Giant Calcified Thrombosed Varices Secondary to a Pial Arteriovenous Fistula Associated With Hereditary Hemorrhagic Telangiectasia

—Case Report of Surgical Removal—

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Abstract

A 28-year-old woman presented with an unusual case of giant thrombosed varix with calcified walls that had mass effects secondary to a pial single-channel arteriovenous fistula (AVF) associated with hereditary hemorrhagic telangiectasia (HHT). She consulted our hospital for chronic headache. She had been diagnosed with HHT based on genetic testing when her 3-year-old son presented with subarachnoid hemorrhage due to spinal AVF. Imaging studies revealed pial single-channel AVF with multiple varices. The varices in the right frontal lobe were over 6 cm in diameter and had laminar thromboses and calcified walls. Because of the mass effect, direct surgical flow disconnection was performed followed by removal of the varices using an internal decompression technique. Postoperatively, the patient was discharged with no neurological symptoms and no longer suffered chronic headache.

Intracerebral varices are occasionally associated with high-flow AVF, and usually treated by interrupting the feeding arteries leaving the varices intact. This extremely rare case of intracerebral giant thrombosed varices with calcified wall and mass effect indicates that surgical removal of varices should be considered.

Key words: calcified thrombosed varix, hereditary hemorrhagic telangiectasia, pial arteriovenous fistula, surgical interruption, surgical removal

Introduction

Intracerebral varices are occasionally associated with high-flow arteriovenous fistula (AVF), and sometimes form large pouches and compress surrounding tissue due to the mass effect.1,4,15–17,20,21) Usually, only disconnection of the feeding artery is sufficient to decompress the lesion because the varices might shrink and immediately lose the mass effect.7) However, if the varices have a hard wall and thrombosed component, only disconnection of the feeding artery might not reduce the volume. Although some cases of large varices associated with AVF have been reported in anecdotal case series,1,4,15–17,20,21) the treatment of such cases has not been well described.

Here we describe an extremely rare case of intracerebral giant laminar thrombosed varices with calcified walls and mass effect, secondary to a single-channel pial AVF associated with hereditary hemorrhagic telangiectasia (HHT). Because of the mass effect of thrombosed varices, direct surgical disconnection of AVF was performed and the varices removed using an internal decompression technique.

Case Report

A 28-year-old woman who had been diagnosed with HHT consulted our hospital. Six months before her consultation, her 3-year-old son had an episode of subarachnoid hemorrhage due to spinal AVF, and genetic tests were performed. Her uncle had also suffered an episode of subarachnoid hemorrhage, but it had not been investigated in detail. The patient had no mucocutaneous telangiectasia or episodes of recurrent epistaxis, but chest computed tomography (CT) revealed a small mass lesion connected with the branch of pulmonary artery and vein in the upper lobe of the right lung that was suspected to be a pulmonary AVF.

The patient suffered chronic headache, but had no neurological deficits or episodes of convulsion. Magnetic resonance (MR) imaging revealed a 6-cm giant laminar tumor-like lesion in the right frontal lobe surrounded by brain malacia compressing the brain, and multiple abnormal flow voids around the sylvian fissure (Fig. 1A, B, D, E). CT revealed that the huge lesion in the frontal lobe had...
thick calcified walls and the frontal bone around the lesion was abnormally thinned, suggesting chronic compression by the lesion (Fig. 1C). Cerebral angiography revealed a pial AVF fed by the anterior branch of middle cerebral artery (MCA) that drained into the superficial sylvian vein through multiple tortuous varices (Fig. 2). The giant tumor-like lesion in the frontal lobe was suspected to be a thrombosed varix.

To decompress the lesion and prevent intracranial hemorrhage, fronto-temporal craniotomy was performed. Opening the dura mater revealed the high-pressure red sylvian vein and varices that refluxed to the precentral vein (Fig. 3A). Opening the sylvian fissure revealed the shunting point fed by the dilated frontal branch of the MCA. Occlusion of the feeding artery using a surgical clip immediately changed the veins and varices to venous color and lowered the pressure (Fig. 3B, C). The non-thrombosed varices were then detached from the pia mater, and giant thrombosed varices were exposed by removal of the frontal brain malacia. The varices had very hard walls, so were fenestrated with a monopolar coagulator and internal thromboses were removed using an ultrasound suction device. After internal decompression, the varices were detached from the brain, the draining vein was ligated, and the lesion was removed en bloc (Fig. 3D–F). Histological examination revealed that the lesion had only the structure of dilated vein with partially calcified and fibrous vascular wall with adherent thrombosis in the lumen, and no elastic fibers in their wall. Hematoxylin and eosin (A) and elastica-Masson (B) staining, original magnifications, ×100.

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After surgery, the clinical course was uneventful. The patient no longer suffered from chronic headache. MR imaging 15 months after surgery revealed that the patient was free from abnormal vascular lesions, and the previously compressed brain had recovered (Fig. 5).

Discussion

HHT is an autosomal dominant disorder of angiodysplasia characterized by a triad of mucocutaneous and visceral telangiectasia, recurrent epistaxis, and family history. Diagnosis is based on the four criteria of spontaneous recurrent nose bleeds; mucocutaneous telangiectasia at characteristic sites (lips, oral cavity, fingers, or nose); visceral involvement such as pulmonary, hepatic, or central nervous system arteriovenous malformations (AVMs); and affected first degree relative. Cerebrovascular malformations (CVMs) occur in 5–11% of patients with HHT. CVMs associated with HHT have specific neuroimaging characteristics. Of all cerebral AVMs in HHT, 28.6–45% were pial single-channel AVFs. AVFs in patients with HHT commonly drain to the superficial cortical veins, usually either an enlarged cortical vein or a venous pouch. Furthermore, 50% of cases of cerebral AVMs in HHT were multiple lesions. Therefore, the pial AVF in this case is a relatively common pattern in patients with HHT. However, no previous cases of giant, calcified, and thrombosed varices secondary to a pial AVF have been reported. In the present case, the diagnosis of pial AVF was relatively easy, because the patient had been already diagnosed with HHT prior to consultation. However, giant thrombosed or serpentine aneurysm should also be considered in the differential diagnosis of such large mass lesions with flow voids. In this case, the acute angled corner of the tortuous varices had a venous pouch, and whorl blood flow in the venous pouch was suspected to result in laminar thrombosis in the varix as seen in thrombosed aneurysms.

Investigation of 28 AVMs (including both AVM and AVF) in patients with HHT found that the bleeding risk ranged from 0.36% to 0.56% per year, significantly lower than the risk for sporadic AVMs. On the other hand, intracerebral hemorrhage arising from such malformations has caused severe deficits. However, the natural history of pial single-channel AVF with HHT remains unknown. Previous experiences of AVMs indicate that single-vein drainage contributes to the risk of AVM hemorrhage as well as small AVM, both of which are characteristics of pial AVF. A family history of intracerebral hemorrhage may be a risk factor for bleeding from CVMs in patients with HHT. The patient’s age at the time of detection should also be taken into consideration.

Most authors have recommended that only the feeding arteries identified by cerebral angiography should be interrupted as closely as possible to the pial single-channel AVF through craniotomy or by endovascular means, leaving the varix intact. However, many small occult feeding arteries that were invisible on preoperative cerebral angiography were identified intraoperatively in one case. In such a situation, surgical observation may help to identify the feeding artery. In the present case, the giant varices in the frontal lobe caused mass effect, and disconnection of the feeding artery may not have been sufficient to decompress the lesion, because the lesions were hard due to internal thrombosis and calcified walls. Instead, resection of the varices was performed. This lesion was surrounded by the brain and a large mass-like tumorous lesion, so internal decompression was performed after flow control of the feeding artery. Although only removal of the thrombosed component, leaving the vascular wall, might have been sufficient to decompress the lesion in this case, the surrounding tissue of the varices was infarcted brain, so detachment of the vascular walls from the surrounding brain was not considered to pose higher risk.

The present rare case of giant thrombosed varices had calcified wall secondary to pial AVF. In such cases, surgical removal of varices should be considered. Furthermore, internal decompression techniques under flow control of the feeding artery may allow detachment of the varices from the surrounding brain.

References

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