Moyamoya-like disease associated with a lenticulostriate region aneurysm

Case report

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A case of moyamoya-like disease associated with an intracerebral hemorrhage and an unusual aneurysm is reported. The patient's clinical status and the initial and follow-up angiographic appearance of the aneurysm are presented. The etiology of the moyamoya phenomenon and of associated aneurysms is discussed.

KEY WORDS  •  cerebral aneurysm  •  intracerebral hemorrhage  •  moyamoya disease  •  pseudoaneurysm

ALTHOUGH moyamoya disease is considered a rare cerebrovascular disorder, more than 1000 cases had been reported by 1985. Far less frequent have been reports of atypical moyamoya-like disease. Unlike typical moyamoya disease, these cases consist of unilateral stenosis of the internal carotid artery, the anterior cerebral artery, or the middle cerebral artery (MCA) in association with moyamoya-type vessels. In 1985, Sato, et al., presented three patients of their own and reviewed 24 other such cases. Among these 27 cases, only four were associated with demonstrable intracranial aneurysms. A case of moyamoya-like disease associated with an intracerebral hemorrhage and an aneurysm arising from a network of moyamoya vessels in the lenticulostriate distribution is presented.

Case Report

This 60-year-old man experienced the sudden onset of headache followed by loss of consciousness. On admission, the patient was comatose and had a right hemiparesis. Computerized tomography revealed a left frontal intracerebral hematoma with extension to the basal ganglia. An emergency craniotomy and evacuation of the hematoma was performed. Postoperatively, the patient became bright and alert, but had a residual right hemiparesis and a mild expressive aphasia. Left carotid angiography performed 1 week after surgery showed an enlarged anterior cerebral artery and a diffusely stenotic proximal MCA with moyamoya vessels arising from the M1 segment. An aneurysm was detected arising from the distal portion of a moyamoya vessel in the lenticulostriate distribution (Fig. 1 left). The right carotid angiogram was normal. Conservative therapy was selected and the patient's hemiparesis resolved and his speech improved. A follow-up angiogram, performed 3 weeks after the initial study, demonstrated the near-disappearance of the aneurysm (Fig. 1 right). The patient was discharged home in good condition.

Discussion

The case presented here is unusual in terms of the atypical, unilateral appearance of the moyamoya phenomenon ("moyamoya-like disease") as well as its association with a uniquely located aneurysm. Of the 27 cases of moyamoya-like diseases previously reported, only four have been associated with an aneurysm. Among this subgroup, no aneurysm was found to have originated from moyamoya vessels arising from the M1 portion of a stenosed MCA.

While the etiology of the moyamoya phenomenon remains unknown, a congenital form of the disease has been suggested by several reports of familial occurrences of this entity. Numerous acquired conditions have also been linked to the appearance of moyamoya vessels. Neurofibromatosis, tuberculosis meningitis, leptospirosis, and previous radiation therapy have all been implicated. The most common causes of moyamoya-like disease are spontaneous MCA occlusion and arteriosclerosis. In the case presented, angiographic evidence of arteriosclerotic changes does not exist, but...
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severe MCA stenosis is evident. Fukawa, et al.,1 have previously described findings at autopsy that confirmed the presence of spontaneous MCA occlusion associated with the moyamoya phenomenon.

As commonly occurs in adult patients with moyamoya disease,8 our patient presented with an intracerebral hemorrhage. The pathogenesis of the presenting intracerebral or subarachnoid hemorrhage in these patients remains unknown. Most early reports assumed the source of hemorrhage to be the rupture of dilated, tortuous moyamoya vessels. However, the appearance of aneurysms in these patients has raised the new possibility of hemorrhage being secondary to aneurysmal rupture. Kodama and Suzuki2 made a distinction between true saccular aneurysms and "pseudoaneurysms" which spontaneously regress on repeat angiography. Histological proof of the existence of pseudoaneurysms was provided by Yuasa, et al.,12 who presented autopsy findings of such a case and found the "aneurysm" wall to be composed of concentric layers of fibrin with red blood cells between them. Whereas true aneurysms may be considered a source of hemorrhage in patients with moyamoya disease in whom they occur, it is thought that pseudoaneurysms most likely indicate the site of hemorrhage secondary to the rupture of small arteries.2

It is now believed that true aneurysms may occur with greater frequency than was previously appreciated. Takahashi9 reported that aneurysms likely represent a significant source of hemorrhage, and predicted that the use of magnification angiography would reveal a higher incidence of aneurysms and microaneurysms than had commonly been accepted. Sato, et al.,7 reported three cases of moyamoya-like disease in which the associated ventricular hemorrhages were attributed to the rupture of microaneurysms in the moyamoya vessels within the subependyma of the ventricle. The location and transient nature of the aneurysm in the case presented here are most consistent with that of a pseudoaneurysm. Whether or not the hemorrhage itself was secondary to unrecognized microaneurysms or simply to the rupture of dilated moyamoya vessels cannot be definitively answered.

The findings of outcome in this patient suggest several points with regard to the management of such cases. Following initial emergency surgical and medical intervention, high-quality magnification angiography must be performed. In cases of moyamoya or moyamoya-like disease, attention must be directed toward the detection of aneurysms. While direct operation is recommended in cases of apparently typical saccular aneurysm,2 conservative management and repeat angiography seem most appropriate for lesions consistent with pseudoaneurysm. Spontaneous regression of the aneurysm on serial angiograms largely confirms the diagnosis of pseudoaneurysm, and further surgical intervention need not be initiated.

References


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