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Case Report

Non-aneurysmal subarachnoid hemorrhage as presentation of moyamoya disease in an adult

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Abstract

Background: The presentation of moyamoya disease (MMD) as an aneurysmal subarachnoid hemorrhage (SAH) is relatively frequent and in the absence of aneurysms is extremely rare.

Case Description: A 53-year-old male patient suddenly developed severe headache associated with dysarthria and an altered state of consciousness. At the time of admission, he was found drowsy with global aphasia, stiff neck, right hemiparesis and right Babinski's sign. A non-contrast brain computed tomography was performed and a small bleeding in the subarachnoid space over the left frontal and parietal cortex was observed. Four-vessel cerebral angiography showed bilateral stenosis of the internal carotid arteries, with multiple tortuous vessels branching from the anterior and middle cerebral arteries. These abnormal vessels were anastomosing with branches from the posterior cerebral and middle meningeal arteries. With this information, a diagnosis of MMD was made. A three-dimensional reconstruction from digital angiography ruled out aneurysms or vascular malformations. After 4 weeks, another angiography was performed and remained the same as previous one.

Conclusion: Clinical and radiological characteristics of this case are consistent with previous reports, supporting the theory that non-aneurysmal SAH in MMD is caused by rupture of fragile moyamoya vessels.

Key Words: Moyamoya, subarachnoid hemorrhage, vascular anastomosis

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INTRODUCTION

Moyamoya disease (MMD), meaning cloud of smoke in Japanese, is a rare vascular disorder of uncertain etiology characterized by a gradual stenosis of supraclinoid segment of internal carotid arteries and their proximal branches. Its incidence is higher in eastern countries, especially in the east of Asia, including Japan and Korea where its prevalence is found to be 3.16 per 100,000 habitants. It is less common in western countries. In South America, it is very rare, making its incidence difficult to determine.[5]

This disease has its highest incidence in two age groups: around 5 years and from 40 to 50 years of age. MMD in

adults presents with transient ischemic attacks, strokes and/or intracranial bleeding. Moyamoya vessels are fragile, dilated and tortuous, with a high risk of rupture. Intraparenchymal and intraventricular hemorrhages are frequently related to this disease and are attributed to these vascular abnormalities. Bleeding is also common in the subarachnoid space as a result of intracranial aneurysm rupture.

Subarachnoid hemorrhage (SAH) in patients without cerebral aneurysms presenting as MMD is extremely rare. In this report is presented a 53-year-old-man who developed bleeding in the subarachnoid space over the left frontal and parietal cortex, with the characteristic angiographic findings of this disease, but in the absence of aneurysms or arteriovenous malformations.

CASE REPORT

A 53-year-old male patient, without relevant personal or family history, was admitted to the emergency room after he suddenly developed severe headache with dysarthria, altered state of consciousness and syncope. At the time of admission, he was found drowsy with global aphasia, stiff neck, right hemiparesis and right Babinski's sign. Kernig's sign was present too. Fundoscopy did not show retinal hemorrhages. Autoimmune diseases, brain tumors, actinic encephalopathy, Von Recklinghausen's disease and Down syndrome were clinically ruled out.

A non-contrast brain computed tomography revealed bleeding in the subarachnoid space over the left frontal and parietal cortex [Figure 1]. Four-vessel cerebral angiography showed bilateral stenosis of the supraclinoid segment of both internal carotid arteries and multiple tortuous vessels branching from the anterior and middle cerebral arteries (moyamoya vessels). These abnormal vessels were anastomosing with branches from the posterior cerebral and middle meningeal arteries. With this information, MMD diagnosis was made. An extensive search for aneurysms and vascular malformations was done, including a three-dimensional reconstruction from each projection of the four-vessel angiography, with no other abnormalities found [Figure 2]. Taking into account the possibility of an angiographically occult aneurysm, digital subtraction angiography was repeated after 4 weeks and remained without changes from the previous.

During hospitalization, the patient developed a massive pulmonary embolism and subsequently died.

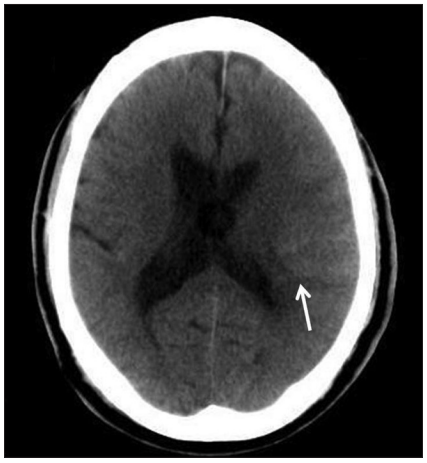

Figure 1: Non-contrast brain computed tomography showing subarachnoid hemorrhage over the left frontal and parietal cortex (white arrow)

DISCUSSION

The main clinical presentations of MMD substantially differ between children and adults. Most children develop transient ischemic attacks or cerebral infarctions, whereas about half of the adult patients present with intracranial hemorrhage. Regarding the ethnic differences, clinical features and the course of MMD in Caucasians clearly differ from those of Asians in the timing of the onset of vasculopathy and rates of hemorrhage, where clearly Asians have a higher prevalence of the latter.[1,9] In the Cooperative Study of the Korean Society for Cerebrovascular Disease that included 334 patients, the results showed that bleeding manifestations occurred in 43% and 62.4% of pediatric patients and adults, respectively.[3]

The most frequent sites of intracranial hemorrhage are the ventricles and cerebral parenchyma, the latter more commonly in the basal ganglia. Two major causes of

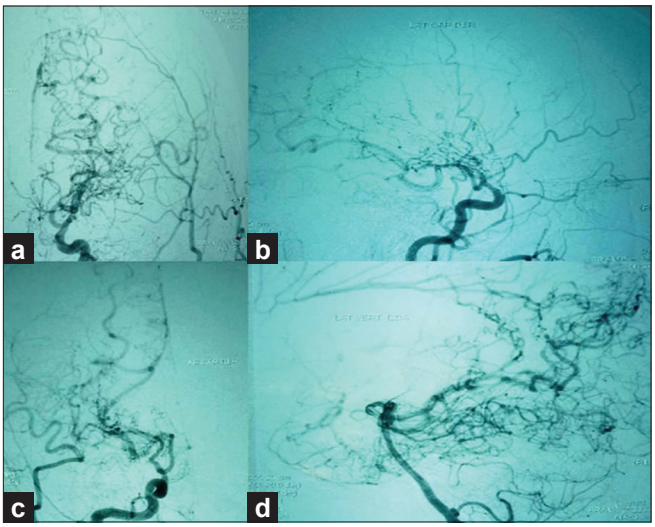

Figure 2: Four-vessel cerebral angiography showing bilateral stenosis of the supraclinoid segment of both internal carotid arteries and tortuous branches from the anterior and middle cerebral artery. These abnormal vessels were found anastomosing with branches from the posterior cerebral and middle meningeal arteries. (a and b) Left and right carotid arteries (antero-posterior view); (c) right carotid artery (lateral view); (d) left vertebral artery (lateral view)

Table 1: Reported cases of non-aneurysmal SAH in patients with moyamoya disease

Author	Gender	Age (years)	Site of bleeding	Associated condition
This report	Male	53	Left frontal and parietal cortex	None
Osanai *et al*.[8]	Female	34	Left frontal cortex	None
Dietrichs *et al*.[2]	Female	21	Left frontal and parietal cortex and left Sylvian fissure	Post-partum
Matsumoto *et al*.[7]	Female	32	Right inter-hemispheric parietal cortex	Post-partum and renal artery stenosis
Marushima *et al*.[6]	Female	38	Left frontal cortex	None
Sönmez *et al*.[10]	Male	31	Left frontal and parietal cortex and left Sylvian fissure	Heroin intoxication

intracranial bleeding in MMD include the rupture of dilated and fragile moyamoya vessels, and of saccular aneurysms in the circle of Willis. The hemorrhage due to persistent hemodynamic stress on moyamoya vessels occurs in the basal ganglia, thalamus, and periventricular region. These vessels and the saccular aneurysms are usually identified on digital subtraction angiography.

The second most frequent cause of intracranial hemorrhage is rupture of saccular aneurysms around the circle of Willis. This most commonly occurs at the basilar artery bifurcation or at the junction of the basilar artery and the superior cerebellar artery. The vertebrobasilar system has a very important role in providing collateral circulation in MMD. Thus, hemodynamic stress probably contributes to the development of saccular aneurysms in this area and their rupture can result in SAH. A third cause of intracranial bleeding in adult patients with MMD is rupture of the dilated collateral arteries on the brain surface, although rare, as in our case.[1]

The development of SAH in patients with MMD in the absence of cerebral aneurysms is extremely rare, and to our knowledge, only five cases have been previously reported.[2,4,6-8,10] Table 1 summarizes the clinical data of the previous cases.

There are certain relevant characteristics of all these cases. All correspond to adult patients, which reinforces the theory that these have been caused by rupture of fragile collateral vessels developed gradually through childhood and adolescence.[8] Pathologic examination of moyamoya vessels shows fragmentation of the internal elastic lamina, attenuation of the media layer and dilation of the arterial wall. Other findings include frequently associated mural thrombi and occasional lipid deposits, although inflammatory cells are conspicuously absent. Accumulating evidence suggests that smooth muscle cell proliferation and phenotypic modulation may underlie these changes.[11]

In the previously reported cases, the involvement of the left frontal cortex has been especially prevalent, suggesting that rupture of collateral vessels branching from the middle cerebral artery could be the cause. There still are no theories that explain the predilection for the left side. Transdural anastomotic vessels often pass from the meningeal arteries to the middle cerebral artery through the subdural and subarachnoid space with little support. Thus, they could be easily damaged in either direct or indirect minor head trauma.[10] Marushima *et al*. described a case of non-aneurysmal SAH in which the bleeding site was coincident with the presence of a large transdural anastomosis adjacent to the frontal lobe, reinforcing the speculation that recognition of these abnormal vessels may have diagnostic and thus therapeutic implications in this group of patients.[6]

Two previous cases of non-aneurysmal SAH as presentation of MMD have occurred during the post-partum period. It has been suggested that during this period and pregnancy, the incidence of hemorrhagic cerebrovascular events is more frequent even as initial presentation of this disease.[1] Other studies have not found evidence that pregnant women with MMD have increased risk of ischemic or hemorrhagic stroke, especially in non-Asian individuals.[1]

In the case presented here, age and location of bleeding is consistent with previous reports. This supports the theory about the development of SAH in patients with MMD even in the absence of cerebral aneurysms.

CONCLUSION

Presentation of MMD as SAH in the absence of intracranial aneurysms is an extremely rare event and it is believed to be caused by rupture of fragile collateral vessels formed through childhood and adolescence. Clinical and radiological characteristics of the presented case are consistent with previous reports, supporting the theory that non-aneurysmal SAH in MMD is caused by rupture of fragile transdural anastomotic vessels in their course through the subarachnoid space.

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