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Superior Cerebellar Artery Aneurysm in a Case of Moyamoya Disease: A Rare Entity

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Abstract

Moyamoya disease is a rare condition characterized by progressive narrowing and occlusion of internal carotid artery and other major arteries of the brain with formation of multiple collaterals. The incidence of aneurysm in moyamoya disease is higher than normal population. Treating the simultaneous pathology of aneurysm and moyamoya disease in an individual is a rare entity. A 35-year-old male presented with headache, loss of consciousness, and generalized tonic-clonic seizure with subarachnoid hemorrhage. Digital Subtraction angiography showed moyamoya disease with left superior cerebellar artery aneurysm. Rupture of aneurysm was suspected to be the source of subarachnoid hemorrhage. Patient underwent clipping of the left superior cerebellar artery aneurysm with encephalo-duro-arterio-myo-synangiosis. Patient survived the procedures. All patients with subarachnoid hemorrhage in moyamoya disease should undergo angiogram and aneurysm should be looked for carefully. The site of aneurysm and the risk for damaging the collaterals should be kept in mind while planning the surgery in such cases.

Keywords

- superior
- cerebellar
- artery
- aneurysm
- moyamoya

Introduction

Moyamoya disease is a rare condition characterized by progressive narrowing and occlusion of internal carotid artery (ICA) and other major arteries of the brain with formation of multiple collaterals. This may lead to ischemic or hemorrhagic stroke. The incidence of aneurysm in moyamoya disease is about 3.4 to 14.8% which is higher than normal population.[1] The pathogenesis of aneurysm involves factors like abnormal vessel architecture, hemodynamic stress, and site of location.

Case Report

A 35-year-old male presented to the emergency department with complaint of headache for 3 days and loss of consciousness followed by fall and one episode of generalized tonic-clonic seizure (GTCS) on the day of presentation with no significant past history and comorbidities. On examination, the patient was intubated on volume-controlled ventilation mode, vitals were stable, and Glasgow Coma Scale (GCS) was E1 Vet M5. On computed tomography (CT) brain he was found to have subarachnoid hemorrhage (SAH) noted in the perimesencephalic cistern, prepontine, and premedullary cisterns, along the sulci of right fronto-parieto-temporal region, left cerebral hemisphere, bilateral Sylvian fissures along with intraventricular hemorrhage noted in the occipital horns of bilateral lateral ventricles and fourth ventricle. On CT brain angiography there was narrowing of bilateral ICAs, bilateral anterior cerebral arteries, bilateral middle cerebral arteries, left posterior cerebral artery, and distal part of right the posterior cerebral artery.

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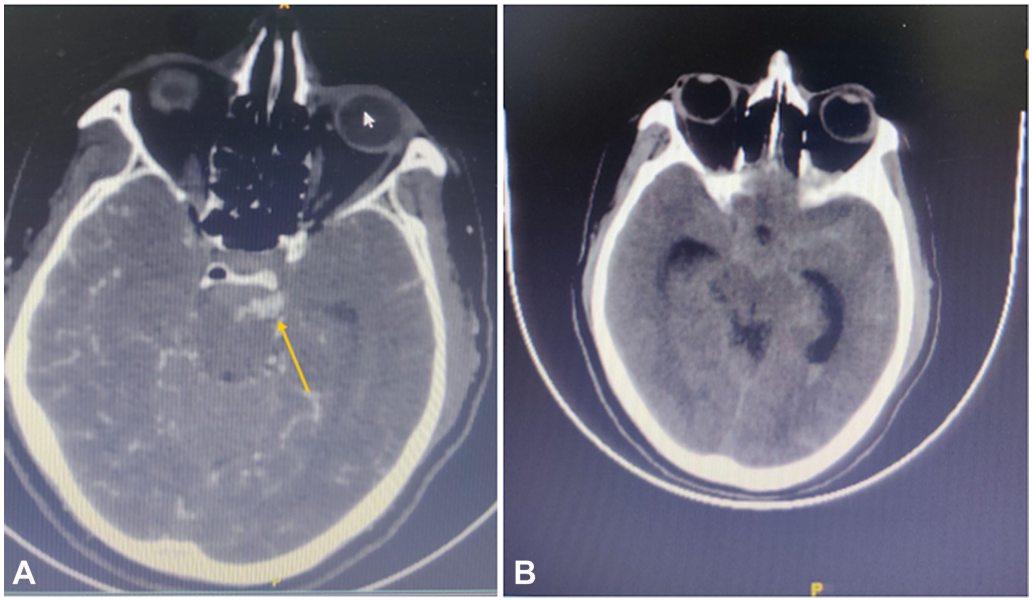

Fig. 1 Computed tomography (CT) angiogram showing left superior cerebellar artery aneurysm (**A**) and CT brain plain showing subarachnoid hemorrhage (SAH) in cisternal and other subarachnoid spaces along with intraventricular hemorrhage (IVH) (**B**).

Multiple collaterals around the circle of Willis were suggestive of moyamoya disease. Small aneurysm was seen arising from the terminal part of the basilar artery (►**Fig. 1**). After initial resuscitation and antiedema measures, his condition improved and he was extubated with GCS of E4 V5 M6. He was planned for digital subtraction angiography (DSA). Cerebral perfusion study was not done as patient presented in emergency with ruptured aneurysm with SAH and in such scenario perfusion study is likely to show false positive results due to vasospasm.

On DSA (►**Fig. 2**), diagnosis of moyamoya disease was confirmed and left superior cerebellar artery aneurysm was found. Patient's relatives were explained regarding possible modalities of treatment of the ruptured aneurysm in the moyamoya disease. But due to financial constraints, patient's relatives selected clipping of aneurysm and revascularization. There were collaterals from middle meningeal artery visible on the right side of the circulation in angiogram, hence the surgery was planned from left-sided approach for aneurysm clipping along with revascularization on the same side.

Preoperatively, the patient's GCS was E4 V2 M5. Patient underwent left pterional craniotomy with anterior temporal approach through trans-Sylvian, transcavernous corridor with mobilization of the third nerve and microsurgical clipping of left superior cerebellar artery aneurysm (►**Fig. 3**) with encephalo-duro-arterio-myo-synangiosis (EDAMS) on the same side (left). He was managed postoperatively with nimodipine and aspirin to prevent vasospasm and infarct, respectively. Postoperatively, the patient was vitally stable and GCS was E4 V2 M5 with left 3rd nerve palsy.

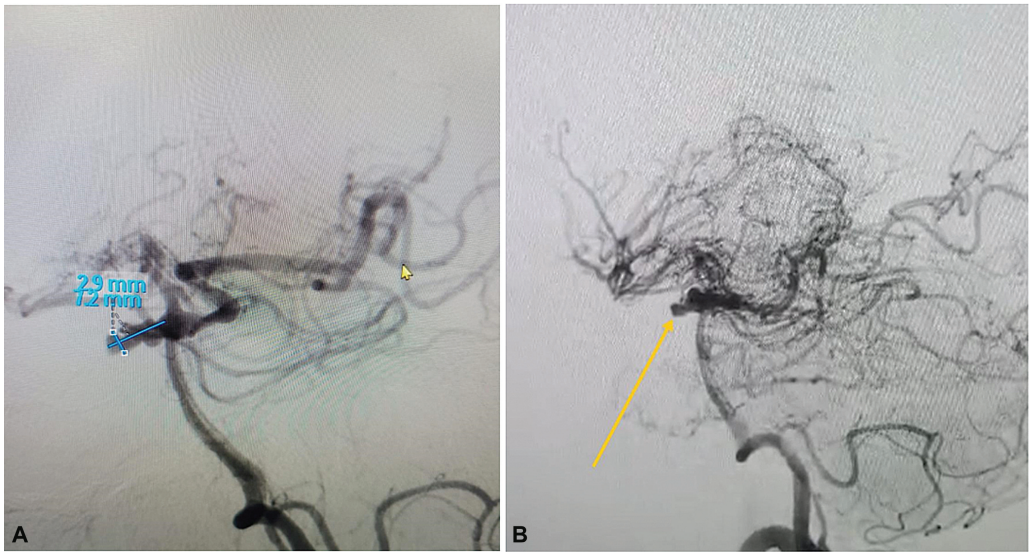

Fig. 2 (A) Showing digital subtraction angiography (DSA) with picture of moyamoya and left superior cerebellar artery aneurysm and (**B**) showing another view of the aneurysm.

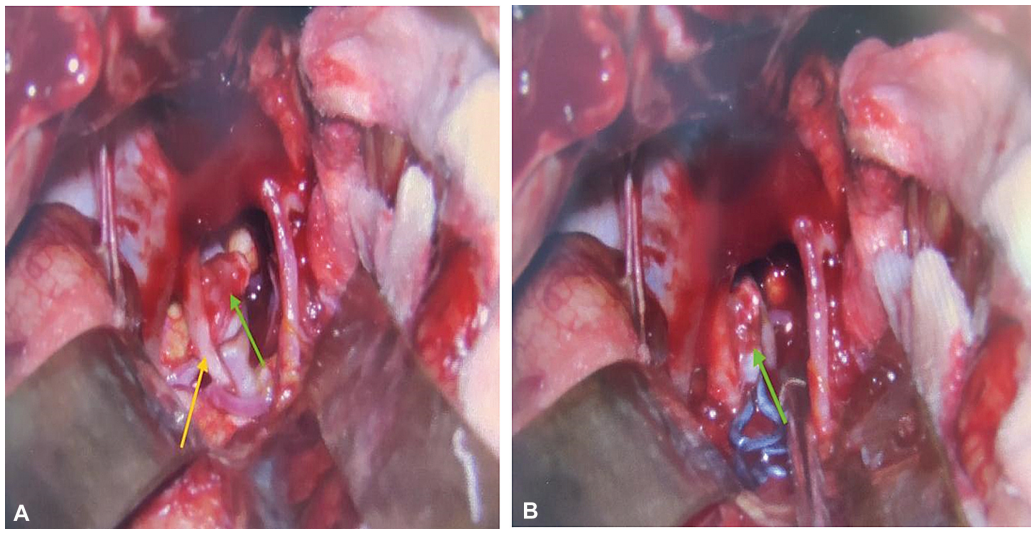

Fig. 3 (**A**) Showing intraoperative picture of left superior cerebellar artery (SCA) aneurysm before clipping and (**B**) showing picture of left SCA after clipping (green arrow shows aneurysm and yellow arrow shows 3rd nerve).

Discussion

Moyamoya disease is a progressive and occlusive cerebrovascular disorder of unknown etiology and is correlated with neither atherosclerosis nor inflammation.[2,3] Moyamoya disease is characterized by steno-occlusion at the terminals of bilateral or unilateral ICAs, and/or the terminal branches, such as middle cerebral arteries and anterior cerebral arteries along with formation of collateral capillary networks.[4] It has also been reported that approximately 3.4 to 14.8% of patients with moyamoya disease can have intracranial aneurysms.[1] In moyamoya disease, the occlusion of the anterior circulation of the circle of Willis predominantly results in a compensatory increase in blood flow in the basilar artery. Such a hemodynamic burden can directly affect major arteries and lead to the formation of aneurysms. Moyamoya disease-associated aneurysms usually develop in the posterior circulation, particularly in the basilar artery.[5,6] Because basilar tip aneurysms are located deeply, direct clipping is difficult due to its anatomical location. Endovascular embolization is the main treatment option for these aneurysms. Aneurysms associated with moyamoya disease have also been found in other areas of the posterior circulation, including the superior cerebellar artery,[5] the P1-P2 junction of the posterior cerebral artery,[7] and the vertebrobasilar junction.[8] The treatment strategies for these differ from those for basilar tip aneurysms and are associated with better outcomes mainly because of their anatomic locations, relatively easy accessibility, and less hemodynamic stress. Numerous studies have shown that revascularization can reduce the rate of stroke recurrence and frequency of transient ischemic attacks in patients with moyamoya disease as well as improve their activities of daily living.[9,10] The aneurysms, which cannot be accessed by direct surgical resection and endovascular embolization, could be treated with surgical revascularization.[11] Thus, in the treatment strategies of moyamoya disease-associated aneurysms patients, microsurgical clipping or endovascular embolization, along with revascularization treatment are the options.[10] Extracranial revascularization of moyamoya disease mainly includes direct or indirect extracranial revascularization, or a combination of the two and other techniques. Direct revascularization involves bypass surgery by anastomosis of the superficial temporal artery with the middle cerebral artery. The indirect revascularization is represented by encephalo-duro-arterio-synangiosis, encephalo-myo-synangiosis, and EDAMS. A 35-year-old male presented with headache, loss of consciousness, and GTCS with SAH. DSA showed moyamoya disease with left superior cerebellar artery aneurysm. Rupture of aneurysm was suspected to be the source of SAH. Patient underwent clipping of the left superior cerebellar artery aneurysm along with EDAMS. Patient survived the procedures with some neurological deficits. Postoperatively, the patient was vitally stable with GCS same as that of preoperatively with left 3rd nerve palsy and was discharged on postoperative day 15.

Conclusion

The simultaneous presence of aneurysm and moyamoya disease in an individual is a rare entity. All patients with SAH and moyamoya disease should undergo angiogram and aneurysm should be looked for carefully. The site of aneurysm and the risk for damaging the collaterals should be kept in mind while planning the surgical treatment in such cases. Microsurgical clipping or endovascular embolization, followed by revascularization treatment improves the outcome of patients in such cases.

Note

The presence of aneurysm and moyamoya both together is a rare thing. All patients with subarachnoid hemorrhage in moyamoya disease should undergo angiogram. Dealing the aneurysm along with revascularization procedure is the management in such cases.

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None.

Conflict of Interest
None declared.

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