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Case Report

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A Case of Probable Moyamoya Disease (Unilateal 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[20]. Affected children typically present with seizures, transient ischemic attacks, and infarctions, whereas adults tend to present with cerebral hemorrhage[12,21]. It is welll known that Moyamoya disease sometimes is accompanied by cerebral aneurysm[20,21,23]. The disease occurs bilaterally but is usually asymmetric. Atypical Moyamoya disease with unilateral involvement also has been reported. These cases of unilateral involvement are believe 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[9,12,20,21]. There are only 15 cases of Moyamoya disease with AVM previously reported in literature, 13 of which have bilateral Moyamoya disease involvment. There are two cases of unilateral Moyamoya disease associated with AVM[1,4,5,8,13-19].

CASE REPORT

A 50-year-old man was presented to the emergency department with a sudden onset of mental deterioraton. On clinical examination, the patient was noted to be stuporous and right hemiplegia. He was afebrile, and vital sign stabilized, but he required nitroprusside for blool 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 a AVM involving around left MCA and choroid plexus in left lateral ventricle (Fig. 2).

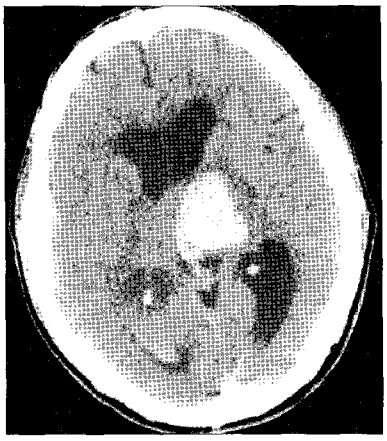

Fig. 1. Initial computed tomography shows intracranial hemorrhage in the left thalamus and both lateral ventricles.

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Fig. 2. Computed tomographic angiography shows a arteriovenous malformation involving around left middle cerebral artery and choroid plexus in left lateral ventricle.

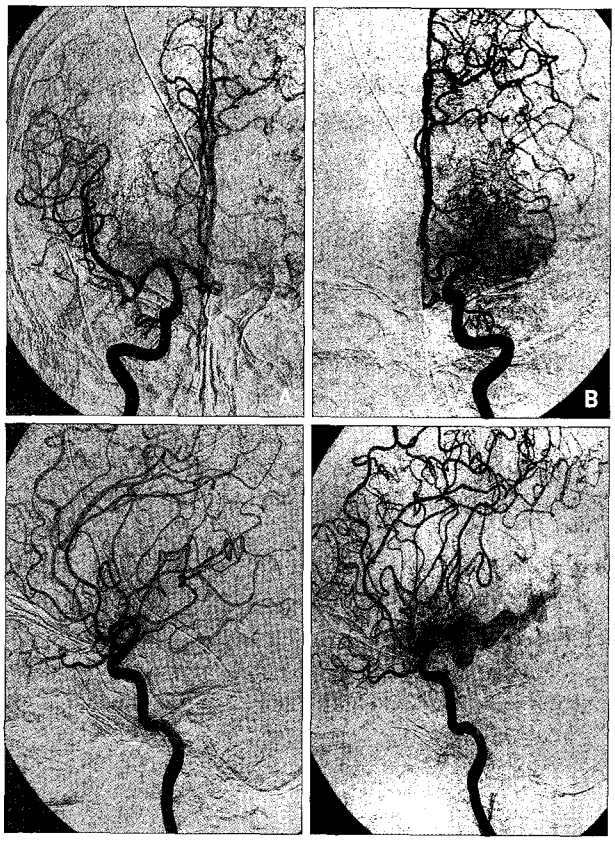

Fig. 3. Digital subtraction angiography shows entangled vessels around left middle cerebral artery and choroid plexus in temporal horn of left lateral ventrticle in early arterial phase anterioposterior view (A) and enlarged left vein of Rosenthal, vein of Galen in midarterial phase lateral view (B) and complete obstruction of left middle cerebral artery compared with right. (A-D).

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 ventrticle 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.

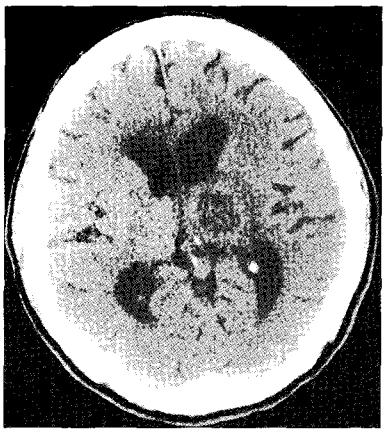

Fig. 4. Brain computed tomography shows much resolved intracranial hemorrhage in left thalamus and slightly resolved intraventricular hemorrhage.

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 invloving left thalamus and slighlty resolved IVH (Fig. 4).

DISCUSSION

Moyamoya disease is an occulsive 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[21]. In addition to the enlarged lenticulostriate and thalamoperforating arteries, multiple leptomeningeal and transdural anastomosis develop[9]. The etiology of this vascular abnormality remains unclear. Opinon 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 majoraty of cases the cause is still inapparent[2,6,7].

Cerebral AVMs are developmental lesion of the cerebral circulation consisting of abnormal dilated tortous 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 ectasias 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[14,22].

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 symtoms 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 transdural communicating arteries[1,4,5,8,13-19].

At present, the pathologenesis 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[13,15].

Many authors have reported that Moyamoya disease might initiate the formation of AVMs[1,5]. Lichtor et al.[9] 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.[1] observed an AVM that enlarged after encephalomyosynangiosis because of new feeders from the external carotid artery. Halatsch et al.[5] 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.[17] 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.[13] 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.[14] 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 extremly 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 perfomed, 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 situation, such that the revascularization procecures requried to treat the moyamoya disease might exacerbate the cerebral AVMs. In our case, the treatment was primary symtomatic 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

1. Akiyama K, Minakawa T, Tsugi Y, Isayama K : Arteriovenous malformation assiciated with moyamoya disease : case report. **Surg Neurol** 41 : 468-471, 1994
2. Debrun G, Sauvegin J, Aireardi J : Moyamoya, a nonspecific radiological syndrome. **Neuroradiology** 8 : 241-244, 1975
3. Friedman P, Salazar J, Sugar O : Embolization and surgical excision of giant arterio-venous malformation. **Surg Neurol** 9 : 149-152, 1978
4. Fuse T, Takagi T, Fukushima T, Hashimoto N, Yamada K : Arteriovenous malformation associated with moyamoya disease. **Childs Nerv Syst** 12 : 404-408, 1996
5. Halatsch ME, Rustenbeck HH, Jansen J : Progression of arteriovenous malformation in moyamoya syndrome. **Acta Neurochir (Wien)** 139 : 82-85, 1997
6. Hardy RC, Williams RG : Moyamoya disease and cerebral hemorrhage. **Surg Neurol** 21 : 507-510, 1994
7. Hial SK, Soloman GE, Gold AP, Carter S : Primary cerebral arterial occlusive disease in children Park II. Neurocutaneous syndromes.

Radiology 99 : 87-93, 1971
8. Kayama T, Suzuki S, Sakurai Y, Nagayama T, Ogawa A, Yoshimoto T : A case of moyamoya disease accompaned by an arteriovenous malformation. **Neurosurgery 18** : 465-468, 1986
9. Lichtor T, Mullan S : Arteriovenous malformation in moyamoya syndrome. **J Neurosurg 67** : 603-608, 1987
10. Luessenhop A, Presper J : Moyamoya disease associated with aneurysm. **J Neurosurg 48** : 565-569, 1978
11. Luessenhop A, Rosa L : Cerebral arteriovenous malformations. **J Neruosurg 60** : 12-22, 1984
12. Maki Y, Enomoto T : Moyamoya disease. **Childs Nerv Syst 4** : 204-212, 1988
13. Mawad ME, Hilal SK, Michesen WJ, Stein B, Ganti SR : Occlusive vascular disease associated with cerebral arteriovenous malformation. **Radiology 153** : 401-408, 1984
14. Montanera W, Marotta TR, Terbrugge K, Lasjaunias P, Willinsky R, Wallace MC : Cerebral arteriovenous malformation associated with moyamoya phenomenon. **Am J Neruoradiol 11** : 1153-1156, 1990
15. Okada T, Kida Y, Kinomoto T, Sakurai T, Kobayashi T : Arteriovenous malformation associated with moyamoya disease : case report. **Neurol Med chir (Tokyo) 30** : 945-948, 1990
16. Parkinson d, Bachers G : Arteriovenous malformations. **J Neurosurg 53** : 285-299, 1980
17. Schmit BP, Burrows PE, Kuban K, Goumnerrova L, Scott RM : Acquired cerebral arteriovenous malformation in a child with moyamoya disease : case reprot. **J Neurosurg 84** : 677-680, 1995
18. Scott RM : Arteriovenous malformation and moyamoya disease. **Childs Nerv Syst 13** : 357, 1997
19. Seol HJ, Kim DG, Oh CW, Han DJ : Radiosurgical treatment of a cerebral arteriovenous malformation in a patient with moyamoya disease : case report. **Neurosurgery 51** : 478-482, 2002
20. Suzuki J, Kodama N : Moyamoya disease-a review. **Stroke 14** : 104-109, 1983
21. Suzuki J, Takaku A : Cerebrovascular moyamoya, Disease showing abnormal net-like vessels in base of brain. **Arch Neurol 20** : 288-299, 1969
22. Willinsky R, Lasjaunias P, Terbrugge K, Pruvost P : Brain arteriovenous malformation (BAVMS) : analysis of the angioarchitecture in relationship to hemorrhage. **J Neuroradiol 15** : 225-237, 1988
23. Yoon YS, Kim TS, Kim KM, Rhee BA, Kim GK, Leem W : Moyamoya Disease associated with Aneurysm. **J Korean Neurosurg Soc 13** : 765-772, 1984