Ruptured Arteriovenous Malformation Anterior to the Brainstem to a Child with Subsequent Spontaneous Thrombosis: Case Report and Literature Review

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Conflict of interest: None declared

Patient: Female, 6-year-old
Final Diagnosis: Arteriovenous malformation
Symptoms: Nausea • vomiting
Medication: —
Clinical Procedure: Computed tomography • digital subtraction angiography
Specialty: Neurosurgery

Objective: Unusual clinical course

Background: Cerebral arteriovenous malformations (AVMs) are considered to be abnormalities of congenital origin, presumably arising due to a disorder in the process of embryogenesis, in the phase of differentiation of premature vascular domes into mature arteries, capillaries, and veins. The end result of that process is the formation of direct arteriovenous communications, without intervening capillary beds.

Case Report: We report the case of a 6-year-old female who suffered an abrupt deterioration of her level of consciousness due to a subarachnoid hemorrhage located in the basal cisterns. Radiological investigation with magnetic resonance arteriography-magnetic resonance venography (MRA-MRV) was negative, but digital subtraction angiography (DSA) revealed a micro-AVM in the vicinity of the brainstem. The patient subsequently developed communicating hydrocephalus and the repeat DSA, performed 1 month later, failed to re-imagine the lesion. Further workup with DSA 1 year after the ictus was negative for pathological findings.

Conclusions: There are a lot of controversies regarding the optimal imaging modality for surveillance of pediatric AVMs, the time period needed to follow-up a given lesion, even if it is considered treated, and the underlying mechanism of spontaneous thrombosis of untreated, yet ruptured, AVMs. All these issues, along with the unusual mode of evolution of the clinical picture of this lesion are discussed in detail, along with a review of the available literature.

MeSH Keywords: Arteriovenous Malformations • Brain Stem • Intracranial Thrombosis

Abbreviations: CT – computed tomography; MRI – magnetic resonance imaging; MRV – magnetic resonance venography; MRA – magnetic resonance arteriography; ICU – Intensive Care Unit; VP – ventriculoperitoneal shunt; ICP – intracranial pressure; DSA – digital subtraction angiography; TOF – time-of-flight; AVM – arteriovenous malformation; GRE – gradient echo; FSPGR – fast spoiled gradient echo; DWI – diffusion weighted imaging; SE – spin echo; FSE – fast spin echo; FRFSE – fast relaxation fast spin echo; FLAIR – fluid-attenuated inversion recovery; CUBE FLAIR – it is simply the General Electric (GE) name of their sequence and not an acronym

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Background

Arteriovenous malformations (AVMs) are lesions whose primary characteristic is the existence of a direct arteriovenous communication via the presence of a nidus of coiled and tortuous vascular connections that connect, without intervening capillaries, feeding arteries to draining veins [1–3]. Studies bases on autopsy cases illustrated that the estimated prevalence of AVMs is considered to be in the range of 0.06 and 0.11% [2,4]. AVMs are considered to be the most frequent abnormality of intracranial circulation in childhood, and they are the most common underlying pathological substrate in cases of spontaneous intra-parenchymal hemorrhage in children [2,5,6].

Although AVMs are considered as lesions of congenital origin, they less commonly come to clinical attention in children than in adult counterparts, with children consisting only 3% to 19% of AVM population patients [1,5,7].

Their clinical presentation refers to cases of non-traumatic intracerebral hemorrhage that are brought to clinical attention after an episode of AVM rupture. Conventional 4 vessel cerebral angiography (both internal carotid and vertebral arteries), including external carotid artery opacification, continues to be considered the diagnostic modality with the better accuracy for the detection of AVMs. Compared to other contemporary diagnostic modalities, it is most appropriate to define the AVM dimensions, feeding vessels, draining veins, location of the nidus, and the existence of any associated vascular lesions. It also offers the capability for evaluation of the dynamic blood flow through, as well as, around AVMs with the ability to detect small malformations that could be overlooked by other modalities [2,8]. However, one potential limitation is that early angiography performed in the immediate post-rupture period could potentially not imagine a portion of the AVM- this could be attributed to the compression exerted by the resultant hematoma [2,9–11].

Intracerebral hemorrhage is the most frequently reported clinical manifestation of AVMs in children, with 80% to 85% of pediatric patients suffering a hemorrhagic ictus as the initial symptom. They have been correlated with a 25% mortality rate and an annual rate of re-bleeding in the pediatric population in the range of 2–4% [1,12,13].

Sporadically, vascular malformations that are angiographically occult have been reported; most of them concern cavernous lesions. Besides that, angiographically occult AVMs are often discovered in the territory of the middle cerebral artery associated with small hemorrhages, and these malformations may be the underlying offending pathology for intracerebral hemorrhage of unknown etiology [1,14]. Delayed imaging as the clot dissolves and retracts may reveal the underlying pathology, even though the initial radiological workup was negative [1,15].

Children harboring a previously undiagnosed intracranial AVM and present with a hemorrhagic ictus, are reported to suffer a 12% chance of sudden death [16]. Subsequent diagnostic imaging workup verified that a posterior fossa AVM was present in 57% of fatal cases, and all of them were associated with malformations located in areas with an associated increased risk of potential herniation, as in our case.

Case Report

We describe the case of a 6-year-old female who was admitted to the emergency department of our hospital due to an abrupt deterioration of her level of consciousness. Initial clinical evaluation revealed that her neurological status was impaired, with a relevant initial GCS score 12/15 (E: 3, V: 4, M: 5).

The patient underwent immediately a computed tomography (CT) scan, which revealed intraventricular hemorrhage in the region of the third and fourth ventricle, along with subarachnoid hemorrhage with hyperintense hemorrhagic deposits situated across the basal cisterns (namely, cerebellomedullary, preptpine, cerebellopontine angle, perimedullary, as well as the subarachnoid space surrounding the upper cervical cord) (Figures 1, 2).

The patient was intubated, as she depicted rapidly worsening neurological deterioration (GCS 8/15), and another CT scan was performed 3 days later, depicting blood clot within the third ventricle and the dilation-rounding of the frontal horns, indicative of ongoing hydrocephalus (Figures 3, 4). There were some evidence of progression of bleeding, which could explain the relatively rapid onset of hydrocephalus. Although it is not so uncommon that the patient develops hydrocephalus after subarachnoid hemorrhage, we mention it for the sake of completeness of our presentation. Due to her clinical course, further diagnostic workup was performed 1 day after the repeat CT, including magnetic resonance imaging (MRI), magnetic resonance arteriography (MRA) and magnetic resonance venography (MRV), spin echo (SE), fast spin echo (FSE), fast relaxation fast spin echo (FRFSE), fast spoiled gradient echo (FSPGR), diffusion weighted imaging (DWI), T2* gradient echo (GRE), fluid-attenuated inversion recovery (FLAIR), and CUBE FLAIR (the General Electric (GE) name of their sequence and not an acronym) sequences were used, with 3-dimensional (3D) reconstruction, with and without injection of paramagnetic substance. 3D time-of flight (TOF) technique was utilized for the MRA and 2-dimensional (2D) technique for MRV (Figures 5–9).

MRI revealed intraventricular blood with hemosiderin deposits located in the region of the third and fourth ventricle, as well as in the occipital horns of the lateral ventricles. Similar findings were recorded in the region of the basal cisterns, finding that is identical to that of the CT scan.
There was a noticeable enlargement in the dimensions of the third and lateral ventricles, with an associated periventricular edema, most notable in the territory of the occipital horns. MRA revealed only an asymmetrical imaging of the vertebral arteries, the right vertebral artery occupying a narrower lumen than its counterpart. MRV did not reveal any pathological findings. Due to controversy regarding the precise source of the subarachnoid hemorrhage, a digital subtraction angiography (DSA) of both carotid and vertebral arteries was executed. It revealed the presence of a micro-AVM anterior to the brainstem, fed by multiple small penetrating arteries, originating from branches of the vertebral arteries (Figures 10, 11). More precisely, it seems that the AVM is nourished by branches (direct and circumflex) of the anterior medullary, lateral medullary and tonsillomedullary segments of the posterior inferior cerebellar artery, which is a branch of the vertebral artery. Its venous outflow is through median and lateral anterior medullary veins and transverse medullary veins, whose terminal ends form bridging veins that cross the subarachnoid and subdural spaces to reach venous dural sinuses. There was left-sided vertebral artery dominancy and both vertebral arteries injection was needed in order to appreciate the lesion correctly.
Figure 12 illustrates the right vertebral artery, after its selective catheterization. No contribution to the AVM was verified. An effort was made in order to perform embolism of the malformation at the same time, but it was ineffective due to inability to catheterize the feeding arteries because the diameter of their lumen was particularly narrow.

The patient remained intubated in the Intensive Care Unit (ICU) and an intraparenchymal intracranial pressure (ICP) monitoring was introduced. Due to a gradual but consistent elevation of ICP catheter measurements, a new CT scan was performed the next day, which revealed further dilation of the ventricular system and hypodense areas surrounding the frontal horns, indicative of periventricular edema. Based on those findings, an external ventricular drain (EVD) was inserted in the right frontal horn for management of the evolving hydrocephalus.

Two days later, the patient was extubated and discharged from the ICU; there was neither depression in her level of consciousness, nor focal neurological deficits. She was completely oriented, with slightly blurred speech and mild intention tremor.
During her hospitalization, a weaning process was executed in order to remove the EVD, but that was not feasible due to neurological and imaging deterioration during the procedure. Two weeks after the initial insertion of the EVD, a permanent ventriculoperitoneal shunt was inserted (Codmann, Medos-Hakim, opening pressure at 100 mmHg).

One month after the initial ictus, a repeat DSA was performed (Figures 13, 14), which did not reveal any aneurysm, AVM, or dural arteriovenous fistula (AVF). Her neurological course was uneventful, with signs of neurological improvement at 1-year follow-up after the initial event. A DSA was performed 6 months and 1 year after the initial ictus, verifying the absence of any underlying pathological vascular abnormalities.

Discussion

Embryologically, AVM formation is related to either the maintenance of a primitive arteriovenous connection or the
development of a new communication after completion of the relative normal closure process. It is considered that most of these malformations are developing during the third week of embryogenesis [2].

It is well known that the majority of AVMs are located supratentorially, with less common sites of involvement considered to be the cerebellum, brainstem, and the ventricular system. Regarding the posterior fossa, the cerebellum is most commonly involved [5,17–19]. This is one peculiar characteristic inherent in our case, as the AVM was located anterior to the lower part of the brainstem, near the cervicomedullary junction.

Another unusual feature refers to the mode of clinical evolution, which involved the progressive development of communicating hydrocephalus. As far as the pediatric population is considered, AVMs are thought to represent the most common cause of spontaneous intraparenchymal hemorrhage. Pediatric AVMs could also present with recurrent seizures or headaches [2]. More specifically, 80% to 85% of pediatric patients suffer a hemorrhagic ictus as the initial presenting symptom [1], something that was not evident in our patient. It is reported that only 4 cases have been reported in the literature regarding unruptured AVM located in brain stem or posterior fossa affecting children less than 18 years old [17]. The novel characteristic of our patient was the development of hydrocephalus that was neither associated with obstruction of the aqueduct of Silvius, nor with intraventricular hemorrhage, but rather with subarachnoid hemorrhage.

MRI with and without gadolinium contrast enhancement, along with MRA sequences are considered to be valuable diagnostic tools in the investigation of patients suspected of harboring an AVM. It facilitates the precise localization of the lesion and it is of outmost importance in the differential diagnosis between these lesions, tumors and cavernous malformations. Finally, MRA is a noninvasive method for delineation of the vascular anatomy of the offending lesion [1]. A potential point of interest is based on the inability of that diagnostic modality to elucidate the underlying pathology, probably because it was intermingled with blood in the subarachnoid space, which could compress and obscure the lesion.

Regarding the disappearance of the lesion, this was depicted on the initial DSA, we would like to mention that sporadic cases of thrombosis of AVMs after hemorrhage are described in the literature [20–23]. In cases with previous hemorrhage, the most widely accepted underlying pathophysiologic mechanism is associated with compression of the nidus precipitated by the resultant hematoma.

Regarding surveillance after the initial ictus, it is generally that imaging 6–12 months after a negative postoperative DSA is mandatory in order to rule out recurrent or residual AVM [24–27], as there is an increasing volume of evidence that AVMs recur in children [26–30]. When best modality for surveillance of these children is considered, most available data support the concept that delayed postoperative DSA is mandatory in order to detect recurrent AVM in children [24]. In contrast with DSA, MRI and MRA were proved to be unreliable and ineffective in the detection of recurrence [28].

Figure 13. Repeat, after 1 month, DSA, early arterial phase. No AVM is imagined. DSA – digital subtraction angiography; AVM – arteriovenous malformation.

Figure 14. Same DSA, as previously, at late arterial phase. Again, the AVM is not depicted. DSA – digital subtraction angiography; AVM – arteriovenous malformation.
Besides that, the most updated reviews on that matter continue to support the concept that DSA is considerably more effective in surveillance and detection of recurrences [31,32]. Based on that data, we decided to survey our patient with repeat DSA at 1 month and 12 months after the ictus, even though the aforementioned data refer to patients treated for AVMs and not for those whose lesion was spontaneously thrombosed. Further supporting our decision is the fact that the initial MRI/MRA were unable to detect the abnormality, so they could not be a reliable and sufficient surveillance imaging modality. This mode of surveillance is supported by another published investigation [26].

Additionally, a recent publication [33] suggested, for cases of surgically treated AVMs, an early postoperative angiogram, followed by a repeat angiographic study at 1 year to confirm the absence of a nidus. This surveillance scheme is very similar to the one that was followed by our interventional neuroradiology team. According to the previous study, if MRI/MRA are added to the follow-up protocol, the potential diagnostic benefit, in terms of increase of the diagnostic sensitivity or prevention of symptomatic recurrence, is not well documented and the potential additional value of combining MRA with DSA is not justified. To the best of our knowledge, the most recently published analysis about the issue of AVM surveillance concludes that MRI as a surveillance imaging modality for that patient population should, at present, be supplemented by conventional angiography. This is based upon the fact that when TOF-MRA and contrast enhanced MRI are collectively used and interpreted together, their complementary predictive value is in the order of 85%, compared with conventional angiography [34].

A repeat MRI scan was not performed, as its diagnostic value for the follow-up of verified AVMs is proved to be inferior to that of DSA. Apart from that, no neurological deficits were recorded to our patient, which would indicate the performance of another MRI scan in order to elucidate the possibility of an underlying pathological substrate (e.g., infarction), which could be responsible for the resultant neurological deterioration.

Regarding the issue of management of this particular AVM in case it was not automatically thrombosed, and considering the inability to approach it via an endovascular route and surgically due to its proximity to eloquent brain regions, the only acceptable therapeutic possibility would be radiosurgery. Currently, stereotactic radiosurgery (SRS) is thought to be a gradually effective and relatively safe treatment option for pediatric AVMs, for which surgery seems to be associated with severe inherent risks. The potential drawback of this treatment modality is that the patient remains at risk during the time interval until obliteration is complete. Currently, the most suitable candidates for SRS are pediatric patients with relatively small-diameter and volume AVMs located in eloquent brain regions [35].

Conclusions

AVMs in the pediatric population are relatively rare lesions that primarily present with hemorrhage and potential neurological deficits. We present a case harboring an AVM in the vicinity, and not the substance, of the lower brainstem, presenting with an abrupt decline in the level of consciousness and progressively established communicating hydrocephalus. It was only detected via DSA and not with MRI/MRA. The combination of the aforementioned clinical and imaging characteristics, along with the spontaneous resolution of the AVM via thrombosis after the hemorrhagic event, constitutes a constellation of findings that initially placed diagnostic dilemmas and followed a clinical course that was difficult to predict. We would like to outline the rare possibility of AVMs occurring in the vicinity of the brainstem who present with an atypical clinical course such as communicating hydrocephalus and spontaneous thrombosis after initial bleeding.

Department and Institution where work was done

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Conflicts of interest

None.
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