A Case of Probable Moyamoya Disease (Unilateral Moyamoya Disease) Coexisting Arteriovenous Malformation

The authors report a unique case of unilateral Moyamoya disease with a rare combination of arteriovenous malformation (AVM) who presented with intracerebral hemorrhage (ICH). A 50-year-old man suffered from sudden onset of mental deterioration and right hemiparesis. Brain computed tomography (CT) showed intracerebral hemorrhage on left thalamus. Brain CT angiography and cerebral digital subtraction angiography (DSA) revealed AVM combined with unilateral moyamoya disease involving left middle cerebral artery (MCA) and choroid plexus in left lateral ventricle. Intraventricular hemorrhage and hydrocephalus were managed conservatively. A rare case of unilateral Moyamoya disease accompanied by a cerebral arteriovenous malformation is described and discussed with review of pertinent literature.

KEY WORDS: Moyamoya disease - Arteriovenous malformation.

INTRODUCTION

Moyamoya disease is an uncommon cerebrovascular disease characterized by bilateral stenosis or occlusion involving the arteries of the circle of Willis with an abnormal network of fine collateral vessels at the base of the brain. Affected children typically present with seizures, transient ischemic attacks, and infarctions, whereas adults tend to present with cerebral hemorrhage. It is well known that Moyamoya disease sometimes is accompanied by cerebral aneurysm. The disease occurs bilaterally but is usually asymmetric. Atypical Moyamoya disease with unilateral involvement has also been reported. These cases of unilateral involvement are believed to be secondary to variety of causes, including tuberculous meningitis, irradiation, or atherosclerosis. But, in the majority of cases, the cause of the arterial occlusion is not apparent. There are only 15 cases of Moyamoya disease with AVM previously reported in literature, of which have bilateral Moyamoya disease involvement. There are two cases of unilateral Moyamoya disease associated with AVM.

CASE REPORT

A 50-year-old man was presented to the emergency department with a sudden onset of mental deterioration. On clinical examination, the patient was noted to be stuporous and right hemiplegia. He was afebrile, and vital sign stabilized, but he required nitrprusside for blood pressure control. Respiratory and gastrointestinal systems were normal. Laboratory studies including electrolytes, coagulation profile and urinalysis were normal. There was no history of previous tuberculosis or drug abuse. The patient was not on any medication.

His initial examination included a CT angiography, which depicted a hemorrhage in the left thalamus and both lateral ventricles (Fig. 1) and an AVM involving around left MCA and choroid plexus in left lateral ventricle (Fig. 2).
He underwent unilateral ventricular drainage for the associated intraventricular hemorrhage and hyrocephalus.

Cerebral DSA revealed a Spetzler-Martin grade V AVM with entangled vessels around left MCA and choroid plexus in temporal horn of left lateral ventricle and enlarged left vein of Rosenthal, vein of Galen. Furthermore, complete obstruction of left MCA and numerous collateral vessels in left sylvian fissures and early draining to left internal cerebral vein and cerebral cortical veins were revealed (Fig. 3). It was categorized as having a stage III probable moyamoya disease according to the six-stage by Suzuki and Takaku, in this stage the moyamoya vessels appear very prominent, the middle and anterior cerebral arteries develop stenosis, and changes begin to occur in the other intracerebral main arteries.

The AVM and Moyamoya disease appeared to be inoperable and were managed conservatively. After two weeks, he gradually regained full consciousness but mild right hemiparesis remained and his follow up brain CT showed almost resolved hemorrhage involving left thalamus and slightly resolved IVH (Fig. 4).

DISCUSSION

Moyamoya disease is an occlusive cerebrovascular disease characterized by cerebral angiographic features consisting of intracranial stenosis or occlusion of the internal carotid artery or its terminal branches associated with telangiectatic collateral vessels at the base of the brain. In addition to the enlarged lenticulostriate and thalamoperforating arteries, multiple leptomeningeal and transdural anastomosis develop. The etiology of this vascular abnormality remains unclear. Opinion is still divided between a congenital versus acquired hypothesis. The congenital hypothesis postulates that moyamoya vessels are the result of congenital cerebrovascular dysplasia. On the other hand, the hypothesis of acquired abnormalities suggests that moyamoya vessels are collateral pathways arising because of gradually progressing vascular stenosis of the bifurcation of the bilateral internal carotid arteries. It is usually a bilateral disease, however, atypical unilateral involvement has been reported. Although a number of
factors, in the majority of cases the cause is still inapparent.

Cerebral AVMs are developmental lesion of the cerebral circulation consisting of abnormal dilated tortuous arteries and veins with a tangle of closely packed pathologic vessels, resulting in shunting of blood from arterial to venous side without intermediary capillary bed. It is known to be accompanied by a variety of dysplasias involving the feeding arteries and draining veins. Occlusions, stenosis, aneurysm and ectasia are the dysplastic changes affecting the feeding arteries and draining veins. Although congenital, the network of feeding arteries and draining veins of an AVM is not a static entity. Morphologic changes may occur during the evolution of these lesions.

The combined presence of AVM and Moyamoya disease is extremely rare in contrast to the presence of saccular aneurysm in patients with this occlusive disease. To date, there are only 15 cases of Moyamoya disease accompanied by AVM reported in the literature. Ten of the patients presented with symptoms of cerebral ischemia, while the remaining five presented with basal ganglia hemorrhage. In all except two cases, cerebral angiography demonstrated bilateral involvement of the internal carotid arteries with Moyamoya disease. Thirteen out of the 15 AVMs were supplied by moyamoya collateral vessels. One patient had two independent AVMs located in the collateral vessels through transudal communicating arteries.

At present, the pathogenesis and relationship between Moyamoya disease and AVM are still obscure. The question to be answered is whether the Moyamoya disease caused or was caused by the AVM. It is also possible that two independent vascular abnormalities occur in one patient.

Many authors have reported that Moyamoya disease might initiate the formation of AVMs. Lichtor et al. postulated that in Moyamoya disease the perforating vessels and end capillaries become distended in response to ischemia. However, the capillary linkage is not rich enough to reach the cortex, instead the increased blood flow is channeled into the normal draining veins. As a result, these veins become dilated, taking on the appearance of an AVM. Akiyama et al. observed an AVM that enlarged after encephalomyosynangiosis because of new feeders from the external carotid artery. Halatsch et al. reported the growth of an AVM after superficial temporal artery-middle cerebral artery anastomosis.

On the other hand, some authors believe that Moyamoya disease might be induced by AVMs. They observed new or progressive occlusions and stenosis of the arteries that originally supplied the AVMs. Schmit et al. demonstrated that the development of AVM in a patient with Moyamoya disease by repeated angiography over a 9-year period. These reports suggest that such AVMs were not congenital, but developed as a consequence of angiogenic failure. Mawad et al. reported with cerebral AVMs, and they observed arterial occlusions proximal to the vascular malformation in 10 cases. Those authors suggested that progressive vascular occlusion might be induced by AVMs. That is, increased blood flow at the carotid artery bifurcation, leading to focal intimal hyperplasia, with subsequent internal carotid artery stenosis and development of the moyamoya phenomenon. Montanera et al. observed two cases of AVMs with occlusion or stenosis of the arteries that originally supplied the cerebral AVMs.

Despite these possible explanations, it is not known why combined presence of cerebral AVMs and Moyamoya disease is extremely rare. As well as our case, developmental sequence of AVM and Moyamoya disease is obscure and coincidental combined presence of both disease can not be excluded. Moyamoya disease is a progressive and, surgical treatment is often necessary soon after disease detection. Direct and indirect bypasses between the internal and external carotid artery have been performed, and the result are known to be satisfactory. Essentially, the treatment of Moyamoya disease involves the establishment of as much blood flow as possible. The treatment of cerebral AVMs, however is designed to stop blood flow into the lesions. In cases of AVMs with moyamoya disease, the management principles may cause conflicting situations, such that the revascularization procedures required to treat the moyamoya disease might exacerbate the cerebral AVMs. In our case, the treatment was primary symptomatic and supportive as the patient recovered.

CONCLUSION

The author report a rare case of combined presence of unilateral moyamoya disease and cerebral AVM. This is very unusual owing to its coexistence of Moyamoya disease and AVM, atypical unilateral involvement of the moyamoya disease.

References