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Case Report

Acute Ischemic Stroke in Moyamoya Disease: A Case Report

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ABSTRACT

Introduction: Moyamoya disease (MMD) is also known as spontaneous occlusion of the circle of Willis, is a chronic, occlusive cerebrovascular disease with an unknown etiology.[1] It is characterized by progressive stenosis or occlusions of the intracranial internal carotid artery (IAC) and or the proximal portion of the anterior cerebral artery (ACA) and middle cerebri artery (MCA).[1,2] This steno-occlusive pattern is associated with a compensatory development of a collateral network of vessels at the base of the brain, appearing as a "puff of smoke" on conventional angiography ("moyamoya" in Japanese).[1,2] The clinical presentations of MMD include transient ischemic attacks, ischemic strokes, hemorrhagic strokes, seizure, headache, and cognitive impairment.[1,2,3]

Case Report: This case study presents a 48-year-old male with symptoms hemiparesis sinistra. Brain CT scan result was subacute infarct at right anterior cerebri artery territory. Digital Substract Angiography (DSA) result was occlusion in the right media cerebri artery (MCA) and right anterior cerebri artery (ACA), severe stenosis right verterbral artery until basilar artery, moyamoya vessels at anterior and posterior circulation.

Keywords: Moyamoya disease; ischemic stroke; occlusive cerebrovascular diease

INTRODUCTION

Moyamoya disease (MMD) is uncommon condition with substantial morbidity and mortality.[1,2] MMD is a cerebrovascular steno-occlusive condition characterized by progressive stenosis of the terminal portion of the internal carotid artery and the formation of the abnormal network of dilated, fragile perforators at the base of the brain.[1,2] The most recent the Research Committee on Moyamoya Disease (RCMD) guidelines from 2021 presented important new accords regarding the locations and magnitude of involvement of the necessary for the definition of MMD, which is the steno-occlusive involvement of the arteries centered on the terminal portion of the intracranial carotid artery in the absence of the other causes that can produce arterial stenosis/occlusion.[2]

When a patient meets the diagnostic criteria for MMD but has other comorbidities that are associated with the vasculopathy, the condition is designated moyamoya syndrome (MMS).[2] Disorders associated with MMS are autoimmune, meningitis, brain tumor, down syndrome, neurofibromatosis type 1, head irradiation, sickle cell disease.[2]

Much of the epidemiological data on MMD come from Asia, where the disease is most prevalent.[2] In 2 large studies of MMD from Japan, the women-to-men ration was up to 1.9:1, and 11% to 12% had family history of MMD suggesting a strong genetic component.[2] Although, overall, there was a bimodal age distribution (with peaks around age 10 and 40).[2]

MMD and MMS may present with a broad variety of clinical symptoms.[2] Presenting symptoms and "events" can be grouped into (1) ischemic and hemorrhagic events, (2) other neurological manifestations, and (3) symptoms of associated diseases, in the case of MMS.[2] Ischemia typically involves the anterior circulation, with the border zone areas most commonly affected.[2] Hemorrhage most commonly presents with intracranial or intraventricular patterns.[2]

Digital Subtraction Angiography is considered the gold standard for the diagnosis of moyamoya vasculopathy.[2]

The control of additional cerebrovascular stroke risk factors is important in the medical management of MMD. Diabetes, hypertension, dyslipidemia, increased body mass index and homocysteine might also be associated with a higher risk of MMD.[2]

Patients with symptomatic MMD should be referred for consideration of revascularization therapy.[2] There is sufficient evidence and class 2a recommendation from the AHA/American Stroke Association Guidelines for surgical revascularization in adult patients with symptomatic MMD.[2]

CASE REPORT

A 48-year-old male presented to the emergency unit at Santosa Central Bandung with symptoms of hemiparesis sinistra for 4 days ago. There were no seizure and headache. He had history of hypertension. There were no comorbidities such as diabetes mellitus, cardiac problem, head trauma, autoimmune diseases, nor history of familial disorders.

During the physical examination, the patient exhibited a Glascow Coma Scale (GCS) score of 15 E4M6V5. and hypertension (180/110 mmHg). Neurological examination revealed paresis N VII and XII central sinistra, hemiparesis sinistra with a motor strength level score 1 (maximum score 5) for upper extremity and score 0 for lower extremity.

Laboratory investigation revealed dyslipidaemia (direct LDL-Cholesterol 141 mg/dl), hyperuricemia (uric acid 7.5 mg/dl). Chest X ray was normal.

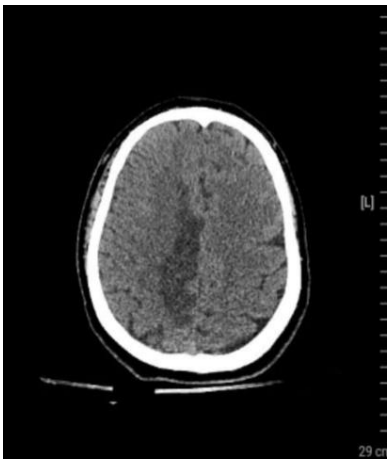

Figure.1. Brain CT scan: subacute infarct at right anterior cerebri artery territory

Electrocardiography was sinus rhythm. CT scan result was subacute infarct at right anterior cerebri artery territory (figure 1). DSA result was occlusion in the right media cerebri artery (MCA), right anterior cerebri artery (ACA), severe stenosis right vertebral artery until basilar artery (figure 2a,2b), moyamoya vessels at anterior and posterior circulation (figure 3).

The patient was treated for five days with oral administration amlodipine 5 mg, acetylsalicylic acid 80 mg, rosuvastatin 40 mg, allopurinol 100 mg. Citicoline 1 gr intravenous twice a day. And consulted a medical rehabilitation doctor for physical rehabilitation program.

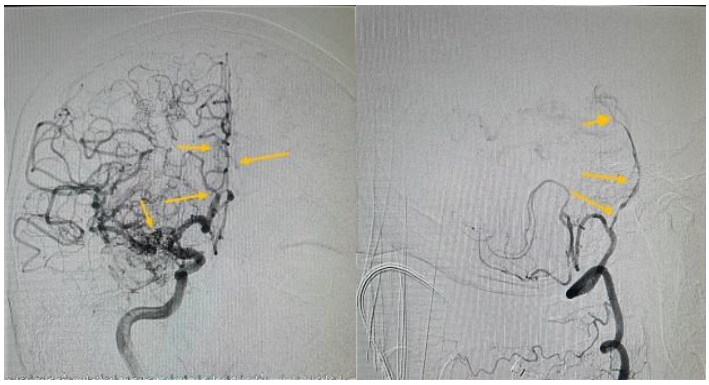

Figure 2 A Figure 2 B

Figure 2a. DSA: steno-occlusive right media cerebri artery (MCA) and right anterior cerebri artery (ACA) (AP position) **Figure 2b.** DSA: steno-occlusive right vertebrobasilar artery (lateral position)

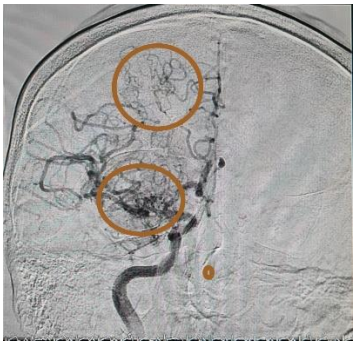

Figure 3. DSA: moyamoya vessels at anterior and posterior circulation

DISCUSSION

The diagnosis MMD in this patient is established based on the present of hemiparesis sinistra in adult (48 age year) without history of other comorbidities vasculopathy. Clinical manifestation and feature in this patient where ischemic stroke with brain CT scan result was subacute infarct at right anterior cerebri artery territory. In MMD ischemia typically involves the anterior circulation, with the border zone area most commonly affected.[2] DSA result was occlusion in the right media cerebri artery (MCA) and right anterior cerebri artery (ACA) , severe stenosis right vertebral artery until basilar artery and moyamoya vessels at anterior and posterior circulation. Based on Fujimura,2022, DSA required findings 1) stenosis or occlusion in the arteries centered on the terminal portion of the intracranial internal carotid artery 2) moyamoya vessels (abnormal vascular network) in the vicinity of the occlusive or stenotic lesions in the arterial phase.[2]

Hypertension and dyslipidaemia in this patient were additional cerebrovascular stroke risk factors.[2] The control of additional cerebrovascular stroke risk factors is important in the medical management of MMD.[2] Antiplatelet use for prevention of ischemic events in surgical and nonsurgical patients with ischemic moyamoya vasculopathy may reasonable, per the AHA 2021 guideline.[2]

Several significant limitations should be considered. There were not laboratories result to exclude comorbidities vasculopathy.

CONCLUSION

MMD is a rare condition that is often under reported in Indonesia. It is crucial to investigate and evaluate for MMD in adult patients presenting ischemic stroke.

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