CASE REPORT

A Rare Case of Focal Multiple Medullary Venous Malformations with Ipsilateral Cerebral Surface Varix

Meguru WATANABE1*, Keiichi ISHIGAME1, Yoshihisa NISHIYAMA2, Hiroyuki KINOUCHI2, and Tsutomu ARAKI1

1Department of Radiology, University of Yamanashi
1110, Chuo-city, Yamanashi 409-3898, Japan
2Department of Neurosurgery, University of Yamanashi
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We report here a rare case of focal multiple venous malformations (VMs) in the white matter, via a draining vein arising from each VM, connecting with an ipsilateral cerebral surface venous varix. The male teen was asymptomatic neurologically. A diagnostic process using of MRI/MRDSA in this extremely rare entity is important as the more incidental discovery is expected with increasing opportunities of performing brain CT/MRI for various indications.

Keywords: cerebral surface varix, focal multiple medullary venous malformations, MRDSA

Introduction

A case of VMs draining into a single surface varix is very rare. This case report describes 1) focal multiple discrete VMs with multiple draining veins connecting with a varix, 2) in a neurologically asymptomatic patient, 3) diagnosis processes with MRDSA. In this report, we demonstrated the significance of MRI/MRDSA in making a diagnosis, and discussed follow-up management for this asymptomatic patient.

Case Presentation

A 15-year-old male patient was hit on his head after accidentally falling to the floor while playing contact sports. There was no loss or altered level of consciousness immediately following the accident. He was brought to a nearby community hospital for assessment. Physical examination revealed no neurological signs. Family history and past medical history were unremarkable. CT scan of the head was taken. The brain CT images demonstrated a dilated venous structure on the right frontal lobe but did not show any other pathologies. He was transferred to a larger university hospital for further evaluation of this incidental CT finding.

Contrast-enhanced MRI of the brain demonstrated a multilobulated structure on the cerebral surface in the left frontal lobe, consistent with a varix (Fig. 1A). Also, a linear contrasting area resembling caput medusa was typically seen in the periventricular area of the left white matter of the frontal lobe (Fig. 1B), representing a typical medullary venous malformation (MVM). Of note, sagittal T1 weighted images post-contrast demonstrated multiple discrete MVMs in the left frontal lobe. Three drainage veins arising from each MVM were seen (Fig. 2A, B, C). These drainage veins were connected to a single varix located in the varix described above (Fig. 3). Contrast-enhanced magnetic resonance digital subtraction (MRDSA) demonstrated no arterial component in these lesions (Fig. 4).

The left thalamostriate vein was tiny, while the left subependymal veins were dilated. There was no stenosis or dilatation at the junction between the cortical varix and superior sagittal sinus.

The patient was followed with MRI study every 6 month. He has been asymptomatic with almost identical MRI findings of the lesions during a 15-month follow-up period.

*Corresponding author. Phone: +81-55-273-1111, Fax: +81-55-273-9766, E-mail: meguruw@gmail.com
Fig. 1. Axial T₁-weighted images post-contrast demonstrated, (A) A bead-like structure on the surface of the left frontal lobe, which was homogeneously enhanced with contrast material. No flow void was seen within the structure. These findings were consistent with a dilated vein, or varix. (B) Multiple linear contrast-ting areas resembling caput medusa in the periventricular area of the left white matter of the frontal lobe. The finding is consistent with venous malformation.

Fig. 2. Sagittal T₁-weighted images post-contrast demonstrated multiple medullary VMs in the left frontal lobe. Three discrete drainage veins were identified arising from each VM. Figure 2A, B, C, from medial to lateral, demonstrated three discrete VMs, each of which had a prominent single draining vein into the cerebral surface varix. These drainage veins were connected to a single varix located in the ipsilateral cortical surface. In Fig. 2A and B, there was a small left thalamostriate vein.
In this patient, at least three MVMs were identified in the left frontal lobe. Each MVM had a prominent single draining vein. The three draining veins were directly connected with the ipsilateral

**Discussion**

In this patient, at least three MVMs were identified in the left frontal lobe. Each MVM had a prominent single draining vein. The three draining veins were directly connected with the ipsilateral
varix on the brain surface. There was no arterial component within these structures. Multiple MVMs with multiple draining veins connecting with cerebral surface varix has not been reported in the English literature so far.

The patient described in this report had coexisting MVM and ipsilateral cerebral surface varix, which was directly connected via a draining vein. These lesions were incidentally found on head CT and brain MRI of this patient, who was neurologically asymptomatic. Coexistence of MVM and ipsilateral cerebral surface varix, as seen in this case, is very rare. Particularly, these lesions have not been identified in a neurologically asymptomatic patient in the English literature to date.

In the case reports published in 1980’s, prior to the common clinical use of MRI, a definitive diagnosis was made with an invasive diagnostic procedure such as angiography. However, a correct diagnosis can be made with contrast-enhanced MRI and MRV nowadays. T₂*-weighted images should be taken to rule out hemorrhage and mixed malformation. In addition to T₂*-gradient echo imaging and post contrast T₁, weighted images, recent studies demonstrated that SWI is helpful to identify venous structures without requiring contrast material.

VM is a slow flow malformation, which does not involve arterial component. VMs in the central nervous system are also called developmental venous anomaly (DVA) or medullary venous malformation (MVM), and are also wrongly known as venous angioma.

A radiographic diagnostic clue of MVM is dilated medullary white matter veins, which are arranged in a radial fashion. VMs drain into a parenchymal venous system to the cerebral surface veins. VMs can be seen in contrast-enhanced CT and contrast-enhanced MRI and MRA. MRDSA typically helps to detect arterial component in arteriovenous malformation and arteriovenous fistula. No arterial components are seen in VMs. MVM is typically seen at the angle of ventricle. Associated abnormalities include co-existing cavernous malformations, which are seen in 15–20% of the patients with MVMs.

While cavernous malformations often bleed, MVMs present with no symptoms. Headache, seizure, and hemorrhage can be seen, but are uncommon. Coexisting cavernous malformation, if any, increases a risk of hemorrhage with focal neurologic deficit. It is reported that the risk of hemorrhage from MVMs was 0.15% per lesion per year. Stenosis or thrombosis of draining vein as well as coexistence of cavernous malformation increases hemorrhage risk. For solitary MVMs, therefore, conservative therapy is recommended.

Follow-up management of the current patient included observation of the patient at outpatient clinic with brain MRI every 6 month. Since MVMs or cerebral surface varix usually cause no symptoms, a patient with these abnormalities needs no treatment. A standard follow-up regimen has yet to be established due to the rarity of the coexistence.

Mechanisms of developing the connection of MVM and ipsilateral cerebral surface varix via a draining vein have not been elucidated. There have been no scientific data on MVM attributes to the development of varix, or vice versa.

Knowledge of the rare coexistence of MVM and Varix allows a diagnostic radiologist to make a correct diagnosis with appropriate diagnostic modalities and protocols. During workups for a patient with incidentally found MVM associated with varix, no invasive diagnostic modality is necessary if the patient is asymptomatic.

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