

Trigonal Cavernoma with Developmental Venous Anomaly Causing Intraventricular Haemorrhage: A Once in Blue Moon Association

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ABSTRACT

Intraventricular cavernomas are rare lesions with a high propensity for intraventricular hemorrhage and rehemorrhage. Those located in the trigone of the lateral ventricle are even rarer with <20 cases of trigonal cavernomas reported till date. We present a case of a mixed vascular malformation, a trigonal cavernoma with associated developmental venous anomaly (DVA) causing intraventricular hemorrhage.

Key words: Cavernoma; developmental venous anomaly; intraventricular hemorrhage; mixed malformations

Introduction

Vascular malformations of the brain are classified as those with “high flow” and those with “low flow”. The arteriovenous malformations and fistulas comprise the high flow group whereas the cavernous malformations capillary telangiectasia, venous angiomas comprise the low flow group. Mixed vascular malformations comprise a combination of these subgroups most commonly a cavernoma and venous angioma or a DVA. We report a case of such mixed malformation of a trigonal cavernoma associated with DVA and describe its imaging appearance and its importance in pre surgical evaluation.

Case Report

A 23-year-old young adult Indian female presented with the complaints of acute-onset headache of 3 days duration which was associated with vomiting and altered sensorium. She was immediately transferred to our hospital and was admitted to casualty. On admission, her Glasgow Coma Scale (GCS) was 13 points (E4V4M5), but there was no paresis. The pupils were equal in size and reacted to light. Corneal reflexes were present

bilaterally. Magnetic resonance imaging (MRI) scan was advised and done which showed a heterogeneous mixed intense lesion on T1-weighted, T2-weighted images [Figure 1] in the trigone of the right lateral ventricle. On SWAN, blooming was noted suggestive of hemorrhage in the trigone and the occipital horn of lateral ventricle on the right side. On contrast, MRI with maximum-intensity projection processing typical “medusa” head appearance was noted from the veins adjacent to the cavernoma draining into a collector vein further draining into the cortical vein [Figure 2]. On the post contrast image, the faint patchy heterogeneous enhancement was noted [Figure 3]. Based on the MRI features a final diagnosis of trigonal cavernoma with a developmental venous anomaly (DVA) was considered.

Her complete routine blood counts and biochemistry parameters were normal. The patient was stabilized and kept under observation. Her GCS slowly improved and after a week surgery was planned. At surgery, a right temporoparietal craniotomy was performed, and the occipital horn of the lateral ventricle was explored. The small residual hematoma was seen which was evacuated and the lesion was approached. A well-defined lesion with a thin capsule was noted adherent to the lateral ventricular margin wall with a very small component extending to the adjacent parenchyma. The lesion was displacing the choroid plexus superiorly. The lesion was resected carefully, however leaving the DVA component untouched. Postoperative period was uneventful and patient was later discharged. Histological evaluation of the lesion showed vascular spaces of variable sizes with hemorrhages and lined by endothelial cells with and walls containing fibrous adventitia suggestive of cavernous malformation.

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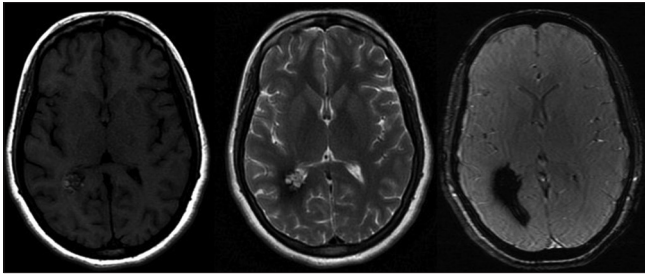


Figure 1: Axial T1-weighted, T2-weighted, gradient images showing heterogeneous lesion in the trigone of a right lateral ventricle with surrounding hypointense hemosiderin rim in adjacent parenchyma on T2-weighted images. On gradient images blooming effect due to intraventricular bleed noted in the occipital horn of lateral ventricle

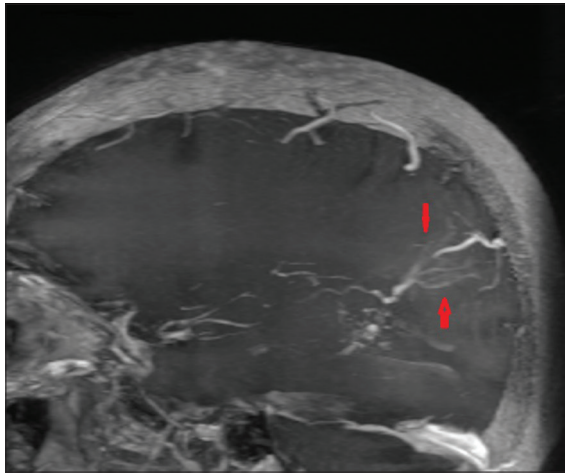


Figure 2: Postcontrast processed maximum-intensity projection magnetic resonance imaging is showing typical medusa head. Appearance (arrow) of venous malformation draining into cortical vein

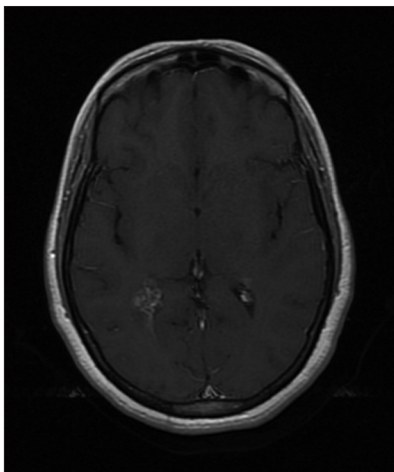


Figure 3: Postcontrast T1-weighted showing heterogeneous contrast enhancement

Discussion

Vascular malformations in the central nervous system have traditionally been classified into four categories: Arteriovenous malformations, capillary malformations

(telangiectasia), venous malformations, and cavernous malformations.^[1] However, this classification scheme does not always accommodate the histopathologic findings. A number of terms have been used to describe the histopathology of cavernous malformations; these entities are referred to chiefly as cavernous angiomas, cavernous hemangiomas, cavernomas, angiographically occult vascular malformations (OVMs), occult cerebrovascular malformations, and OVMs. Similarly, venous angiomas have been known by a variety of terms, including venous malformations, medullary venous malformations, and more recently, DVAs.^[1] The combination of these malformations is termed as “mixed” malformations. Though the incidence of these malformations is not rare to occur individually, the occurrence of mixed malformations is very uncommon. We report a case of such mixed malformation (venous angioma and trigonal cavernoma) and its significance is discussed. Trigonal cavernomas are a very rare entity with only very few cases reported in the literature. With the advent of contrast-enhanced MRI, DVAs, once considered rare, have become the most frequently diagnosed intracranial malformation. Prevalence on contrast-enhanced MR scans ranges from 2.5% to 9%. They are found in all ages without any gender predilection. They are considered to be either an extreme variant of normal venous drainage or supposed to develop due to arrested medullary vein development. The most common location of DVAs is in the deep white matter adjacent to the frontal horn of the lateral ventricle followed by the area adjacent to the fourth ventricle.

Approximately 98% of all DVAs are asymptomatic with most of them being incidentally discovered at autopsy or on imaging. The remaining 2% may present with hemorrhage or infarct owing to the stenosis or spontaneous thrombosis of collector vein.

Coexisting vascular lesions may cause symptomatic intracranial hemorrhage. The most common of such mixed cerebrovascular malformation is a cavernous venous malformation.

Classically DVAs are composed of radially arranged medullary veins^[2] that converge on a transcortical or subependymal large collector vein giving the classic “medusa head” or “upside-down umbrella” appearance.

Cavernoma, also known as cavernous angioma are the third most common cerebral vascular malformation (after DVA and capillary telangiectasia). They are benign malformative vascular hamartomas found in approximately 0.5% of the population with the intraventricular type accounting for only 2.5–10.3% of patients with cerebral cavernous malformations.^[3]

The classic imaging feature is that of a well-circumscribed mixed density/signal intensity mass surrounded by a complete hemosiderin rim (“popcorn ball”).^[4] On noncontrast-enhanced

computed tomography scans the lesion may be undetectable unless large when it appears hyperdense with scattered intralesional calcifications.^[4] They do not exhibit mass effect unless there is recent hemorrhage.^[5] It is the presence of an associated cavernoma in a mixed malformation that results in the symptoms - because of hemorrhage, as it is in our case.

In our case, there was a mixed vascular malformation with the patient presenting with the headache of acute onset associated with vomiting. A trigonal cavernoma with intraventricular bleed and an associated DVA were noted on MRI. The intraventricular bleed probably accounts for the symptoms experienced by the patient.^[6]

Various differentials of trigonal cavernomas include trigonal meningioma, subependymoma, choroid plexus papilloma, central neurocytomas, which have unique MRI features including contrast enhancement and absence of hemorrhages.

Conclusion

It is important to identify the DVA in a mixed malformation preoperatively, in order not to ligate the collector vein or remove its tributaries, which may lead to venous infarction.^[7]

Solitary DVA's do not require treatment and seldom cause hemorrhage. Therefore, it is important to search for an associated vascular malformation in cases of DVA presenting with hemorrhage.^[8]

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