

Coexistent Cerebral Cavernous Malformation and Developmental Venous Anomaly: Does an Aggressive Natural History Always Call for Surgical Intervention?

Abstract

Coexistence of cerebral cavernous malformations (CMs) and developmental venous anomaly (DVA) represents the most common form of mixed intracranial vascular malformations. Existing literature supports not only a possible causative role of DVA for *de novo* CMs but also a potentially detrimental effect on an associated CM, increasing the chances of hemorrhagic complications and growth in the latter. A 52-year-old gentleman presented to us with a 17-year long history of simple motor seizures on the left faciobrachial region. On magnetic resonance imaging (MRI) of the head, a 1.5 cm × 1.5 cm CM without any evidence of recent hemorrhage was identified in the left high frontal premotor area. There was a linear enhancement in the adjoining superior frontal sulcus on contrast MRI. On intra-arterial angiogram, this hyperintensity was confirmed to be a venous channel draining into the superior sagittal sinus. Thus, a diagnosis of cavernoma associated with a DVA was made. The patient was advised conservative treatment and he was doing well at follow-up. Unless diligently looked for, DVA associated with CM may be easily missed. The coexistence has pathophysiological and management implications. Despite the reported aggressive natural history, there is a scope for conservative treatment for these complex vascular malformations.

Keywords: Angiography, cavernous malformation, conservative, developmental venous anomaly, hemorrhage, venous angioma

Introduction

Intracranial vascular malformations are a spectrum of developmental disorders of the cerebral vasculature with an incidence of 2%–4%.^[1] McCormick initially categorized these into four subtypes: arteriovenous malformations (AVMs), cavernous malformations (CMs), capillary telangiectasia, and developmental venous anomaly (DVA)^[2] and “mixed” subtypes were recognized subsequently.^[1,3-5] Out of all the possible combinations of these malformations, the combination of CM and DVA is said to be the most common one.^[6] The natural history and subsequently their treatment planning for these mixed malformations are more complex.^[7] Herein, we report an interesting case of a 52-year-old gentleman with this combined vascular anomaly and review the literature.

Case Report

We admitted a 52-year-old gentleman with a history of three episodes of seizures over

the last 17 years. The last seizure episode was 2 weeks before the admission. At that time, he had undergone a computed tomography (CT) of the head elsewhere and subsequently referred to us for evaluation. His neurological examination was unremarkable.

The CT head showed a small, rounded slightly hyperdense lesion in the left high frontal lobe [Figure 1a]. We investigated him further with a magnetic resonance imaging (MRI) of the head which showed a small, heterogeneous mass in the left high frontal lobe with a peripheral hypointense rim [Figure 1b and c]. The lesion was situated in the premotor area at the posterior edge of the superior frontal sulcus (SFS) [Figure 1b-d]. There was a small, rounded hyperdensity in the SFS just anterior to the lesion [Figure 1b]. On contrast imaging, there was patchy enhancement of the lesion [Figure 1d] along with a linear enhancing structure in the SFS [Figure 1e and f]. On digital subtraction angiogram, the lesion

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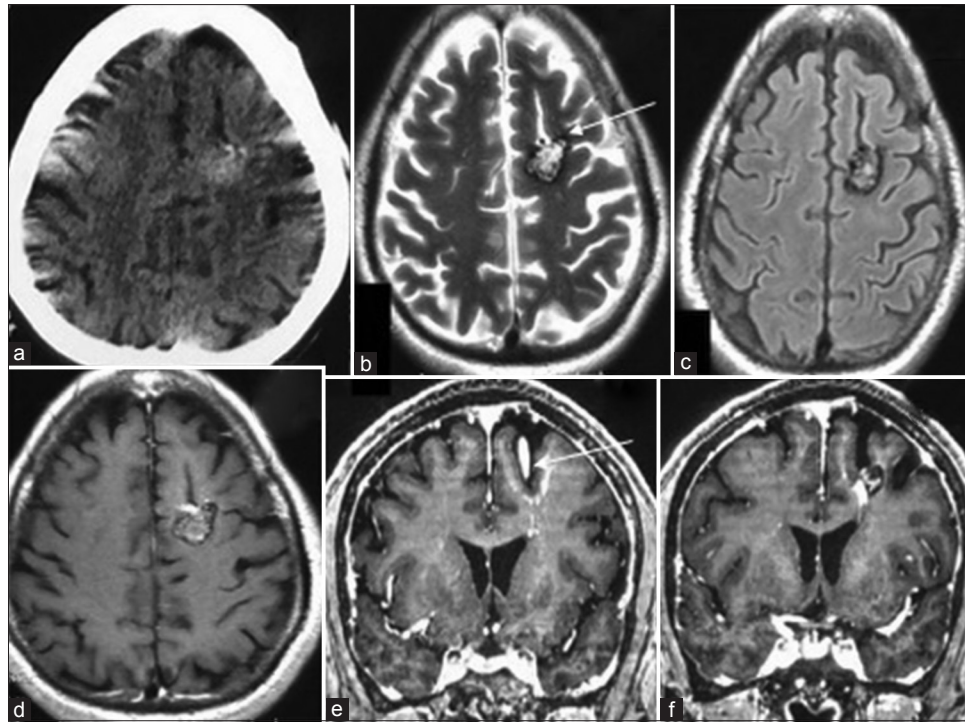


Figure 1: Computed tomography head showed a small, rounded iso-to-hyperdense lesion in the left high frontal lobe without any perilesional edema (a). A popcorn-shaped mass in the left high frontal lobe (marked with white arrow) with a peripheral hypointense rim was visualized on T2 and inversion recovery magnetic resonance imaging images (b and c). The lesion was situated in the premotor area at the posterior edge of the superior frontal sulcus (b-d). On contrast imaging, there was patchy enhancement of the lesion (d) along with a linear enhancing structure in the superior frontal sulcus (e and f) (marked with white arrow)

remained angiographically occult. However, a prominent vein draining into the superior sagittal sinus was seen [Figure 2a and b]. Therefore, an angiographic diagnosis of CM with an associated DVA was made.

Considering the lack of any previous hemorrhagic events, pressure symptoms, and medically well-controlled seizures despite harboring this “high-risk” vascular malformation for so many years, a decision of conservative treatment was taken. We started him on tablet levetiracetam 500 mg twice daily before discharge from the hospital. He was doing well without any new seizures at 6 months of discharge.

Discussion

DVA, also known as venous angioma, accounts for nearly 60% of all intracranial vascular malformations.^[3] Characteristically, it drains normal brain parenchyma without any abnormal arteriovenous shunts. On the other hand, CMs are clusters of venous sinusoids lined by endothelium without any intervening brain parenchyma.^[7] A combination of DVA and CM accounts for the most common type of mixed cerebral vascular malformations.^[6]

It is widely believed now that DVAs almost never cause symptoms on their own and whenever intracranial bleed is associated with a DVA, the dictum is to find out an associated CM. Small size, isodense nature on CT scan, and nonvisualization on diagnostic angiogram contribute

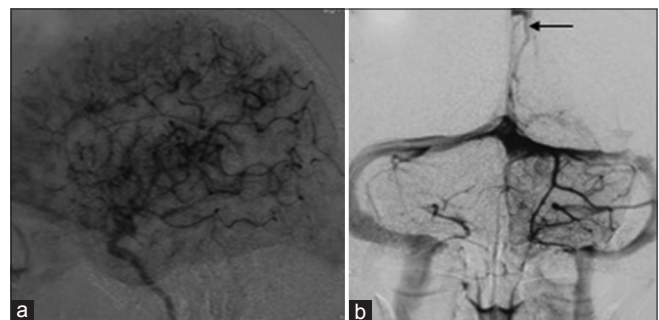


Figure 2: On intra-arterial angiogram, there were no abnormal arteries (a); however, a prominent vein (marked with black arrow) was seen at that location which was seen draining into the superior sagittal sinus (b)

to the missed diagnosis of CMs.^[7] The reverse is also true. The diagnosis of DVA may also be missed on conventional MRI. These missed cases are often detected as small venous channels in the resection cavity during surgery.^[8] Some authors have also suggested that the DVAs may, at times, even be angiographically occult, adding further to their missed diagnosis.^[9]

The association of these two anomalies has pathophysiological and management implications that we specifically like to highlight here. CMs are dynamic lesions that usually grow by microhemorrhages then neoangiogenesis.^[1,6,7] It is believed that DVA has a direct causal-evolutional role in the CM genesis and growth. Chronically elevated pressure inside DVA, as documented

by Wilson *et al.*, can cause hemodynamic pressure load at the point where the medullary veins join the venous trunk.^[10] It may lead to microhemorrhages, which in turn give rise to CM by a process called “hemorrhagic angiogenic proliferation.” Alternatively, the elevated venous pressure may lead to opening up of the hitherto silent arteriovenous collaterals or cause venous ischemia and subsequent recruitment of vasogenic factors. A combination of these effects may lead to *de novo* CM development.^[11]

The fact that the CM in our patient was located at the base of the DVA, we have a strong reason to believe that it resulted from primary venous hypertension in the venous trunk in the SFS. Apart from *de novo* CM formation, associated DVA has been implicated in recurrences following surgical extirpations. Interestingly, the recurrent lesions may be histopathologically different, as noted by Wurm *et al.*^[12]

Does the associated DVA change the natural history of CM when the two lesions coexist? Usually, the natural history of CM is that of progressive growth with neurologic deficits or hemorrhage and rarely a spontaneous regression.^[1,7] CMs also frequently lead to epilepsy. The risk of bleeding in CM *per se* (0.1%–0.6% per year) is much lower than that of an AVM (3%–4% per year).^[7] Various authors have noted that CMs behave more aggressively when there is a DVA at the same site, perhaps due to a communication between the two.^[11,12] Abdulrauf *et al.* noted a 24% increased chances of hemorrhage, while Wurm *et al.* noted that as high as 93.5% of CMs bled when associated with a DVA.^[11,12] Therefore, a combined anomaly must be considered “more seriously” than either of these anomalies in isolation.

Hence, how does one manage a case of DVA and associated CM? Does it call for surgical excision in all cases, even if there are not much symptoms? Our patient had a small CM producing medically controlled seizures and an associated DVA that was draining the pre motor/motor area. No other symptom related to this malformation was present in our patient despite having this anomaly for so many years! Hence, we decided against surgical excision, as the probable complications of surgery were deemed higher than the anticipated benefit. Otherwise, when indicated, surgical opinion largely favors excision of the CM only with sparing of the associated draining venous channel, primary due to the fear of catastrophic venous infarction.^[13-15] Interestingly, some authors have recommended excision of the venous channels lining the cavernoma cavity, not the draining vein *per se*.^[8] Such a strategy has been associated with lesser recurrences and a better seizure control. In a thought-provoking article, Wurm *et al.* attempted a division of the sulcal venous trunk of the DVA and have been able to demonstrate a reduced postoperative recurrence without any major

complications. However, it must be understood that the overwhelming majority of the neurosurgeons would not attempt to divide any angiographically or surgically identifiable sulcal venous trunk for the fear of devastating venous infarction.

Conclusion

The association of DVA with CM is actually more than what meets our eyes. There are important pathophysiological and management implications. Although these lesions may have an aggressive natural history, there is still a scope for conservative treatment even for these “dangerous” lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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