advocate leaving the DVA intact to prevent hemorrhagic infarction.³⁰ In contrast, if the DVA is a promoting factor in the development of cerebral CM, resection of associated DVA components may prevent regrowth of a partially excised CM.¹⁵ In Case 1 the DVA was removed during CM extirpation without clinical sequelae.

Histology of Cerebellar CMs

Our cases exhibited typical pathological characteristics, in accordance with those in the published literature on CMs in cerebellar and other locations.^{11,27} The lesions displayed abnormally dilated vascular spaces of various sizes and degenerative changes with irregular fibrous hyaline

FIG. 5. Case 5. **A:** Coronal CT obtained on admission, showing a large left cerebellar roundish hemorrhagic lesion with mixed but mostly hyperintense density. **B:** Axial T2-weighted MR image with mixed density and perilesional edema. There was mass effect on the fourth ventricle and brainstem. **C:** Corresponding SWI sequence. **D:** Sagittal T1-weighted MRI sequence with hypo- and hyperintense signal intensities. **E and F:** Sagittal and axial post-Gd MR images with visible potential DVA but no further pathological enhancement.

thickening, old hemorrhages, and thrombosis. The vessels were packed closely together typically with no normal neural tissue in between. The cerebellar cortex close to the vascular malformation exhibited signs of atrophy and gliosis, also a common finding in the literature on CCM. Additionally, in all of our cases, there was an accompanying inflammatory reaction.

Treatment Options and Outcome

The overall annual risk of hemorrhage of incidentally

diagnosed CMs was reported to be low at 0.4%–2% per year; however, the risk of a second hemorrhage in the next year was estimated to be 4%–5%.^{3,22,33} Cavernous malformations in the pediatric age group were reported to be more prone to bleeding with a more dramatic clinical course than those in adults.^{18,20,26} Surgical removal of symptomatic CCM is considered the treatment of choice with a good overall outcome.^{2,5,9–11,16,19,24} All patients in our series also underwent excision of the CCM and associated hematomas with an excellent overall clinical outcome.

FIG. 6. Case 1. **A:** Numerous closely packed abnormal venous structures. **B and C:** Portions of an abnormally long and thick vessel wall (arrows) lacking black elastic fibers. This abnormal venous structure is directly opposed to cerebellar parenchyma (behind the arrows) and is suspected to represent the presumed DVA on neuroimaging. H & E (A and B), Movat's pentachrome stain (C), original magnification $\times 100$ (A–C). Figure is available in color online only.

TABLE 2. Literature summary of CCM cases in the pediatric population

Authors & Year	No. of Cases	Age	Sex	Symptoms	CT/MRI	Size of Lesion (cm)	Op	Outcome
de Tribolet et al., 1982*	1/1	12 yrs	F	Sudden occipital HAs, cerebellar dysfunction, acute IH, coma	"Hematoma"	4	Yes	Good; nystagmus & homonymous hemianopia remained 8 mos after op
Simard et al., 1986	1/12 (8.3%)	6 yrs	F	"Signs and symptoms of mass effect"	—	—	Yes	—
Rigamonti et al., 1988	3/24 (12.5%)	—	—	—	—	—	—	—
Epstein et al., 1990*	1/1	12 yrs	F	Nausea, vomiting, vertigo, occipital HAs, gait ataxia, dysmetria, rt ear pain for 12 days; followed for 4 yrs (improved & stable); †acute onset of occipital HAs radiating into her face	Hyperdense/Type I; Type II	—	No; † yes	Good
Ciricillo et al., 1994	5/37 (13.5%)	—	—	Cerebellar signs in 3 patients	—	—	—	—
Scott et al., 1992	2/19 (10.5%)	11 yrs; 8 yrs	—	—	—	—	Yes; yes	—
Giulioni et al., 1994	2/18 (11.1%)	13 yrs; 15 yrs	M; F	Cerebellar dysfunction; cerebellar dysfunction	—	—	Yes; yes	Unchanged; unchanged
Fujii et al., 1996*	1/1	9 mos	M	Lethargy, focal neurological deficit due to additional cavernoma in the pons, multiple cavernomas	Hyperintense on T1 & T2	—	Yes	Cerebellar symptoms good
Di Rocco et al., 1996, 1997	2/22 (9%)	—	—	—	—	—	Yes	—
Lee et al., 2008	1/33 (3%)	—	—	—	—	—	—	—
Mottollese et al., 2001	4/36 (11%)	—	—	—	—	—	—	—
Lena et al., 2007	5/57 (8.77%)	—	—	Cerebellar dysfunction	—	—	—	Good
de Oliveira et al., 2006	1†	14 yrs	M	Sudden HAs, vomiting, ataxia, dysmetria, & nystagmus	Hyperdense/Type II	4.8	Yes	Excellent
Acciarri et al., 2009	3/42 (7.1%)	15 yrs; 13 yrs; 5 yrs	F; M; M	IH, cerebellar dysfunction; IH, headache, cerebellar dysfunction; IH, HAs (duration of illness before op in all cases 1 day)	—	2 × 1.5; 1 × 2; 1 × 1.2	Yes; yes; yes	Good; good; fair (moderate cerebellar dysfunction)
Xia et al., 2009	4/66 (6%)	—	—	Headaches (n = 3), neurological deficits (n = 2)	—	—	—	—
Consales et al., 2010	2/32 (6.2%)	4 yrs; 16 yrs	M; M	IH; IH, nystagmus	Hematoma; hematoma	—	Yes; yes	No deficits; improved
Amato et al., 2013	2/30 (6.6%)	14 yrs; 11 yrs	M; F	IH, ataxia; coma	Hematoma/mixed density; Type II	—	Yes; yes	Excellent; excellent
Bliginer et al., 2014	3/36 (10.8%)	3 yrs; 15 yrs; 9 yrs	M; M; F	Transient low-extremity monoparesis, ataxia (cerebellar, "incidental finding"); vertigo, ataxia, facial, & abducens nerve palsy (pons-cerebellar pedicle); confusion, dysarthria (parietal & cerebellar lesion)	—	2.3 × 2.2 × 2.1; 1.3 × 1.3 × 0.1; 1.2 × 1 × 1.2	No; yes; yes	—; good; excellent

IH = intracranial hypertension; — = data not available.

* Indicates case report.

† Four years after first presentation, age 16 years.

‡ Adult series of 10 cases; 1 case with age < 18 years.

Considering their location, CCMs are straightforward to access surgically. However, they can become challenging to resect completely when parts of the lesion are scattered diffusely throughout adjacent brain parenchyma, as occurred in one of our patients.

Conclusions

Cerebellar CM presents acutely, in previously asymptomatic patients, with cerebellar parenchymal hemorrhage. The hematoma(s) are commonly large and can be mistaken for hemorrhage into a tumor. Cerebellar CM should be considered when the MRI characteristics suggest hemorrhage of varying age and the coexistence of a DVA. Surgery is a safe treatment option providing excellent clinical outcome in pediatric patients who are treated in a timely fashion. Subtotal resection should prompt ongoing clinical and imaging monitoring, as growth of residual CCM with repeat hemorrhage is possible.

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