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Multiple Cerebral Vascular Malformations and Spontaneous Regression

—Case Report—

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Abstract

A female infant manifested a rare case of spontaneous regression of a vascular malformation in the occipital lobe after removal of another arteriovenous malformation in the frontal lobe. She was born with multiple nevi on the face, body, and upper and lower extremities. She demonstrated developmental retardation at 8 months of age. Computed tomography at 11 months of age demonstrated ventricular enlargement and a mass in the subdural portion of the left anterior fossa. Magnetic resonance images demonstrated signal void signs in the left frontal lobe, which suggested vascular malformation. Cerebral angiograms disclosed two vascular malformations. The malformation in the frontal lobe was totally removed. Cerebral angiograms 25 days after the operation failed to demonstrate either vascular malformation previously observed. Hemodynamic change following the removal of the arteriovenous malformation may have contributed to the occlusion of the remaining malformation.

Key words: multiple vascular malformations, spontaneous regression

Introduction

Cerebral vascular malformation is thought to be a congenital lesion,[6] although presentation in childhood is infrequent. Initial hemorrhage occurs below age 10 years in only 7% of patients, and under the age of 5 years in less than 1%.[12] Cerebral vascular malformations in infants are frequently accompanied by venous varix,[2,4] and commonly manifest as congestive heart failure and enlargement of head circumference.[3,4] Spontaneous regression of arteriovenous malformation (AVM) in the parenchyma is rare,[7,9,11,13] and only three cases have been reported in infants.[5,8,10] We describe a female infant with multiple cerebral vascular malformations, of which one disappeared after excision of the other.

Case Report

This female infant was delivered uneventfully at 40 weeks of gestation after a normal pregnancy, and was the mother's second child. The birth weight was 3480 g. Multiple nevi were noted on her face, body, and upper and lower extremities. The family history included no neurological diseases. Her development was normal at 3 months of age. However, psychomotor retardation, evidenced by failure to sit, was noted at 8 months. Her head circumference was 42 cm, and her weight was 10.5 kg. A bruit was audible on the left supraorbital portion. No cardiac murmur was audible, and no cardiac abnormalities were demonstrated by chest roentgenography. However, an electrocardiogram revealed slight hypertrophy of the left ventricle. Neurological examination discovered no focal deficit, but mild developmental retardation was recognized.

Computed tomography (CT) at 11 months of age demonstrated ventricular enlargement, and a small mass on the surface of the left frontal lobe (Fig. 1 *left*). Magnetic resonance (MR) imaging demonstrated flow void signs on the left frontal lobe (Fig. 1 *right*). Bilateral carotid angiograms revealed a vascular malformation fed by the left orbitofrontal,

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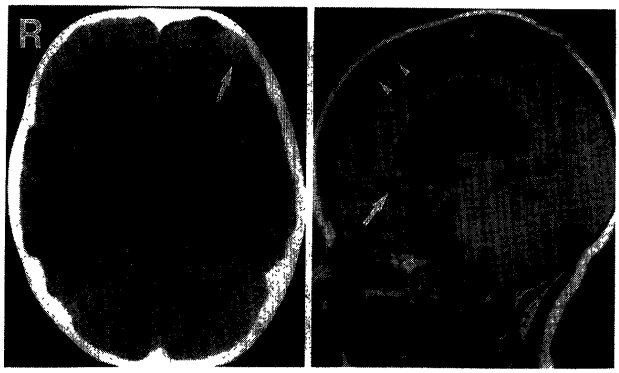

Fig. 1 *left*: Precontrast CT scan demonstrating ventricular enlargement and a mass in the left frontal subdural area (*arrow*). *right*: Sagittal T_1-weighted MR image demonstrating flow void signs in the left frontal area (*arrow*) indicating feeders of the vascular malformation, and dilation of the superior sagittal sinus (*arrowheads*).

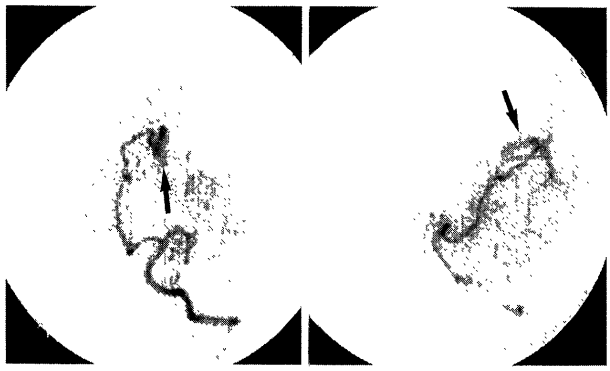

Fig. 3 Left vertebral angiograms, anteroposterior (*left*) and lateral views (*right*), showing a vascular malformation fed by the right calcarine artery (*arrow*).

prefrontal, and frontopolar arteries, and drained by the dilated superior sagittal sinus (Fig. 2). The straight sinus was filled retrogradely from the confluens sinuum, suggesting that the pressure in the superior sagittal sinus was increased due to the shunt flow. Left vertebral angiograms demonstrated early venous filling from the right calcarine artery, and suggested the presence of another vascular malformation (Fig. 3).

A left frontotemporal craniotomy was performed under a diagnosis of multiple vascular malformations causing developmental disturbance when she was 12 months old. The frontal malformation was located on the frontal lobe. The nidus was fed by branches of the middle cerebral artery and frontopolar artery. Two drainers to the sagittal sinus were observed. The main drainer included a venous pouch which was visible on the CT scan. The feeders were cut and the nidus was dissected from the frontal lobe. The main drainer was cut between the venous pouch and the sagittal sinus, and the malformation was removed. Histological examination of the surgical specimen revealed an AVM with marked venous endothelial thickening due to endothelial elastofibrosis.

Bilateral carotid angiograms 25 days after the operation demonstrated no AVM in the frontal lobe, and left vertebral angiograms failed to demonstrate the malformation in the occipital lobe observed preoperatively (Fig. 4). MR imaging 2 weeks after

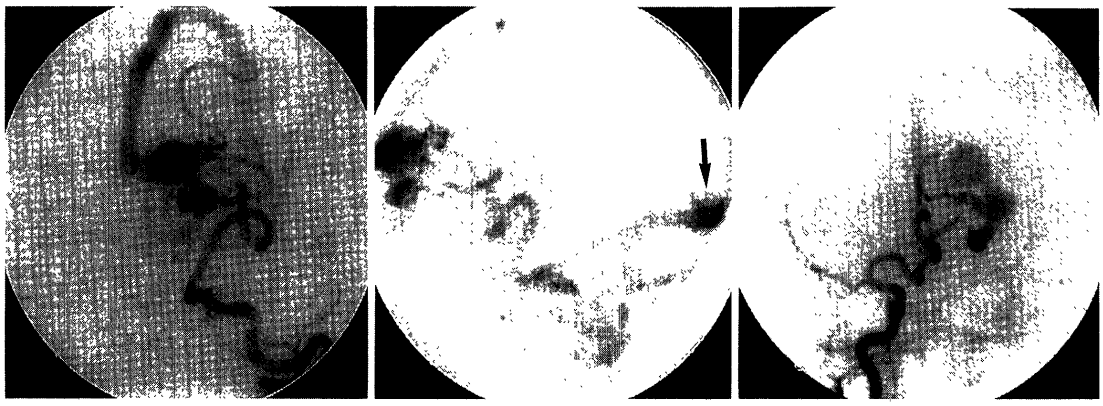

Fig. 2 Left carotid angiograms, left anterior oblique (*left*) and lateral views (*center*), and right carotid angiogram (*right*) showing a vascular malformation with a venous pouch fed by the left orbitofrontal, prefrontal, and frontopolar arteries, and drained by the dilated superior sagittal sinus. The straight sinus was filled retrogradely from the confluens sinuum (*arrow*). The deep venous system was poorly demonstrated.

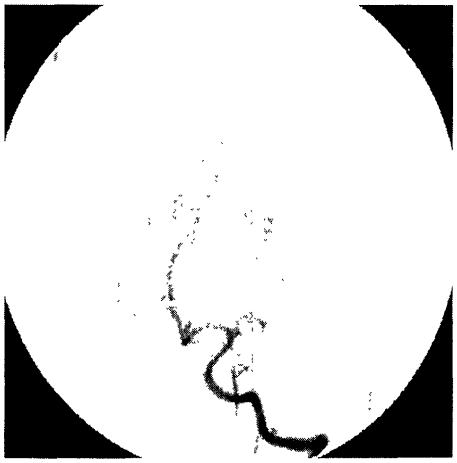

Fig. 4 Postoperative left vertebral angiogram, anteroposterior view, showing absence of the vascular malformation fed by the right calcarine artery.

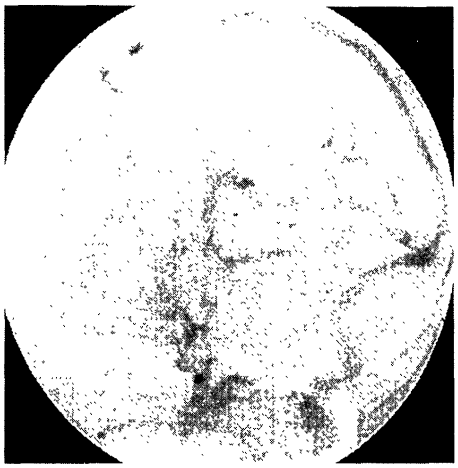

Fig. 5 Postoperative left carotid angiogram, lateral view, demonstrating the deep venous system clearly and the straight sinus with normal filling.

the operation revealed moderate improvement of hydrocephalus. Two months after the operation, hydrocephalus had improved to Evans ratio 0.36 from 0.63 preoperatively, but CT showed bilateral subdural fluid collections. Her developmental score entered the normal range, and she was discharged without any neurological deficit.

Discussion

Our patient had two cerebral vascular malformations located in the frontal and occipital lobes. Angiograms 25 days after the removal of the frontal AVM could not demonstrate the occipital malformation. Closure of arteriovenous fistulas may result from the pressure effect from hematoma and/or hydrocephalus inducing vascular stasis or intraluminal thrombosis, resulting in fistula occlusion.[5] In our patient, hydrocephalus improved after removal of the frontal malformation, so a mass effect cannot have caused the arteriovenous shunt closure in the occipital lobe. Preoperative angiograms revealed retrograde filling of the straight sinus and demonstrated the deep venous system poorly, but postoperative angiograms revealed normal venous flow (Fig. 5). These findings suggest that the flow posteriorly in the occipital malformation increased after the operation.

Angiography frequently shows vessel wall irregularity and stenosis in malformations.[14] Histological examination has found marked phlebosclerotic change of vessel walls in arteriovenous fistula.[1] In our case, marked venous endothelial thickening was observed. These findings suggest that increased shunt flow after the operation promoted the degeneration of vessel walls or acted as a shearing stress on the previously degenerated endothelial cell layer, resulting in occlusion of the shunt channel. Whatever the mechanism, the occipital malformation disappeared in a process related to the removal of the frontal AVM.

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