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Intracranial Pial Arteriovenous Fistula

—Case Report—

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Abstract

A 33-year-old woman presented with a rare intracranial pial arteriovenous fistula manifesting as monoparesis and hypesthesia of the right lower extremity. Computed tomography demonstrated an approximately 10-mm diameter subcortical hematoma in the left postcentral gyrus. Two months after suffering the ictus, angiography demonstrated a pial arteriovenous fistula in the late arterial phase fed by the left paracentral artery and drained into the left precentral vein. No nidus or dural arteriovenous fistula was detected. Left parietal craniotomy was performed and the pial arteriovenous fistula was extirpated by electrocoagulation. Intraoperative angiography demonstrated disappearance of the fistula. She experienced no postoperative neurological deterioration, but hypesthesia of the right leg persisted. Obliteration of the pial arteriovenous fistula was reconfirmed by postoperative angiography. She suffered no rebleeding episodes during the 36-month follow-up period. Pial arteriovenous fistula causing mild symptoms should be treated by flow disconnection because the direct arteriovenous shunt and attendant high blood flow usually results in huge venous varices. To determine whether direct surgery or endovascular treatment is appropriate, the position and shape of the lesion must be known.

Key words: arteriovenous fistula, flow disconnection, pia mater, single venous drainage

Introduction

Intracranial pial arteriovenous fistulae (AVF) are rare cerebrovascular lesions of the brain that are considered to be distinct from other arteriovenous malformations (AVMs).[13] Pial AVF differs from dural AVF in that the arterial supply is derived from pial or cortical arterial vessels and the location is not within the dural leaflets.[11] Pial AVF consist of one or more arterial connections to a single venous channel without any intervening network of vessels. Intracranial pial AVF is rare with only about 90 cases reported since 1970, and account for only 1.6% of a series of 320 AVMs.[11]

We treated a patient with intracranial pial AVF manifesting as only mild symptoms.

Case Report

A 33-year-old woman presented with monoparesis and hypesthesia of the right lower extremity. Computed tomography (CT) obtained at a local hospital demonstrated an approximately 10-mm diameter subcortical hematoma in the left postcentral gyrus (Fig. 1A). Magnetic resonance (MR) imaging failed to disclose the cause of the hemorrhage (Fig. 1B–D) and she was treated conservatively. Her monoparesis improved but hypesthesia persisted.

Two months after suffering the ictus she was referred to our hospital for further examination. Angiography demonstrated a pial AVF in the late arterial phase fed by the left paracentral artery and draining into the left precentral vein. No nidus was detected (Fig. 2). Dural AVF could be excluded because there was no shunt from the external carotid artery (ECA).

Left parietal craniotomy exposed a small red vein joining the left precentral vein below the thickened white arachnoid membrane (Fig. 3A, B). The left precentral vein was not so red but colored like a normal vein, because the red vein joined the precentral vein near the superior sagittal sinus. Retrograde dissection of the red vein disclosed an AVF in the left central sulcus (Fig. 3C, D). Some fine arteries, apparently peripheral branches of the left paracentral artery, connected with the red vein. This lesion was unlikely to be a small AVM, because no nidus or gliosis were present. Moreover, dural AVF could be excluded because no feeding artery was seen from the

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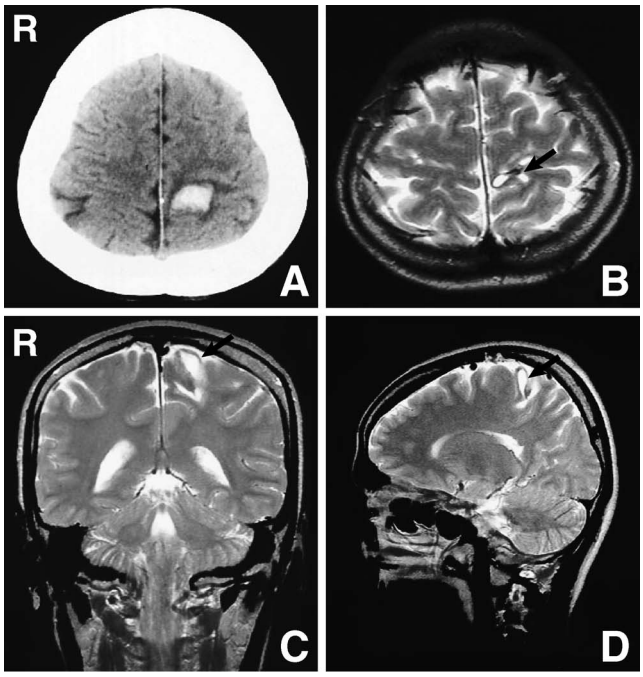

Fig. 1 A: Computed tomography scan demonstrating an approximately 10-mm diameter subcortical hematoma in the left postcentral gyrus. B–D: T_2-weighted magnetic resonance images showing only a small hematoma (*arrow*).

ECA. No varix formation or dilation of the red vein were observed. The AVF was extirpated by electrocoagulation from the fistula to produce flow from the normal dark vein into the red vein (Figs. 3C–E and 4). Intraoperative angiography demonstrated disappearance of the fistula (data not shown).

She experienced no postoperative neurological deterioration, but hypesthesia of the right leg persisted. Obliteration of the pial AVF was reconfirmed by postoperative angiography (Fig. 5). She suffered no rebleeding episodes during the 36-month follow-up period.

Discussion

Pial AVF may have congenital, traumatic, or iatrogenic causes. Congenital pial AVF usually presents in childhood as part of syndromes such as Rendu-Osler-Weber disease[5,7] and Klippel-Trenaunay-Weber syndrome.[15] Although not much is known regarding the pathophysiological mechanisms giving rise to these lesions, a defect in the embryologic development of the cerebrovasculature has been suggested.[11] Likewise, the pathophysiological mechanisms underlying acquired pial AVF remain to be elucidated, but, like dural AVF, development has been attributed to venous hypertension following head injury, brain surgery, and to venous thrombosis.[11,16,18] Our patient was an adult female without Rendu-Osler-Weber or Klippel-Trenaunay-Weber disease. She had no obvious history of head trauma or brain surgery, but transient venous hypertension following slight trauma or of asymptomatic venous thrombosis with recanalization cannot be excluded completely.

The natural history of pial AVF remains unknown because these lesions are so rare. Spontaneous closure of pial AVF has only been reported once,[17] so these lesions are not expected to close spontaneously. The risk of hemorrhage and rerupture of pial AVF is also uncertain, but may be similar to the risks posed by small AVMs with a single drainer, because pial AVF exhibit characteristics similar to these lesions. The flow and perfusion pressure in pial AVF is high because of the direct arteriovenous shunt,[11] so huge venous varices are not uncommon.[1,3,4,8,20,21] Five of eight patients with conservatively treated pial AVF died.[14] Review of 88 patients with pial AVF found mortality and morbidity rates of 7% and 11%, respectively, following flow disconnection by direct or endovascular surgery.[11] Although our patient had only slight vessel abnormality without venous varix recognizable on CT or MR imaging, our angiographic findings suggested radical treatment was essential to prevent future rebleeding.

We recommend that pial AVF be treated by flow disconnection. The absence of a nidus renders resection of the lesion unnecessary. Elimination of the connection to the high-flow system simultaneously obliterates the anomaly and associated elements. Flow disconnection can be accomplished by either direct or endovascular surgery. Conventional flow disconnection involves clipping or cauterization of the vessels, which are effective but carry a high surgical risk for neurological morbidity if the lesions are deep-seated or located in eloquent areas. Abrupt disconnection may result in hyperemia upon evocation of the normal perfusion pressure breakthrough phenomenon.[2,4,6,7,9,10,19,20]

In contrast, endovascular techniques avoid the risks associated with surgical approaches to deep or eloquent areas. Staged endovascular treatment may further reduce the risk of post-therapeutic hyperemia.[21] Endovascular treatment is not risk-free. First, catheterization of the target points may be difficult due to the angiogeometric configuration.[11] Multiple arterial connections may present one and more technical difficulties in terms of access. Second, endovascular treatment risks the migration of embolization material into the draining vein, lung, or

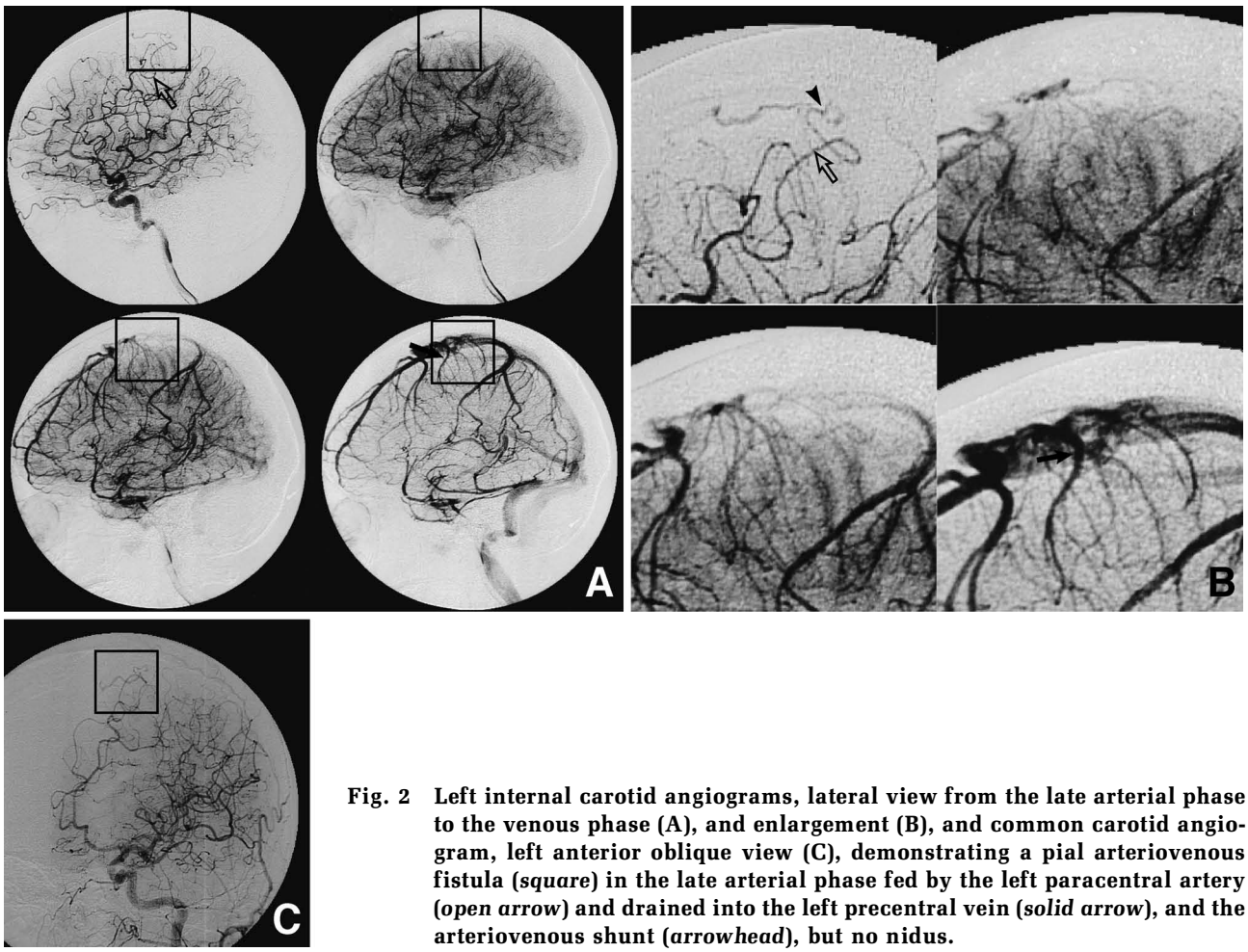

Fig. 2 **Left internal carotid angiograms, lateral view from the late arterial phase to the venous phase (A), and enlargement (B), and common carotid angiogram, left anterior oblique view (C), demonstrating a pial arteriovenous fistula (*square*) in the late arterial phase fed by the left paracentral artery (*open arrow*) and drained into the left precentral vein (*solid arrow*), and the arteriovenous shunt (*arrowhead*), but no nidus.**

elsewhere in the cerebral vasculature.[3,8,12,20] Embolization of a normal vein draining into the venous channel of the pial AVF may result in venous infarction. On the other hand, proximal embolization of the feeder may give rise to new arterial connections and to fistula recurrence.[3,9] Therefore, the therapeutic strategy with pial AVF must be developed for individual patients. We chose direct surgical flow disconnection because the present lesion was located superficially and we were able to preserve the normal cortical vein that drained into the venous channel of the pial AVF.

The present case of pial AVF manifested as subcortical hemorrhage due to rupture. Pial AVF is rare and the natural history remains unclear. Even patients presenting with only mild symptoms should undergo flow disconnection to prevent the risk of rebleeding. To determine whether direct or endovascular surgery is appropriate, the position and shape of the lesion should be ascertained.

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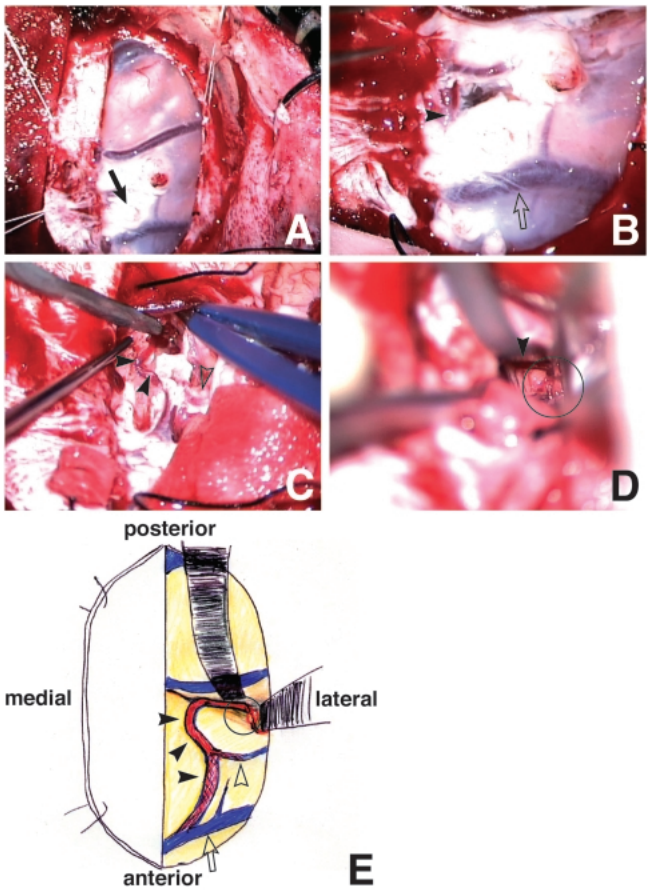

Fig. 3 **A–D: Intraoperative photographs showing (A) the thickened white arachnoid membrane (*solid arrow*), (B) a small red vein (*solid arrowhead*) joining the left precentral vein (*open arrow*) below the thickened white arachnoid membrane, which was not so red but colored like a normal vein, because the red vein joined the precentral vein near the superior sagittal sinus, (C) a normal cortical vein (*open arrowhead*) draining into the red vein (*solid arrowheads*), and (D) an arteriovenous fistula in the left central sulcus (*circle*). Some fine arteries, apparently peripheral branches of the left paracentral artery, connected with the red vein (*solid arrowhead*), but no nidus, gliosis, feeding artery from the external carotid artery, varix formation, or dilation of the red vein was seen (D). E: Schema of the intraoperative findings.**

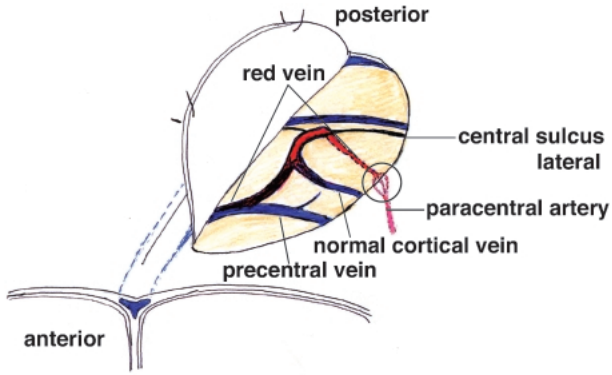

Fig. 4 **Schema of the present case. The red vein was extirpated by electrocoagulation from the fistula (*circle*) to produce flow from the normal dark vein into the red vein.**

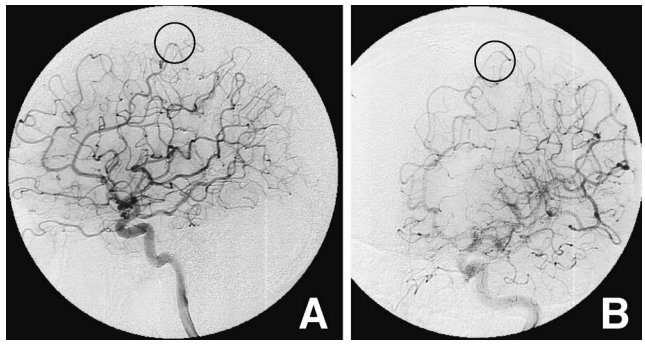

Fig. 5 **Postoperative left internal carotid angiograms, lateral view (A) and left anterior oblique view (B), showing the pial arteriovenous fistula was obliterated (*circle*).**

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