Dilemmas in managing coexisting arteriovenous and cavernous malformations: Case report

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Abstract:
Coexisting arteriovenous malformations (AVMs) and cavernous malformations (CMs) are rare. Here, we present our dilemmas in managing a patient with a cerebral AVM and a pontine CM. A 47-year-old patient suffered from headaches, vomiting, and transient swallowing difficulties. The cerebral computed tomography showed a pontine hyperintense lesion, while the axial magnetic resonance imaging of the head disclosed a frontal interhemispheric AVM and a CM located in the rostral and ventral aspect of the pons. Despite a pontine hemorrhage, the patient underwent microsurgical excision of the frontal lesion in the first place, due to the increased bleeding risk, followed by stereotactic radiosurgery of the pontine CM. On the 6 months follow-up, the patient’s clinical status was stable. A reasonable treatment strategy based on risk stratification is paramount in managing patients with coexisting AVMs and CMs. The optimal outcome frequently requires a staged multidisciplinary approach.

Keywords:
Arteriovenous malformation, cavernous malformation, microsurgery, radiosurgery

Introduction
Cerebral cavernous malformations (CMs), also known as cavernous angiomas or cavernomas, are low-flow venous lesions composed of an abnormal cluster of enlarged, thin-walled capillaries, with no significant feeding arteries or draining veins. On the other hand, arteriovenous malformations (AVMs) consist of variable size vessels, where feeding arteries shunt directly into veins, with no intervening capillary bed. The feeding arteries are frequently enlarged due to the low resistance (as blood bypasses the capillary beds), leading to high-flow conditions.

CMs and AVMs frequently present with headaches, dizziness, and seizures. Both lesions have been associated with significant morbidity and mortality due to the continued risk of intracranial hemorrhage. On angiography, CMs are occult lesions, and the diagnosis largely depends on identifying the characteristic “popcorn” appearance in magnetic resonance imaging (MRI) with a rim of signal loss due to peripheral hemosiderin deposition. Cerebral MRI, computed tomography (CT), and angiography of the intracranial vessels are helpful to visualize the size, shape, and location of a cerebral AVM.

The treatment of CM and AVM is primarily directed to eliminate the risk of intracranial bleeding and seizure control. The neurosurgeon’s armamentarium includes microsurgery, endovascular treatment, and stereotactic radiosurgery (SRS), solely or combined. The choice of treatment depends on the lesions’ location (eloquence and adjacent structures), size, vascular associations, and other parameters such as...
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Coexisting cerebral AVMs and CMs are exceedingly rare, and the underlying pathogenesis remains unclear. The natural history of a dual lesion including an AVM and a CM is unknown. Only 5 cases of mixed cerebral AVM and CM have been reported to the authors’ best knowledge until now. Moreover, there are no reports of cerebral AVMs associated with pontine CM. Therefore, this entity’s management remains under debate. Here, we present the rare case of a 47-year-old male with coexisting cerebral AVM associated with pontine CM and discuss the pathogenesis of this rare occurrence.

Case Report

A 47-year-old male patient presented with an 18-month history of progressive headache, dizziness, increasing frequency of vomiting attacks, and a recent episode of transient confusion. The neurological examination identified moderate swallowing difficulty in fluids, consistent with lower cranial nerve palsy, whereas the family history was unremarkable. The patient was awake, oriented, and cooperated well during the clinical examination. Otherwise, there were no signs from the long tracts, basal ganglia, cerebellum, and the remaining cranial nerves.

The patient underwent a full workup, including radiological and laboratory examination. The head CT/CT angiography (CTA) demonstrated a large hyperdense region at the middle anterior cranial fossa with arterial supply from pericallosal branches and deep venous drainage suggestive of an AVM and a smaller hyperdense area at the brain stem with evidence of recent bleeding, without precise vascular supply [Figure 1]. MRI/MR angiography (MRA) showed a lesion of focal altered signal intensity – hypointense with hyperintensities on T1-weighted images, hyperintense with hypointense foci, and a hypointense perimeter on T2-weighted images situated in the midline of the anterior cranial fossa, extending into the right frontal lobe, consistent with a diagnosis of AVM (Spetzler–Martin Grade 3). In addition, a CM was identified in the midbrain – specifically in the rostral and ventral aspects of the pons. The CM displayed focal altered signal intensities – hyperintense with a peripheral hypointense hemosiderin rim in T2-weighted images, which served as evidence that both malformations had bled at least once [Figures 2 and 3].

Due to the increased rebleeding risk, we decided to remove the AVM surgically in the first place. According to our interventional radiologist, complete AVM obliteration through the endovascular approach was technically demanding. Indeed, the AVM was surgically removed en-bloc, together with its 4 cm nidus, through an interhemispheric approach [Figure 4]. The postoperative CT verified the complete AVM resection without any hemorrhage [Figure 5], whereas the patient’s sensorium improved after surgery (GCS 15). The patient was perfectly conscious (GCS 15) with no motor deficit, cerebellar syndrome, nor cranial nerve impairment. Three months after the index surgery, the patient was unwilling to accept the risks associated with the surgical excision of the pontine CM, which was ultimately treated with SRS. At the 6-month follow-up, no neurological alteration was noted, and the pontine CM remained unchanged on MRI. For the proper understanding of the underlying lesion, we recommended to the patient for genetic counseling and profiling from a dedicated genetic laboratory. Furthermore, we recommended a DSA examination 1 year after surgery for the AVM and annual MRIs of the head for the initial three years of follow-up for both lesions.

Discussion

This case report demonstrated that AVMs and CMs might coexist, and a detailed diagnostic imaging workup is required in these cases. In addition, the treatment of coexisting AVM and CM should be carefully designed based on the anticipated hemorrhage risk and should consider all available treatment alternatives and combinations.

Diagnostic approach

With the broad use of MRI, CM is increasingly identified as a source of midbrain hemorrhage. The value of the cerebral CT was limited in identifying the underlying pontine hemorrhage and setting the suspicion for a frontal lesion. However, the CTA revealed the coexisting AVM, whereas the cerebral MRI showed the presence of the hemosiderin deposition, indicative of at least
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one bleeding episode. CTA and MRA were sufficient in experienced hands for the preoperative planning of frontal AVMs, as in the current case, obviating thus the need for an invasive diagnostic process, such as digital subtraction angiography. Particular lesion features, including the alternating patterns of hyper-/hypointensity areas and the ill-defined nidus on CTA or MRA, could question the ultimate diagnosis. However, the intraoperative findings and the histopathological diagnosis provided the final diagnosis of a partially thrombosed AVM.

Treatment strategy
Our treatment strategy primarily focused on the lesion with the highest bleeding risk, which was the AVM. Of note, the rates for AVM re-hemorrhage in the 1st year and the entire life are as high as 6%–18% and 17%–90%, respectively.[7,8] Likewise, the equivalent risks in patients with CM are as high as 0.25% and 10%, respectively.[2] In the present case, we also had to take into consideration the role of prior CM hemorrhage (hazard rate: 5.6) and midbrain location (hazard rate: 4.4).[9]

Choice of treatment
The optimal management, surgery indications, and timing of intervention of cerebral malformations, including AVMs and CMs, are still under debate.[4,9] The current medium-sized AVM with deep venous drainage (Grade 3 Spetzler–Martin lesion) was amenable to microsurgical resection.[6] Surgical resection offers prompt and complete nidus obliteration, thus being a permanent solution while also allowing removal of the concurrent intracerebral clot in cases of ruptured AVM.[10,11] Other treatment modalities are endovascular occlusion or SRS, either as a sole treatment or as a part of a downgrading process to render the AVM amenable for surgery.[11] Endovascular treatment was of limited value due to the small-sized feeding arteries from the pericallosal artery and the tortuous draining veins to the deep cerebral veins. In addition, preoperative embolization was not required since it was considered
of high rebleeding risk in an already unstable vascular lesion. Furthermore, the embolization material would make tissue handling during surgical removal more difficult. We decided to reserve endovascular treatment if surgery resulted in partial lesion removal. Of note, endovascular treatment alone rarely achieves complete eradication and often requires repeat sessions for a satisfying result while still carrying nonnegligible perioperative morbidity.[12] We decided to treat the pontine CM with SRS after an extensive discussion with the patient on the advantages and disadvantages of all treatment alternatives. The option to observe a clinically unstable CM was rejected. SRS constitutes a debated alternative for the management of CM but is of value in deep-seated lesions and/or when surgical removal is contraindicated, as in poor surgical candidates, aiming to reduce morbidity and mortality due to bleeding while avoiding the risks associated with complex surgery in selected patients.[4,9,10] The option to simply observe an unstable CM was rejected after discussing with the patient the advantages and disadvantages of all treatment alternatives.

Risk of seizures
Both AVMs and CMs are characterized by a variable seizure risk, which is not affected by the treatment choice and previous bleeding.[13] Therefore, the risk of seizures was not a primary modifier of our treatment strategy. Nevertheless, the patient was treated with antiepileptics until a near-normal EEG.

Conclusion
The management of coexisting cerebral AVM and pontine CM is complex. Appropriate lesion prioritization based on the bleeding risk, location of the lesions, eloquence, and efficacy/safety of available treatment alternatives treatment is of paramount importance. In the overall treatment plan, utmost importance is the patient’s own informed decision, having considered all treatment choices, possible complications, and outcomes.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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