Intracranial pial single-channel arteriovenous fistula presenting with significant brain edema

Case report

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The authors report a rare case of pial single-channel arteriovenous fistula presenting with significant brain edema. A 51-year-old woman was admitted with a 5-day history of headache and nausea, followed by consciousness disturbance. Computed tomography showed cerebellar swelling with obstructive hydrocephalus. Magnetic resonance imaging revealed extensive vasogenic edema in the cerebellum bilaterally. Angiography demonstrated 2 different arteriovenous shunts (AVSs) at peripheral branches of the right anterior inferior cerebellar artery. One was located on the suboccipital surface. It drained through a dilated inferior vermian vein and emptied retrogradely into the contralateral cerebellar veins with marked stagnation. Focal stenosis of the dilated draining vein was present. The other AVS was located on the petrosal surface, which had a slow flow with no angiographic evidence of venous congestion. Given that the latter was believed to be asymptomatic, the former AVS was excised, and histological examination revealed that the lesion consisted of a direct communication of multiple arterial feeding vessels with a single vein, consistent with a diagnosis of pial single-channel arteriovenous fistula. The restriction of venous drainage presumably caused venous hypertension, leading to the brain edema and neurological symptoms.

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Key Words • brain edema • pial arteriovenous fistula • venous congestion

Intracranial pial AVFs are rare vascular lesions of the brain that have only recently been considered a distinct pathological entity from other vascular malformations.8 They differ from pial AVMs owing to the lack of a nidus and from dural AVFs in that they derive their arterial supply from pial or cortical arterial vessels, and the lesion does not lie within the dura mater. Single-channel pial AVFs consist of a single or multiple arterial feeding vessels and a single draining vein without any intervening nidus of vessels or capillaries.5 Although patients with pial single-channel AVFs may present with hemorrhage or seizure, congestive brain edema is a very rare clinical condition, especially in adults. We report the case of a 51-year-old woman with a pial single-channel AVF who developed cerebellar symptoms and hydrocephalus due to significant brain edema.

Case Report

History and Examination. This 51-year-old woman presented with a 5-day history of headache and nausea followed by consciousness disturbance. She had no previous history of stroke, trauma, or other illness. On admission, she was lethargic and exhibited mild truncal ataxia. A laboratory examination of blood and a coagulation profile showed no abnormalities. Computed tomography scanning showed marked cerebellar swelling with hydrocephalus, but no hemorrhage. An extensive hyperintense area in the cerebellum bilaterally was noted on T2-weighted MR imaging (Fig. 1A). The area appeared
hypointense on diffusion weighted images, suggesting vasogenic edema (Fig. 1B). Stringlike enhancement over the area was noted on Gd-enhanced T1-weighted MR images, suggesting dilated pial vessels (Fig. 1C). Conventional angiography revealed 2 different AVSs fed by branches of the right AICA (Fig. 2). One was located on the suboccipital surface. It drained through a tortuous and dilated inferior vermian vein and emptied retrogradely into the contralateral inferior hemispheric veins with marked stagnation, suggesting venous congestion. The other AVS was located on the petrosal surface. It drained through a hemispheric vein into a tentorial sinus without stasis. The external carotid arteries did not seem to have feeding arteries to these fistulas. Focal stenosis was evident in the dilated draining vein of the former AVS on 3D CT angiography (Fig. 3). Based on these findings, we believed that the former AVS was responsible for the patient's clinical symptoms and that the latter was asymptomatic.

**Operation.** A posterior fossa decompressive craniectomy was performed immediately, with removal of the former symptomatic AVS. There were no abnormalities in the dura mater. The right inferior vermian vein was dilated and appeared red. An abnormal vascular pouch was found on the suboccipital surface. Some small feeding vessels from the AICA converged into the vascular pouch and drained into a vein (Fig. 4). The vascular pouch, including adjacent vessels, was excised.

Serial sections through the surgical specimens show-
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ed the lesion consisting of a direct communication of multiple arterial feeders with a single vein, without a nidus (Fig. 5). Discontinuation of the IEL and prominent thickening of the vascular wall were observed in the vascular pouch. The thick intimal and medial layers were strongly positive for \(\alpha\)-smooth-muscle actin (data not shown). The histological diagnosis was a pial single-channel AVF.

Postoperative Course. Complete obliteration of the symptomatic fistula was confirmed on postoperative angiography. The MR images obtained 1 month after the surgery revealed marked resolution of the abnormal hypointense area (Fig. 1D). The patient was discharged from the hospital without any neurological deficit and resumed her previous work. Given the untreated asymptomatic AVS, a 1-year follow-up angiogram is planned.

Discussion

Our patient had a pathologically confirmed AVF that consisted of multiple arterial feeders and a single vein, consistent with a diagnosis of pial single-channel AVF. Intracranial pial single-channel AVFs are rare vascular lesions. In a series of 320 AVMs reported by Halbach et al.,\(^3\) pial single-channel AVFs accounted for only 1.6% of all lesions. They are often associated with cerebral AVMs, or are sometimes multiple as in our case.\(^1\) Although multiple AVFs often occur in patients with Rendu-Osler-Weber disease, an autosomal dominant angiodysplastic disorder characterized by mucocutaneous telangiectases and visceral AVMs,\(^2\) our patient had no family history or clinical features of Rendu-Osler-Weber disease.

Because of its rarity, little has been reported on the...
The histological features of intracranial pial AVF. In the vascular pouch, we observed irregular intimal and medial thickening caused by proliferation of smooth-muscle cells and disappearance of the IEL. These findings are similar to those of the arterialized draining vein in spinal or dural AVFs.\textsuperscript{1,4} Communication between an arterial feeder directly into a draining vein without an intervening tangle of vessels creates a condition in which the draining vein is exposed to increased pressure and blood flow. Abnormal hemodynamic stress may induce chronic endothelial alterations that induce increased endothelial permeability and subsequent intimal fibrous proliferation with destruction of the IEL.

Pial AVFs are considered to be congenital in nature and usually present in childhood or early adulthood. Hoh et al.\textsuperscript{5} reported 9 cases of pial single-channel AVFs; 6 of these patients presented with hemorrhage, 1 presented with a seizure, and 2 had incidental fistulas. The high incidence of hemorrhage seems to be correlated with single-venous drainage and small size, both of which are associated with an increased risk of hemorrhage in pial AVMs.\textsuperscript{9,10} However, a significant brain edema is a rare presentation, especially in an adult patient.

The mechanism of acute development of extensive brain edema in the present case is not clear. The T2-weighted MR images revealed an abnormal hyperintensity in the cerebellum, which reflected vasogenic edema resulting from venous hypertension. This is analogous to the venous congestive myelopathy of spinal AVF. In cases of spinal AVFs, the major factor contributing to the presentation in the 5th and 6th decade of life may be a reduction in spinal venous drainage due to a reduced number of radiculospinal veins and delayed drainage of these radiculospinal veins.\textsuperscript{11} In cases of intracranial dural AVMs, stenosis or obstruction of venous drainage systems is recognized to be a cause of venous hypertension.\textsuperscript{7} We believe that an analogous situation exists in our case: the occurrence of significant brain edema is attributable to venous outflow obstruction due to the focal stenosis of the draining vein. As indicated by 3D CT angiography (Fig. 3), cerebellar veins at prestenotic sites were tortuous and dilated. Moreover, cerebellar edema and medullary vascular enhancement were prominent especially in the suboccipital surface on both sides, corresponding to the draining area of the cerebellar veins at the prestenotic site. These findings may give support to the aforemen-
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tioned mechanism. The cause of the focal stenosis of the draining vein was not clear. Kurata et al. reported on a patient in whom a tentorial dural AVM presented with visual disturbance due to venous ischemia. In their case, the draining cortical vein severely narrowed, and they speculated that the draining vein became stenosed as a result of venopathy caused by shearing stress due to long-term turbulent and high-pressure flow of arterial blood.

Owing to the absence of a nidus, closure of the shunt by either endovascular or surgical technique represents a satisfactory therapeutic procedure. Endovascular occlusion is sometimes effective, especially for deep-seated AVFs. Our lesion was considered to be unfavorable for endovascular treatment as the access routes are too complicated to navigate with the microcatheter. Moreover, the lesion was superficial and easy to reach via a suboccipital craniectomy. Therefore, the present case was treated by resection, which resulted in successful extirpation. Early diagnosis and appropriate treatment for patients with venous congestion are important. If treated at an early stage, venous congestion may be reversible.

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Disclaimer

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References


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