CASE REPORT

Middle Cerebellar Peduncle Cavernous Malformation (Cavernoma): Case Report

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Port J Pediatr 2022;53:429-34
DOI: https://doi.org/10.25754/pjp.2022.21867

Abstract

Cavernous malformations are almost asymptomatic cerebral vascular malformations. In children, these low-blood flow lesions are the main causes of cerebral hemorrhage besides arteriovenous malformations. Cavernous malformations can mimic - radiologically and clinically - a bleeding tumor. Magnetic resonance imaging scans provide the diagnosis and then histopathology confirms it. We report a case of a 14-year-old male patient who presented with a five-day history of dizziness, imbalance, tinnitus, hearing loss, and paresthesia of the face and tongue on the right. A magnetic resonance imaging scan revealed a multi-cystic, right middle cerebellar peduncle lesion, showing hemorrhages with various thrombus formation stages as well as developmental venous anomaly. Cerebral angiography ruled out an arteriovenous malformation. Surgery was performed for the excision of the lesion. The decision regarding the surgery was based on the location and symptoms. Histopathology confirmed a cavernous malformation. The patient presented no new neurological deficit and had an uneventful postoperative recovery. The follow-up magnetic resonance confirmed the total removal of the lesion.

Keywords: Adolescent; Hemangioma, Cavernous, Central Nervous System/diagnostic imaging; Hemangioma, Cavernous, Central Nervous System/pathology; Hemangioma, Cavernous, Central Nervous System/surgery; Middle Cerebellar Peduncle

Introduction

Cavernous malformations, also known as cavernous angioma, cavernous hemangioma or cavernoma, are a type of low flow, angiographically occult vascular lesions.[1,2] Cavernous malformations typically appears in patients 20-50 years old.[3] Their prevalence in children is around 0.37%-0.53%.[4,5] Cavernous malformation sizes range from a few millimeters to a few centimeters, with a mean size of 14.2 mm in diameter.[6] Most cavernous malformations are supratentorial (80%) and are commonly located in the parietal lobe and thalamus. Intracranial extra axial cavernous malformations are relatively rare.[2,7]

Symptomatic patients may present with seizures, hemorrhages, mass effect, and neurologic deficits. Cavernous malformations in children may lead to increased hemorrhage risk and be more aggressive compared to adults.[4,5]

Magnetic resonance imaging (MRI) plays a key role in cavernous malformation identification and diagnosis.[8] Over 40% of all cavernous malformations are found incidentally in an MRI.[9] Cavernous malformations might be suspected in cases of large hemorrhagic brain mass, with 'bubbles of blood' of multi-cystic appearance, surrounded by a hemosiderin ring, fluid-fluid levels, and accompanying edema-mass affect.[1]

Complete excision of cavernous malformations is considered the gold standard of treatment. A complete excision results in a long-term cure with the complete recovery of pre-existing neurological deficits in most cases.[3]

We report a case of middle cerebellar peduncle cavernous malformations in a 14-year-old patient and a literature review on clinical presentation, radiological features, surgical management, and outcome.

Case Report

A 14-year-old male patient, with no relevant medical and family history, was admitted to the emergency department, with symptoms of dizziness, imbalance, tinnitus and hearing loss in the right ear, and paresthesia of the face and tongue on the right, already occurring for five days. His condition worsened progressively after

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Received: 21/12/2020 | Accepted: 17/06/2021 | Published online: 03/01/2022 | Published: 03/01/2022

visiting a water amusement park. When questioned, he denied any type of trauma, namely cranial, and said that the symptoms had started on the same day that he had visited the park. He was seen by a doctor three days later, who prescribed ciprofloxacin 1 g/day, presuming acute otitis media on the right. He returned to the emergency department two days later due to the persistence of symptoms, associated with the onset of nausea and vomiting, without otalgia or otorrhea. This time, a pediatrician and neurologist saw the patient. A neurological examination showed a tandem gait with slight imbalance, a positive Romberg test and spontaneous horizontal nystagmus, maximum in levoversion, with rapid left phase, exhaustible. Additional physical examination reported no major changes, including otoscopy. The patient received intravenous metoclopramide and was evaluated by an otorhinolaryngologist, for a condition compatible with peripheral vestibulopathy. After evaluation, antibiotic therapy was discontinued and an audiogram was performed, which revealed hearing loss for low frequency sounds on the right. He was medicated with prednisolone, betahistine dihydrochloride, and metoclopramide, and was referred for consultation with an urgent computed tomography scan (CT). The patient returned to the emergency department two days later, with complaints of intermittent horizontal diplopia with the persistence of the aforementioned symptoms, despite the medication already prescribed. An ear CT scan was performed, which revealed a moderately bulky hematoma centered on the right cerebellar peduncle, with a maximum diameter of about 25 mm, and a reduction in the amplitude of the cerebellum-pontine angle and the lateral recess of the fourth ventricle, without spatial conflict or other alterations, namely alterations suggestive of fracture. In this context, it was decided to perform a cerebral angiography, which excluded the presence of vascular malformations and considered the possibility of cavernous malformation. The patient was transferred to neurosurgery where he underwent an MRI scan. The CT scan showed an intraparenchymal hematoma in the right middle cerebellar peduncle suggestive of the presence of cavernous angioma and perilesional edema with some local mass effect with the impression of the lateral wall of the fourth ventricle (Fig. 1). The MRI scan revealed a focal area with rounded contours in the right middle cerebellar peduncle, heterogeneous, with hemosiderin deposits and focal areas of hypersignal in T2, giving a 'popcorn' appearance suggestive of cavernous angioma. The angioma measured about 24 (anteroposterior) x 15.5 (transverse) x 19.5 (craniocaudal) mm and conditioned the molding of the fourth ventricle without compromising its permeability and without drainage to the fourth ventricle or to the subarachnoid space. The presence of developmental venous anomaly was also observed in planes juxtaposed to the lesion, with an upward path in the cistern of cerebellum-pontine angle. Magnetic resonance showed no arteriovenous malformations or aneurysmal malformations (Fig. 2). Cerebral angiography ruled out an arteriovenous malformation.

Because of the favorable clinical evolution and reversal of neurological symptoms and signs, he was discharged after 17 days of hospitalization, with continued vigilance and indication for a repeated MRI. One week later, an MRI showed signs of the partial reabsorption of the hemorrhagic component located before the cavernous angioma in the right middle cerebellar peduncle. The hemorrhagic content showed signs compatible with late subacute evolution (hypersignal at T1 and T2). Resolution of the parenchymal edema was observed adjacent to the hemorrhage areas and around the cavernous angioma. There was a greater patency of the fourth ventricle and right cerebellum-pontine angle in relation to the reduction of hemorrhage and edema. The cavernous angioma had dimensions approximately overlapping those of the previous MRI. Pending clinical recovery, it was decided to conduct follow-up checks and repeat MRI scans. Despite the patient clinical improvement and subsequent MRI stability (two months later), excision of the cavernous angioma was proposed and performed three months after the onset of the lesion (brain hemorrhage and diplopia), age (higher risk

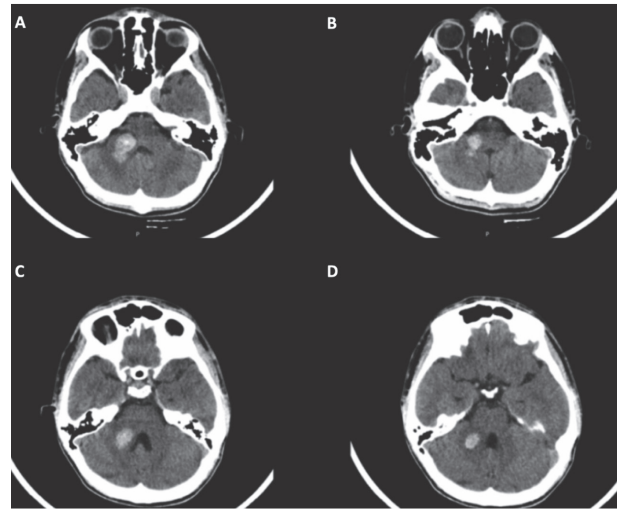

Figure 1. Initial computed tomography scan in 2 mm axial planes without contrast, showing an intraparenchymal hematoma in a right middle cerebellar peduncle, about 24 x 16 mm, with a small rounded area of hyperdensity, and perilesional edema extended to the subcortical white substance of the right cerebellar hemisphere, to the right cerebellar peduncle and right posterolateral slope of the midbrain, with a mass effect on the lateral wall of the fourth ventricle.

of recidivist hemorrhage in youths) and localization (fear of brain stem damage in case of a new hemorrhage).

Surgery was performed with the support of neuronavigation Stealthstation S7 and electrophysiological monitoring with evoked potentials motor, somatosensitive, auditory, and facial nerve. The patient was put in the prone position, with the head in cervical flexion with the aid of a Mayfield support. A median occipital craniotomy was performed with the craniectomy of the border of the magnus foramen and exposure of the posterior C1 arch. Arachnoid dissection, exposure of the fourth ventricle, superior deviation of the cerebellar tonsil, dissection until the superior loop of the right posterior inferior cerebellar artery, identification of a large drainage vein inferomedial to the selected entry zone, and finally a pial incision were performed for detecting the lesion. Evoked potentials without comparative changes at the end of the procedure. The development venous anomaly was spare. The patient was admitted to the pediatric intensive care service for the immediate postoperative period. A control cerebral CT scan showed the presence of intracranial air, namely in the surgical site. There was a small intraventricular hemorrhage and hemorrhagic traces in the interpeduncular cistern. No other significant alterations were observed, namely the deviation of midline structures and signs of hydrocephalus. The removed tissue was sent for anatomopathological study, whose result was compatible with cavernous angioma. He also underwent a control MRI at day three showing a surgical site resulting from the removal of cavernous angioma with cerebrospinal fluid content with a possible protein/blood component, around an intense hyposignal halo in T2, related to the presence of hemosiderin (Fig. 3). In the immediate postoperative period, he presented diplopia with horizontal nystagmus, exhaustible in the left and levoversion, without oculomotricity deficits. The patient was discharged 15 days after surgery with progressive clinical improvement. He was advised to consult an ophthalmologist due to diplopia in the extreme right eye (without nystagmus). One year after surgery, the patient has no type of neurological sequela, although he still has residual diplopia and the MRI no longer presents cavernous angioma. The only sequela of the surgery is visible at the level of the right middle cerebellar peduncle (Fig. 4).

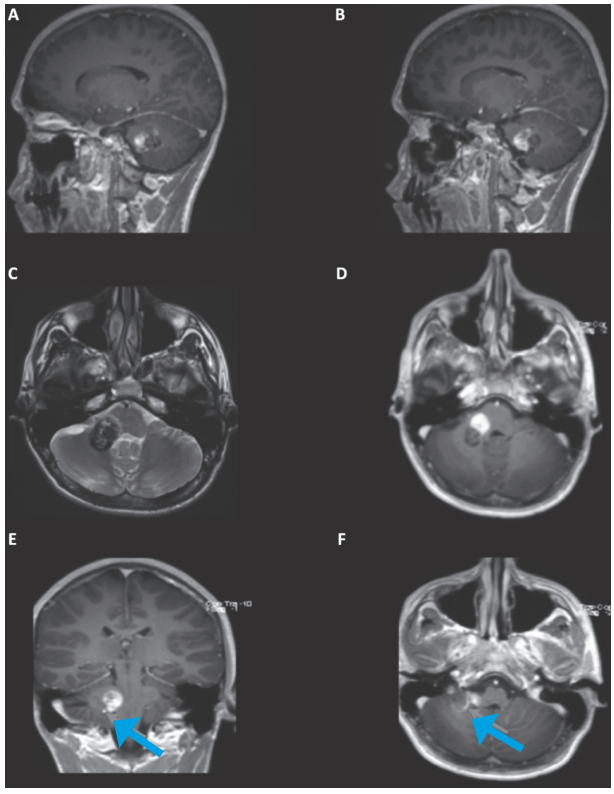

Figure 2. Diagnostic brain magnetic resonance revealing suggestive images of cavernous angioma measuring about 24 (anteroposterior) x 15.5 (transverse) x 19.5 (craniocaudal) mm (A, B: T1 weighted sagittal planes with gadolinium) - focal area in the right middle cerebellar peduncle with marked gradient echo blooming, with hemosiderin deposits and focal areas of T2 hypersignal (enlarged endothelial spaces) giving it a "popcorn-like" aspect (C: T2 axial plane). In the anterior and superior aspects, a hyperintense component in T1 and hypointense in the sequences of long RT, translating hemorrhage into an early subacute phase, with mild adjacent edema (D: T1 axial plane with gadolinium). We can also see a developmental venous anomaly in planes juxtaposed to the lesion, in the posterior and inferolateral aspects, with an upward path in the cistern of the right cerebellum-pontine angle (marked with arrows in figures E: T1 coronal plane with gadolinium, and F: T1 axial plane with gadolinium).

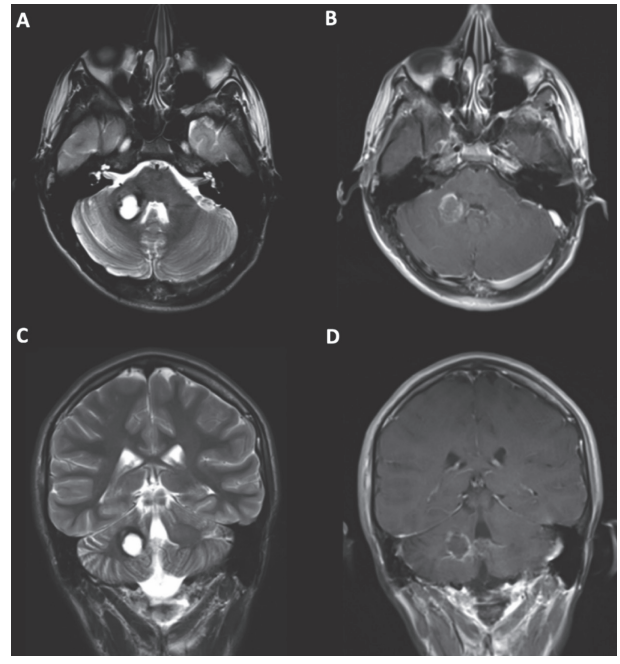

Figure 3. Third day postoperative brain magnetic resonance showing the surgical site resulting from removal of cavernous angioma - 18 (anteroposterior) x 11 (transverse) x 12 (craniocaudal) mm with content with hypersignal in T1 weighted image, cerebrospinal fluid with possible protein/hematic component (intense hyposignal halo in T2 axial plane (A) and T2 coronal plane (C), due to the presence of hemosiderin).

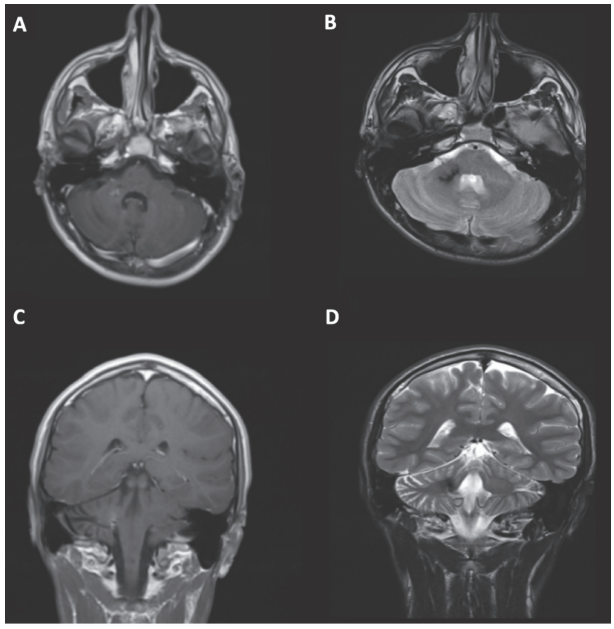

Figure 4. Late postoperative brain magnetic resonance showing the surgical site resulting from the removal of cavernous angioma, with a hypersignal in the right cerebellar peduncle in T1 weighted image C (developmental venous anomaly preserved).

Discussion

Cavernous malformations can be congenital or acquired cerebral vascular malformation that occur in approximately 0.5% of the population.[2] They represent 5%-13% of all central nervous system vascular malformations.[1] They occur sporadically, are usually solitary, and tend to remain asymptomatic. Whereas familial cavernous malformations are multiple in number and disposed to become symptomatic.[10] Cavernous malformations have been reported at every age of the pediatric period, including prenatal and neonatal stages. However, they rarely appear before the first year of life. The mean age of children for clinical presentation is 9-10 years.[11] The overall prevalence among male and female is equal.[12] Most commonly located in the cerebral hemispheres, while most of the posterior fossa cavernous malformations present in the brainstem. Cerebellar cavernous malformations are rare lesions.[1]

Etiology and growth mechanism are still unclear. Amongst the postulated mechanisms are embryogenetic anomalies during vascular development and *de novo* genesis secondary to proliferative vasculopathies.[1] Cavernous malformations can be associated with other vascular malformation and other central nervous system pathologies like tumors, visceral hamartomas, and extra cerebral soft tissue tumors. Cavernous malformations association with venous malformations, capillary telangiectasia and arteriovascular malformations has been described, the most common being developmental venous anomalies (10%-20%). Developmental venous anomalies are more often associated with lesions in the posterior fossa than in the supratentorial compartment.[3,10] In this case report, a developmental venous anomaly was detected.

Most lesions remain asymptomatic throughout life while others present with headache, seizure, or focal neurological deficit due to hemorrhage. The bleeding tendency is greater for familial cavernous malformations *versus* sporadic cases and for cavernous malformations that are associated with developmental venous anomaly or atypical venous drainage. The risk of new bleeding is greater in cavernomas of the brain stem and cerebellar peduncles.[10] Almost invariably, cavernous malformations diagnosis in children is related to radiological investigations for neurological signs or symptoms or, sometimes, for macrocephaly in newborns and infants. Only 14.2% of pediatric cavernous malformations are discovered incidentally.[5,13] The presentation of cavernous malformations is specific to their location. Supratentorial cavernous malformations commonly present with hemorrhage, seizures, and progressive neurologic deficits. Infratentorial cavernous malformations usually present with recurrent hemorrhages and progressive neurological defects.[10,14] Cerebellar cavernous malformations differ from supratentorial ones that present most often with seizures and from brainstem cavernous malformations, which become obvious with focal neurological deficits.[15] Our patient likely had dizziness due to the localization of the cavernous malformations bleeding in the right side of the middle cerebellar peduncle.

Since the introduction of MRI, these vascular lesions have been widely reported in the literature, and their natural history is now better defined.[5] On imaging studies, cavernous malformations are characterized by the presence of blood products of different ages.[9] On MRI, most presented as non-enhancing multi-cystic lesions, representing blood of different ages, and had multiple hemosiderin rings, giving a characteristic "bubbles of blood" appearance. Although not specific, this pattern should prompt the consideration of cavernous malformations in the differential diagnosis. Edema or mass effect may also be present, mimicking neoplasms.[16,17] The main differential diagnosis was hemorrhage into a preexisting cerebellar tumor. Because cerebellar tumors are much more common in children than cerebellar cavernous malformations, the diagnosis of tumor is often the first consideration. The spherical and 'cystic' appearance may mimic that of other cystic tumors, such as pilocytic astrocytomas. However, features like an associated developmental venous

anomaly, multilocular lesions on susceptibility-weighted imaging, and several hemorrhagic episodes (bleeding of different ages) favor cavernous malformations as the preoperative diagnosis.

The overall annual risk of hemorrhage of incidentally diagnosed cavernous malformations was reported to be low at 0.4%-2% per year. However, the risk of a second hemorrhage in the following year was estimated to be 4%-5%.[18,19] Cavernous malformations in the pediatric age group were reported to be more prone to bleeding with a more dramatic clinical outcome than those in adults.[20] The current, well-established indications for the surgical resection of cavernous malformations are recurrent hemorrhage, progressive neurological deterioration, and intractable epilepsy, unless the location is associated with an unacceptably high surgical risk.[6] A complete excision results in a long-term cure with the near complete recovery from the pre-existing neurological defects. [3,5,21,22] The presence of developmental venous anomaly in close proximity to a cavernous malformation is important for the surgeon because injury to such veins can cause devastating venous infarction.[23]

WHAT THIS CASE REPORT ADDS

- Cerebellar cavernous malformations can present acute cerebellar parenchymal hemorrhage in previous asymptomatic patients. Hematomas are commonly large and can be mistaken for hemorrhage into a tumor.
- Cerebellar cavernous malformations should be considered when MRI scans suggest hemorrhage of varying age and the coexistence of a developmental venous anomaly.
- Surgery is a safe treatment option and should be attempted whenever possible because low morbidity, good recovery, and a complete cure in the long term are actual possibilities, always considering that location as well as age (cumulative risks in children) are important decision factors.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

Funding Sources

There were no external funding sources for the realization of this paper.

Provenance and peer review

Not commissioned; externally peer reviewed

Consent for publication

Consent for publication was obtained.

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Malformação Cavernosa do Pedúnculo Cerebeloso Médio (Cavernoma): Caso Clínico

Resumo

As malformações cavernosas são malformações vasculares cerebrais habitualmente assintomáticas. São lesões de baixo fluxo sanguíneo e, nas crianças, são a principal causa de hemorragia cerebral, além das malformações arteriovenosas. As malformações cavernosas cerebrais podem, clínica e radiologicamente, mimetizar o sangramento de causa tumoral. Os estudos radiológicos, nomeadamente a ressonância magnética, são o elemento chave para o diagnóstico. A histopatologia confirma o diagnóstico. Relatamos o caso de um paciente de 14 anos, do sexo masculino, que se apresentou com uma história de cinco dias de tonturas, desequilíbrio, zumbido, hipoacusia e parestesia de face e língua à direita. A ressonância magnética revelou uma massa multiquística no pedúnculo cerebeloso médio direito, mostrando áreas de hemorragia com vários estágios de evolução, e uma anomalia venosa do desenvolvimento. A angiografia cerebral excluiu uma malformação arteriovenosa. Foi submetido a cirurgia, com excisão cirúrgica completa, decisão baseada na localização e modo de apresentação sintomática. A histopatologia confirmou a malformação cavernosa. O paciente não apresentou novos défices neurológicos e teve uma recuperação pós-operatória sem intercorrências. A ressonância magnética de controlo confirmou a remoção total da lesão e a histologia confirmou o diagnóstico imagiológico.

Palavras-Chave: Adolescente; Hemangioma Cavernoso do Sistema Nervoso Central/cirurgia; Hemangioma Cavernoso do Sistema Nervoso Central/diagnóstico por imagem; Hemangioma Cavernoso do Sistema Nervoso Central/patologia; Pedúnculo Cerebelar Médio