CASE REPORT

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Intracerebral hemorrhage and ischaemic stroke due to Moyamoya disease in young patients: a case report

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ABSTRACT

Background: Moyamoya disease (MMD) is a non-atherosclerotic cerebrovascular disorder characterized by progressive stenosis of the distal part of the internal carotid artery (ICA), thus forming an abnormal vascular network in the base of the brain. MMD is often found in women. The common symptoms of MMD are migraine, epilepsy, recurrent transient ischemic attack (TIA), ischemic stroke, and intracerebral hemorrhage. Stroke at young onset has challenges in looking for risk factors and etiology. A detailed examination is needed to prevent recurrent strokes.

Case Presentation: Male 17-year-old complained of a throbbing headache on the right side for 5 months, with a numeric rating scale (NRS) 4. The headache duration was 24-72 hours, with a frequency of 1-2 times a month. Complaints are exacerbated by activity and improve when taking over medication. 2 months after the first onset, he felt severe headache with NRS 7, accompanied by weakness and tingling on the left side. All his laboratory results were within normal limits. Cerebral Digital Subtraction Angiography (DSA) results showed severe stenosis at the base of the right ICA bifurcation segment accompanied by slow flow at the right middle cerebral artery (MCA) and severe stenosis at the base of the left ICA accompanied by cross-filling to the contralateral anterior cerebral artery (ACA). Cortico-cortical collaterals were obtained from the posterior cerebral artery (PCA) branch covering the MCA area. The patient was diagnosed with MMD.

Conclusion: MMD is a rare cerebrovascular disease that can be a risk factor for stroke at a young age. Diagnosis and evaluation of the collateral system need to be carried out through cerebral angiography so that the selection and consideration of appropriate management of recurrent stroke attacks can be carried out.

Keywords: intracerebral hemorrhage, ischaemic stroke, moyamoya disease, young age.
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INTRODUCTION

Moyamoya disease (MMD) is a non-atherosclerotic cerebrovascular disorder characterized by progressive stenosis of the distal part of the internal carotid artery (ICA), thus forming an abnormal vascular network in the base of the brain.[1,2] The incidence rate of MMD is increasing, with a prevalence of 10.5 cases per 100,000 population. The ratio of women to men reached 1.8:1, with the peak prevalence in female patients aged 10-14 years and 20-24 years in male patients.[2] The exact pathogenesis of MMD is still unknown, but the latest study has revealed that genetic variables play a significant role in the development of MMD. Ring finger protein (RNF) 213 gene was recently discovered to be a gene susceptible to MMD. Circulating endothelial/smooth-muscle progenitor cells (SPCs), cytokines, and growth factors were also found to be linked with the pathogenesis of MMD.[1]

Stroke at young onset has its challenges in looking for risk factors and etiology. A detailed examination is needed to prevent recurrent strokes. A structural vascular abnormality should be considered in young stroke patients, as MMD can cause stroke in young patients. Symptoms often encountered in MMD are ischaemic stroke, migraine, epilepsy, recurrent TIA, and intracerebral hemorrhage.[1] The diagnosis is made based on angiography. The development of MMD is estimated to be progressive over 5 years. The outcome is poor without treatment. Current therapy options include symptomatic medications, surgical and medical procedures for preventing strokes. Treatment choices are now made depending on the experience of surgeons and clinicians and the resources available at the medical facility because there are no randomized trials or recommendations.[3] Moyamoya disease was often overlooked, as the study found that 62% of MMD patients were initially misdiagnosed. Therefore, it is important to note the necessity of performing vascular imaging in cases of a stroke at a young age to prevent further delay in diagnosis and treatment.

Based on those mentioned above, this case report aims to show that stroke at a young age may be caused by structural abnormalities of blood vessels, such as MMD.

CASE REPORT

A seventeen-year-old male complained of throbbing headaches on the right side for

5 months with NRS 4. The duration of one attack lasted 24-72 hours, with a frequency of 1-2 times a month. The headache was exacerbated by activity and improved when he took medication. Two months after the first headache onset, he felt a severe headache (NRS 7) accompanied by abrupt weakness and a tingling sensation on the left side of his body. The patient was found to have a risk factor, such as smoking 1-2 cigarettes/day. Examination of vital signs and general status were within normal limits. Neurological examination revealed upper motor neuron (UMN) type left facial and lingual palsy, left hemiparesis and left hemi-hypoesthesia. His routine laboratory results were unremarkable. We also screened for vascular risk factors, including blood sugar, lipid profile, uric acid, and fibrinogen, which were all within normal limits.

Head MRI show hyperintense lesion on sequences T1, T2 and DWI in the area of the corona radiata to the right thalamus, which did not show contrast enhancement, suggesting a bleeding process with a volume of 0.89 cc (Figure 1). In addition, there was hypointense on T1 and hyperintense on T2 in the cortical area of the right frontal lobe, suggesting a cerebral infarction.

Cerebral Digital Subtraction Angiography (DSA) results showed severe stenosis at the base of the right ICA bifurcation segment accompanied by slow flow at the Middle Cerebral Artery (MCA), severe stenosis at the base of the left ICA accompanied by cross-filling to the contralateral Anterior Cerebral Artery (ACA) (Figure 2 and 3). Cortico-cortical collaterals were obtained from Posterior Cerebral Artery (PCA) branches: Perisplenial Artery, Parietooccipital Artery, Posterior Temporal Artery, Medial Temporal Artery, and Anterior Temporal Artery covering the MCA area.

DISCUSSION

MMD symptoms present with various cerebrovascular events, including intracerebral hemorrhage, TIA, cerebral infarction, and epilepsy. While pediatric MMD mostly correlates with ischemic lesions, adult MMD may have both hemorrhagic and transient or permanent ischemia. Research in Japan mentioned

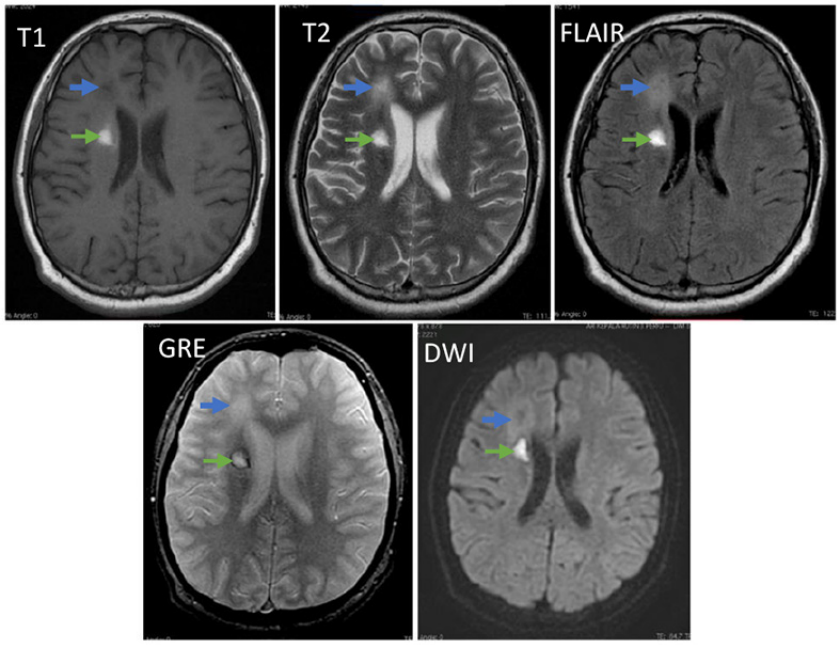

Figure 1. Head MRI results show bleeding in the corona radiata area to the right thalamus (green arrow) and cerebral infarction in the right frontal lobe cortex-subcortex area (blue arrow).

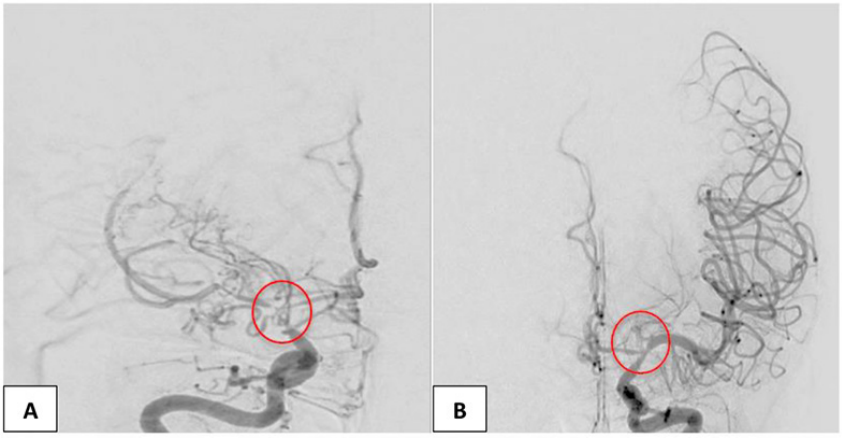

Figure 2. (A) Severe stenosis of the distal right ICA bifurcation segment accompanied by slow flow in the MCA (anteroposterior view). (B) Severe stenosis of left ICA base accompanied by cross-filling to the Contralateral ACA (anteroposterior view)

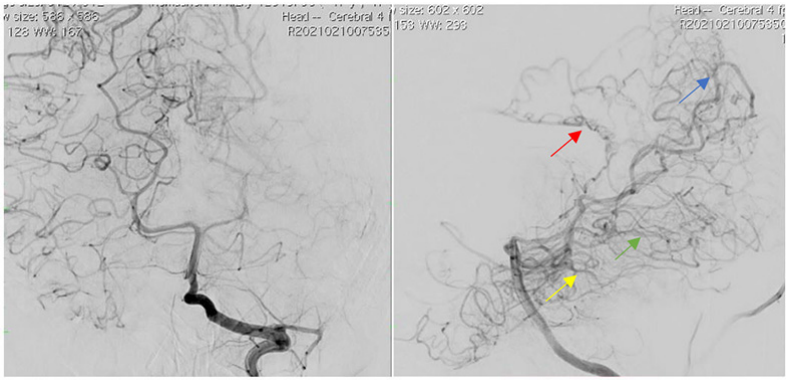

Figure 3. Obtained cortico-cortical collaterals from PCA branches: perisplenial artery (red arrow), parieto-occipital artery (blue arrow), posterior temporal artery (green arrow), middle temporal artery (yellow arrow) covering the ACA and MCA area.

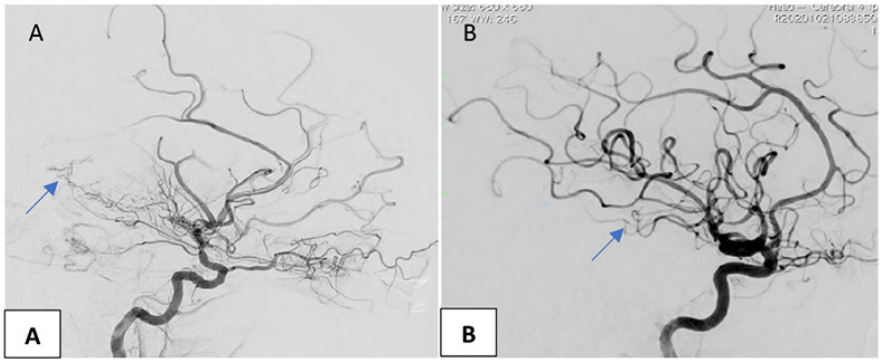

Figure 4. (A) The choroidal anastomosis characterized by dilatation and extension of the anterior choroidal artery. (B) View of the anterior choroidal artery in a normal patient.

4 symptoms of MMD that are often encountered: cerebral ischaemic 63.4%, cerebral hemorrhage 21.6%, epilepsy 7.6%, and other symptoms 7.5%.[4] Currently, there is insufficient data on the prevalence of MMD with ischaemic stroke and cerebral hemorrhage. In MMD patients with intracerebral hemorrhage, bleeding is usually found in the anterior lateral ventricles, basal ganglia, and thalamus. It is caused by the rupture of abnormally thin-walled collateral vessels. The three most common sources of bleeding in MMD are the lenticulostriate arteries, the thalamic arteries and the choroidal arteries.[4–6]

The diagnosis of MMD is based on the presence of (1) stenosis of the Distal ICA or proximal to the MCA or ACA, (2) abnormal vascular tissue (Moyamoya vessels) around the stenotic lesion, and (3) bilateral lesions. Cerebral DSA is the gold standard in MMD cases.[7]

The reduced flow in the major vessels results in compensatory collateral vessel formation by the small vessels. A deep collateral system was found in this patient, characterized by an extension of the anterior choroidal artery.[5-8] The deep collateral system in MMD is divided into 3 types: choroidal anastomosis, thalamic anastomosis and lenticulostriate anastomosis. The choroidal anastomosis is characterized by dilatation and extension of the choroidal artery, with features moving away from the lateral ventricular location to form collaterals with the medullary arteries. Another indicator is the extension of the anterior or posterior lateral choroidal arteries beyond the atria of the lateral ventricles to reach the bodies of the lateral ventricles. Anastomosis is considered positive when the artery is connected to the pericallosal artery by penetrating the corpus callosum. The thalamic anastomosis is an anastomosis between the thalamic perforator originating from the posterior communicating artery and the posterior cerebral artery with the medial end of the medullar or insular arteries.[6-8] On angiographic examination, it is characterized by dilatation and extension of the thalamic perforator extending through the posterior medial choroidal artery. Moyamoya disease patients with lenticulostriate anastomosis are characterized by a connection between the lenticulostriate artery and the medial end of the medullar artery. On angiography, the lenticulostriate artery dilates and extends so that the artery is parallel to the pericallosal artery when viewed from the lateral side. When viewed from an anteroposterior angle, a long lenticulostriate artery is visible, curving laterally with a sharp inflection at the periphery.[5]

Other than deep collaterals, we also found a cortico-cortical collateral system in this patient. A cortico-cortical network was seen, with feeding vessels from a splenial artery, parietooccipital PCA branches, and temporal PCA branches. The splenial artery runs through retrosplenial to anastomose with pericallosal artery, while temporal PCA branches course via medial and convex parietooccipital hemispheric surface to end up in ACA distal cortical branches, and temporal PCA branches run through the inferior temporal hemispheric surface with MCA distal cortical branches as its recipient's vessels.[8]

The radiographic feature of MMD resulted from the abnormal vascular tissue, which gives the characteristic "puff of smoke" appearance due to the formation of a collateral system of small blood vessels around the stenotic lesion area. This "puff of smoke" appearance was mostly found in MMD with ICA stenosis, accompanied by a steno-occlusive PCA lesion on the ipsilateral side of the lesion. In MMD cases without ipsilateral PCA steno-occlusive lesions, the "puff of smoke" appearance was not dominant.[7]

Suzuki et al. in Fujimura M et al. classified MMD into 6 stages. Stage 1: stenosis of the distal part of the ICA, Stage 2: visible moyamoya blood vessels, stenosis of the proximal part of ACA and MCA, Stage 3: clearly visible moyamoya blood vessels, MCA and ACA of the proximal part are not visible, while the distal part appears as a branch of PCA. Stage 4: the moyamoya blood vessels begin to disappear, accompanied by stenosis in the proximal PCA; stage 5: the moyamoya blood vessels increasingly disappear with the major intracranial arteries disappearing, and Stage 6: the moyamoya blood vessels disappear, circulation only comes from the External Carotid Artery (ECA).[1] The patient reported, in this case, was diagnosed with MMD Suzuki stage 2 because he had ICA stenosis, proximal ACA and MCA stenosis, and abnormal vascular tissue formation in the choroidal arteries. Kang S et al. showed that intracerebral hemorrhage related to MMD was mostly found in stages 3 and 4 according to Suzuki's classification, but there was no statistically significant relationship between the MMD stage and the incidence of intracerebral hemorrhage.[9] Latest angiographic study demonstrated a specific angioarchitecture linked to higher re-bleeding risk, including extension and dilatation of choroidal collateral and posterior cerebral artery involvement. However, the exact process underlying the hemorrhagic appearance in MMD is unknown.[10-14]

Management of MMD depends on the aggressive course of the disease. Cases with mild symptoms will be treated with conservative therapy, while cases with severe symptoms are usually treated with revascularization procedures.

Conservative therapy aims to pass the acute phase of stroke due to MMD and prevent stroke recurrence. Several types of MMD treatment, such as aspirin, steroids, antibiotics, mannitol, verapamil, nimodipine, and nicardipine, have been studied. However, these therapies are still considered ineffective for preventing stroke recurrence in Moyamoya disease. Cerebral revascularization can be performed to avoid stroke recurrence. Some of the indications for cerebral revascularization include an infarct stroke with a maximum size of 2 cm presented in computed tomography or a hemorrhagic stroke after full resolution of the hematoma. Revascularization measures can be carried out 2 months after the acute phase of the last attack has passed.[15,16] In this patient, conservative therapy was carried out to pass through the acute phase of stroke by giving symptomatic analgesic therapy for the headache. Cerebral revascularization was not carried out because the patient still had sufficient collaterals originating from the PCA and anterior choroidal artery.

This case report concludes that Moyamoya disease is a rare disease that causes stroke at a young age. This case report presented ischaemic and hemorrhagic strokes with MMD features at a young age. There is insufficient data on MMD with ischaemic stroke and cerebral hemorrhage. Diagnosis and evaluation of the collateral system need to be carried out through cerebral angiography so that the selection and consideration of appropriate management of recurrent stroke attacks can be carried out. Periodic monitoring of intracranial vascular structures is necessary to assess the progress of MMD. As this report is only a single case study, this may not be able to represent MMD as a whole. Future case series of MMD with different features in terms of radiologic appearances, stages, and collateral systems may be able to give a wider and deeper knowledge of this disease.

CONCLUSION

MMD is a rare cerebrovascular disease that can be a risk factor for stroke at a young age. Diagnosis and evaluation of the collateral system need to be carried out through cerebral angiography so that the selection and consideration of appropriate management of recurrent stroke attacks can be carried out.

CONFLICT OF INTEREST

No conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper.

ETHICS CONSIDERATION

Written informed consent was obtained from the patient/legal representative to publish this case report (including all data and images). The patient/legal representative understands that his/her name and initial will not be published, and due efforts will be made to conceal the patient's identity.

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AUTHOR CONTRIBUTIONS

RDAW conceived the idea and collected and analyzed the data. All authors were involved in the drafting and review of the manuscript and approved the final manuscript before submission.

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