

Surgical Treatment of Brainstem Cavernous Malformation with Concomitant Developmental Venous Anomaly

Abstract

Surgical resection of brainstem cavernous malformations (BCMs) is a high-risk procedure and can be challenging to the neurosurgeon. Lateral surgical routes are becoming increasingly used to approach ventrolaterally brainstem cavernoma. Surgical approach decision depends on the location of the cavernoma in the brainstem and a possible association with brainstem developmental venous anomalies (DVAs). DVA can affect the formation and clinical course of cavernous malformation (CM). CMs related to DVAs tend to have more aggressive behavior than isolated CM. In cases of DVAs associated with hemorrhage, CMs are most often the site of bleeding rather than DVAs themselves. In this case report, we present a 24-year-old woman with a pontomedullary CM and associated dorsally located DVA. BCM was operated through a far lateral suboccipital craniotomy. Brainstem entry point was at inferior olive with extension to the pontomedullary sulcus. This approach should be preferred as a safe surgical exposure to the central and paramedian pontomedullary cavernoma, especially in the cases with associated intraparenchymal brainstem DVA. Such surgical exposure allows preservation of the concomitant brainstem DVA.

Keywords: Brainstem cavernoma, developmental venous anomalies, far lateral approach, hemorrhage

Introduction

Cavernous malformations (CMs) of the central nervous system have an estimated prevalence of 0.4%–0.9% in the general population.^[1–3] Brainstem cavernous malformations (BCMs) are rare and account for 8%–22% of all intracranial cavernomas.^[4] Bleeding and rebleeding (recurrent hemorrhages) rates of BCMs are substantially higher than cavernomas in other locations.^[5,6] Hemorrhages from BCs are never clinically silent comparing with some cerebral cavernomas. They cause severe neurological deficits mainly due to repeated hemorrhages.^[6] Developmental venous anomalies (DVAs) are the most frequently encountered common form of vascular malformations. The reported incidence is 2.6% on autopsy studies,^[7] but with the use of modern imaging techniques, the prevalence is estimated to be much higher (6.4%).^[8] Posterior fossa is a frequent location of DVAs, but drainage through the brainstem is exceptional finding.^[9] CMs associated with

a DVAs have a more aggressive clinical course and are more likely to present with symptomatic hemorrhage than CMs alone.^[10] The goal of operative intervention in such association is complete resection of the CM with preservation of the associated venous anomaly.^[11] The experience of many surgeons has shown that these veins drain normal tissue and their obliteration can lead to venous infarcts.^[11] We present a case of a brainstem pontomedullary cavernoma with concomitant venous anomaly operated through a far lateral suboccipital craniotomy and transolivar approach. Complete excision of the lesion in the medulla was achieved, and gliotic tissue at the pontine level around the abnormal vein was left in order to preserve the associated venous anomaly.

Case Report

A 24-year-old female patient presented with a sudden onset of nausea as well as head-and-neck pain 2 weeks before recent admission. She reported hypoesthesia on the left side of the body, left arm, and leg. Clinical examination revealed also unstable gait and weakness in the left leg. Magnetic resonance imaging (MRI) exam revealed a

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brainstem cavernoma located in the medulla and lower pons with signs of recent bleeding [Figure 1a]. At the same level, a prominent vein could be seen near the floor of the fourth ventricle [Figure 1b-d], draining blood from radiating small veins of the pons and cerebellar peduncles. These small vessels constituted a pontomedullary DVA [Figure 1e and f]. The symptoms could be related to bleeding of the cavernoma.

The patient was placed in the three-fourths prone (“park bench”) position with the head flexed and rotated 45° away from the lesion and laterally flexed downward toward the floor. A far lateral approach was used to expose the occipital bone and suboccipital region from the right side as well as the posterior elements of C1. C1 laminectomy, lateral occipital craniotomy, and condylectomy were performed. After opening of the tonsillomedullary fissure and gently retraction of the tonsil vagoaccessory triangle was approached. A brainstem incision (entry point) was performed on the inferior olive up to the pontomedullary sulcus. Cavernoma excision was performed leaving a part of the gliotic tissue and hemosiderosis in the pons to prevent DVA tributaries damage. Intraoperative brainstem auditory-evoked responses and motor-evoked responses did not show any disturbance.

She had postoperative dysphagia, hoarseness, and right cranial nerve VI palsy at the 1st month after the operation. A nasogastric tube was used during this period until the dysphagia had completely resolved. Two months later, the patient was almost asymptomatic with gait disturbance. A year after the operation, the symptoms resolved completely.

Postoperative imaging showed complete excision of the lesion in the medulla. Surrounding gliotic and hemosiderin-laden tissue at the pontine level was left in the order to preserve the associated DVA [Figures 2a-c and 3]. The patient was informed for the estimated individual rebleeding risk and, in addition, all treatment options and possible morbidities related to them. There was no evidence of recurrent hemorrhage during the first 3 years after the operation.

Discussion

BCMs are associated with severe neurological deficits and repeated hemorrhages. After the initial hemorrhage, the rebleeding rate increases to 45% per person per year.^[12-14] Currently, patients with significant neurological deficits and cavernoma coming up to the pial or ependymal surface are surgically treated. Although there is a morbidity rate mainly related to surgical experience,^[3,15] the long-term outcomes in nonsurgical groups tend to be worse than in surgically treated patients.^[3,6,16,17]

Coexistence between cerebral vascular malformation is not an unusual finding. There has been reported association between DVAs and CMs in approximately 13%–40% of cases.^[18-20] DVAs are considered to be anatomical variant of medullary veins. They represent a compensatory venous drainage system due to aplasia, hypoplasia, or early occlusion of normally developing veins.^[21,22] Although DVAs are benign lesions, they can affect the formation and clinical course of associated CMs. There have been case

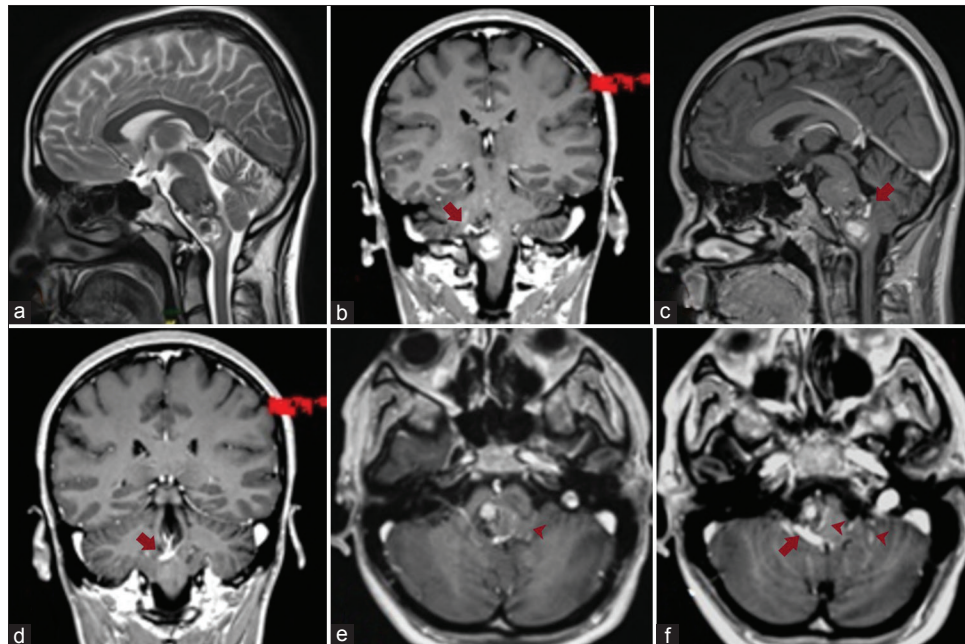


Figure 1: (a) Magnetic resonance imaging T2-weighted sagittal image shows pontomedullary cavernoma after recent bleeding, (b-d) magnetic resonance imaging T1-weighted images with gadolinium. Medullary veins of the pontomedullary developmental venous anomaly exits dorsally to the cavernoma in a large collecting vein near the floor of the fourth ventricle (arrows), and (e and f) magnetic resonance imaging T1-weighted axial images with gadolinium. Transparenchymal small veins – tributaries of the developmental venous anomaly (arrowheads). A large collector draining vein running toward the cerebellomedullary fissure (arrow)

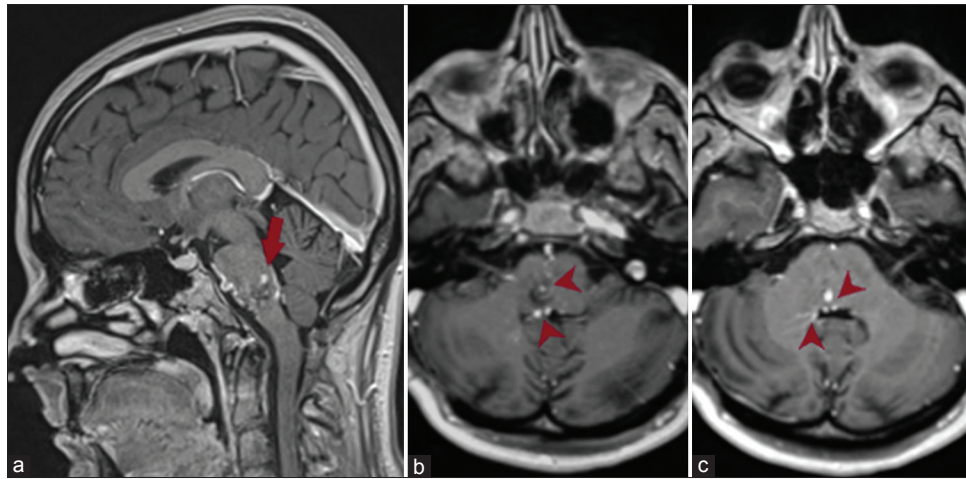


Figure 2: (a-c) Postoperative magnetic resonance imaging T1-weighted sagittal (a) and axial (b and c) images with gadolinium show residual gliotic hemosiderin tissue (arrow) around the small veins – tributaries of the developmental venous anomaly (arrowheads)

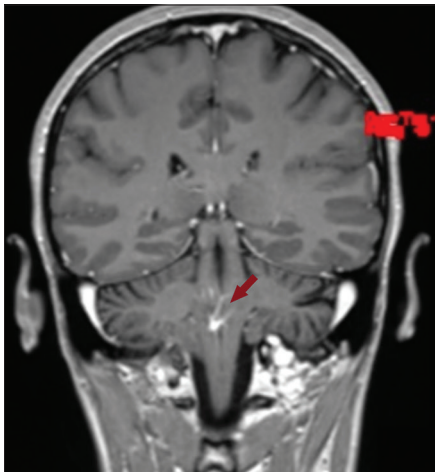


Figure 3: Postoperative magnetic resonance imaging T1-weighted coronal image with gadolinium shows developmental venous anomaly tributaries preservation (arrow)

reports of *de novo* development of CMs in the drainage territory of DVAs.^[23-25] CMs related to DVAs tend to have more aggressive behavior than isolated CMs. In a recent study with persons younger than 45, the presence of a CM in the infratentorial region and the existence of a DVA are key independent hemorrhage risk predictors that may have a key role in treatment decision.^[26] In the case presently reported, we believe that cavernoma resection reduces the venous pressure in the concomitant DVA and facilitates venous drainage of the brainstem.

MRI combined with magnetic resonance angiography (MRA) replaces angiography in most cases of DVAs and CMs as a noninvasive alternative.^[20,27] In the presented case, MRI and MRA were used for the initial diagnosis. Follow-up MRI and MRA proved convenient for long-term follow-up of the patient.

Multiple surgical approaches have been proposed for BCMs. Most of them are exposed through the following

approach: the orbitozygomatic pterional approach, supracerebellar infratentorial approach, retrosigmoid approach, midline suboccipital craniotomy (with or without telovelar dissection), and far lateral approach.^[28]

The central and deep paramedian parts of the medulla and pons are difficult locations for surgical exposure. Central and paramedian pontomedullary CMs are often accessed through a retrosigmoid exposure and a transmiddle cerebellar peduncle approach.^[28,29] Dissection through middle cerebellar peduncle continues approximately 1 cm to reach central or paramedian pons. This could be hazardous for patients with central pontomedullary cavernoma and concomitant deep situated DVA in the brainstem draining through the vein of the middle cerebellar peduncle such in the presented case. Such exposure carries additional high risk from venous brainstem infarction. A recently developed approach through pontomedullary sulcus^[30] could be popularized in such cases with relatively low morbidity and mortality rates. In the presented case, the entry point in the brainstem was through the superior part of inferior olive up to the pontomedullary sulcus. The trajectory passes through the upper part of the vagoaccessory triangle laterally to hypoglossal nerve rootlets, laterally to abducens nerve, and inferomedially to vestibulocochlear and facial nerves. The incision on inferior olive with an extension of the incision in the pontomedullary sulcus expands the surgical exposure to central and paramedian parts of the medulla and the pons simultaneously.

In the presented case, there was a pontine residual cavernoma next to the main drainage of the DVA. Conservative observation and follow-up MRI investigations were performed. We decided that a possible reoperation carries an additional high risk for venous brainstem infarction. There was a nonaggressive appearance of the residual lesion on the MRI and a conservative management was selected in this special case. Three years after the operation, she had no evidence of recurrent hemorrhage.

Conclusion

Inferior olive approach with extension to the pontomedullary sulcus is a safe surgical exposure to the central and paramedian pontomedullary cavernoma. This approach should be preferred, especially in the cases with associated transparenchymal brainstem DVA. Surgical excision of BCMs associated with DVA is related with risk of venous brainstem infarction. Preservation of the associated DVA is mandatory. Such an association could affect the treatment decision.

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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