Angiographically Occult Arteriovenous Malformation in the Septal Region
—Case Report—

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Abstract

An occult arteriovenous malformation (AVM) in the septal region occurred in a 14-year-old boy, manifesting as headache and vomiting. Computed tomography showed a high-density mass in the septal region, faintly enhanced postcontrast. Mild hydrocephalus was also seen. Angiography revealed no abnormalities other than hydrocephalic signs. The lesion was totally removed by the transventricular approach after corticotomy of the left frontal lobe. The histological diagnosis was AVM. He was discharged without neurological or endocrinological deficits.

Key words: occult arteriovenous malformation, septum pellucidum

Introduction

Arteriovenous malformations (AVMs) in the medial sites of the cerebral hemisphere and the limbic system form a special group because of the obscure location and difficulties encountered in removal. The lesion was located in medial sites of the hemisphere or limbic region in 25 of 164 AVM cases (15%) in Stein's series. Of these, two were the septal region or medial sites of the frontal lobe.

Many angiographically occult AVMs have been reported, but occult AVM in the septal region is very rare. We describe a case of occult AVM in the septal region and an alternative surgical approach.

Case Report

A 14-year-old boy complained of headache about 1 month previously, and then vomited a few days before admission. His consciousness was clear and no choked disk was observed. The headache became more severe when he was in the left-upper lateral position. This suggests that the foramen of Monro was obstructed by a mass in that position.

Precontrast computed tomographic (CT) scans showed a high-density mass in the septal region, faintly enhanced postcontrast (Fig. 1). The mass protruded into the left anterior horn. Left carotid and right transbrachial angiograms disclosed no abnormal findings except for hydrocephalic signs (Fig. 2). The preoperative diagnosis was cavernous hemangioma.

Fig. 1 left: Precontrast CT scan, showing a high-density mass in the septal region. Mild hydrocephalus is also present. right: Postcontrast CT scan, showing the faintly enhanced mass.
A left frontal craniotomy followed by a 3 cm corticotomy of the left frontal lobe reached the left anterior horn. A yellowish mass coated with hemosiderin deposits was seen on the anterior side of the foramen of Monro. The mass contained multiple hematoma cavities with old and new clots. Most of the AVM was located in the septal region although it extended into the left anterior horn. Part of the mass wall consisted of abnormal strawberry-like vessels. Partially thrombosed feeding arteries and veins entered from the medial side. The anterior septal vein was located on the medial side of the AVM. The AVM was totally removed without damaging the fornix or anterior septal vein.

Histological examination found no muscular or elastic laminae, but the size and shape of blood vessels were markedly irregular (Fig. 3). The histological diagnosis was AVM.

The postoperative course was uneventful. He was discharged 2 weeks after the operation, without disturbance in mentality, character, or memory function. CT scans showed no AVM (Fig. 4).

**Discussion**

Our case classified according to Stein’s is the septal or medial frontal type. He reported two cases of this type, and recommended removal via low frontal craniotomy and the interhemispheric parafalx approach, with care taken to preserve the main trunk and branches of the anterior cerebral artery and perforating arteries of the anterior perforating space. Dual venous drainage from the lesion often occurred, deep drainage to the ventricular region and superficial to the sagittal sinus.

Generally, the limbic system such as the cingulate gyrus can be damaged more severely by an interhemispheric approach than a transventricular approach. The interhemispheric approach requires retraction of the cingulate gyrus, which may damage some cortical veins or perforating arteries. In our case, part of the AVM protruded into the left anterior horn, so the interhemispheric approach may have required radical sacrifice of the wall of the septum. Therefore, we chose the transventricular approach. Management of blood vessels is certainly easier with the interhemispheric approach than with the transventricular approach. In the present case, however, the AVM was almost completely thrombosed and vessel management was not so critical.

Differential diagnosis between occult AVM and...
cavernous hemangioma is very difficult. Lobato et al. summarized the clinical, radiological, and histological features of 21 angiographically occult intracranial vascular malformations. The clinical presentation, CT appearance, and surgical prognosis were similar for all histological types of vascular malformation.

Occult AVM and cavernous angioma are similar on CT and magnetic resonance (MR) imaging. The characteristic CT appearance of angiographically occult AVMs is usually a well-circumscribed area of increased density with either homogeneous or mottled enhancement postcontrast. Ring enhancement is rarely seen. One-third to one-half of the lesions cause a mass effect. Edema is generally absent. Cavernous hemangioma demonstrates similar patterns. The MR imaging appearance of occult AVMs often includes defined low-intensity areas caused by hemosiderin deposits. These areas are interspersed with mixed signal intensity foci corresponding to different stages of hematoma evolution. Cavernous hemangioma also shows similar patterns.

A final diagnosis of this vascular malformation can only be made by histological examination. AVM contains a mixture of vessel types, veins with thin, collagenous walls, and arteries with muscular and elastic laminae, while many are structural hybrids. The size, shape, and muscularization of the vessels are irregular. In cavernous hemangiomas, the vascular spaces appear as sinusoids, with the walls lined with a single layer of endothelial cells without elastic or muscular layers. In addition, no neural or glial tissue is interposed between the vessels. In this case, muscular and elastic laminae were not present, but the size and shape of vessels were markedly irregular. We therefore diagnosed an AVM.

References


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