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Internal Medicine

MOYA MOYA DISEASE

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ABSTRACT **Introduction:** Moyamoya disease (MMD) is a rare, chronic cerebrovascular disorder characterized by progressive stenosis or occlusion of the intracranial internal carotid arteries and their proximal branches, leading to the development of fragile collateral vessels that produce the classic "puff of smoke" appearance on angiography. The disease may present with ischemic or hemorrhagic manifestations and requires timely diagnosis and revascularization to prevent neurological deterioration. **Case Summary:** We report a case of a 22-year-old male presenting with progressive neurological symptoms including headache, left upper limb weakness, facial deviation, and cognitive disturbances. Neuroimaging revealed multiple chronic infarcts with extensive collateral formation suggestive of moyamoya disease. Digital subtraction angiography confirmed Suzuki Grade IV moyamoya disease. The patient underwent right encephalo-duro-arterio-myo-synangiosis (EDAMS) for cerebral revascularization. Postoperative recovery was uneventful with improvement in neurological symptoms and stable hemodynamic status. **Conclusion:** Early diagnosis and timely surgical revascularization remain crucial in the management of moyamoya disease to prevent recurrent ischemic events. EDAMS provides effective indirect revascularization and can significantly improve cerebral perfusion in symptomatic patients.

KEYWORDS :

INTRODUCTION

Moyamoya disease is a progressive occlusive cerebrovascular disorder involving the terminal portion of the internal carotid arteries (ICA) and the proximal segments of the anterior cerebral artery (ACA) and middle cerebral artery (MCA). Progressive stenosis leads to the development of abnormal collateral vascular networks at the base of the brain, which appear as a "puff of smoke" (moyamoya in Japanese) on angiographic imaging.

The disease was first described by Suzuki and Takaku in 1969, who also proposed the angiographic staging system used today. Moyamoya disease is more prevalent in East Asian populations but has been increasingly recognized worldwide. Clinical manifestations include transient ischemic attacks, ischemic stroke, seizures, headaches, and cognitive impairment.

Definitive diagnosis relies on neuroimaging techniques such as MRI, MR angiography, and digital subtraction angiography (DSA). Treatment is primarily surgical, focusing on revascularization procedures that restore cerebral blood flow and reduce the risk of future ischemic events. Indirect bypass procedures such as encephalo-duro-arterio-myo-synangiosis (EDAMS) promote neovascularization and improve cerebral perfusion over time.

Case Report

A 22-year-old male presented with complaints of holocranial headache for two months, predominantly on the left side. The headache was intermittent and aggravated by stress and lack of sleep but relieved with medication. He also reported episodes of irrelevant speech, reduced concentration, and occasional slow responses.

The patient developed weakness of the left upper limb for two months, with difficulty holding objects and slowness of movements. Two days prior to presentation, he noticed deviation of the angle of the mouth to the right side. There was no history of seizures, vomiting, blurring of vision, double vision, or bowel and bladder disturbances.

Past medical history was unremarkable for diabetes, hypertension, or coronary artery disease. He had no history of smoking or alcohol consumption. There was significant family history of Moya Moya disease in his mother.

On examination, the patient was alert and oriented (GCS E4 V5 M6). Vital signs were stable. Cranial nerve examination revealed left upper motor neuron facial palsy. Extraocular movements were full and pupils were reactive to light. Motor examination showed reduced power in the left upper limb (4+/5) while the remaining limbs had normal power (5/5). Reflexes were normal and plantar responses were flexor. Sensory examination and gait were normal.

MRI brain demonstrated multiple chronic infarcts in bilateral deep border-zone regions, right parietal region, and right anterior temporal region with extensive collateral formation.

Perfusion imaging showed delayed cerebral perfusion and collateral circulation. MR angiography demonstrated abrupt stenosis of the supraclinoid internal carotid arteries with absence of bilateral MCA and A1 segments, consistent with moyamoya disease.

Subsequently, cerebral digital subtraction angiography (DSA) was performed. Angiography revealed diffuse severe narrowing of the supraclinoid internal carotid arteries bilaterally with absence of antegrade filling of the ACA and MCA territories. Numerous collateral vessels were seen arising from perforators and skull base arteries, producing the characteristic "puff of smoke" appearance, confirming Suzuki Grade IV moyamoya disease.

Given the symptomatic disease with perfusion deficit, the patient was planned for surgical revascularization. He underwent right encephalo-duro-arterio-myo-synangiosis (EDAMS) under general anesthesia. The superficial temporal artery (STA) was identified and mobilized. A temporal craniotomy was performed, the dura was opened, and the STA along with temporalis muscle was placed in contact with the brain surface to promote indirect revascularization.

The procedure was uneventful. Postoperatively, the patient was managed with intravenous fluids, analgesics, antibiotics, and antiepileptic medications. Gradual mobilization was initiated, and the patient tolerated oral intake well. He remained hemodynamically stable with no new
neurological deficits.

At discharge, the patient was conscious, ambulant, afebrile, and tolerating a normal diet. He was advised to continue levetiracetam, aspirin, and atorvastatin along with adequate hydration, regular follow-up, and avoidance of strenuous activity. On follow-up review, the patient was clinically stable with no new complaints.

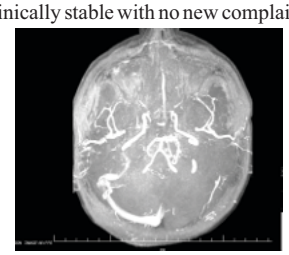

Fig 1: Stenosis of the Supraclinoid Internal Carotid Arteries with Absence of Bilateral MCA and A1 Segments

DISCUSSION

Moyamoya disease is a chronic, progressive cerebrovascular disorder characterized by stenosis or occlusion of the terminal internal carotid arteries and their proximal branches, particularly the anterior cerebral artery and middle cerebral artery. This results in the development of fragile collateral vessels at the base of the brain, producing the characteristic "puff of smoke" appearance on angiography, as first described by Suzuki and Takaku.[9]

The exact etiology remains unclear; however, current evidence suggests a multifactorial process involving genetic predisposition, endothelial dysfunction, and abnormal smooth muscle proliferation leading to intimal thickening and progressive luminal narrowing.[3] As cerebral blood flow decreases, compensatory collateral circulation develops via perforating arteries, leptomeningeal anastomoses, and transdural branches of the external carotid artery.[4] These collateral vessels, although compensatory, are fragile and often insufficient, predisposing patients to ischemic and hemorrhagic events.[6]patients to ischemic and hemorrhagic events.[6] Epidemiologically, moyamoya disease is more prevalent in East Asian populations but is increasingly recognized worldwide.[1] It demonstrates a bimodal age distribution, with peaks in childhood and early adulthood.[7] Adult patients more commonly present with ischemic symptoms or intracranial hemorrhage.[6] The clinical presentation in our patient, including headache, focal neurological deficit, and cognitive disturbance, is consistent with cerebral hypoperfusion.[5] Neuroimaging is central to diagnosis. MRI typically demonstrates chronic ischemic changes, particularly in watershed regions, while perfusion studies reveal impaired cerebral blood flow.[7] Magnetic resonance angiography and computed tomography angiography are useful screening tools; however, digital subtraction angiography remains the gold standard for definitive diagnosis and staging.[8] The Suzuki staging system classifies disease progression, with Stage IV characterized by reduction of moyamoya vessels and increasing reliance on external carotid artery collaterals, as seen in our patient.[9]

Medical therapy alone is insufficient in symptomatic patients, as it does not address the underlying hemodynamic compromise.[10] Surgical revascularization remains the cornerstone of management. Direct bypass procedures provide immediate augmentation of cerebral blood flow, whereas indirect techniques such as encephalo-duro-arterio-myo-synangiosis (EDAMS) promote gradual neovascularization.[11]

In the present case, EDAMS was performed, facilitating indirect revascularization by placing the superficial temporal artery and temporalis muscle in contact with the cortical surface. This promotes angiogenesis and improves cerebral perfusion over time. Surgical revascularization has been shown to significantly reduce recurrent ischemic events and improve long-term neurological outcomes.[12]

The favorable postoperative course in this patient underscores the importance of early diagnosis, appropriate imaging evaluation, and timely surgical intervention in improving prognosis and preventing disease progression in moyamoya disease

CONCLUSION

Moyamoya disease is a rare but potentially debilitating cerebrovascular disorder that can lead to recurrent ischemic events if not diagnosed and treated promptly. Early recognition through appropriate neuroimaging and angiographic evaluation is essential for accurate diagnosis.

Surgical revascularization procedures such as encephalo-duro-arterio-myo-synangiosis (EDAMS) play a crucial role in restoring cerebral perfusion and preventing further neurological deterioration. Timely intervention and multidisciplinary management can significantly improve long-term outcomes in affected patients.

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