

Surgical Management of Giant Basilar Tip Aneurysm Associated with Moyamoya Disease: A Case Report and Literature Review

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Key words

- Giant aneurysm
- Headache
- Hemiplegia
- Moyamoya disease
- Subarachnoid hemorrhage

Abbreviations and Acronyms

CT: Computed tomography EVD: External ventricular drainage IVH: Intraventricular hemorrhage SAH: Subarachnoid hemorrhage

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INTRODUCTION

Moyamoya disease is a slowly progressive occlusive disease, with the initial manifestation of stenosis of the bilateral supraclinoid carotid arteries and associated development of abnormal collateral circulation at the base of the brain and basal ganglia (12, 10).

The vascular changes in this pathology typically result in ischemic strokes in children and cerebral hemorrhages in young adults, although bleeding can occur at any age. Moyamoya disease is now recognized worldwide, even in the Middle East. The cause of this disease has not been identified (13, 18).

Cerebral aneurysms associated with moyamoya disease are uncommon. When they occur, they are generally located around the circle of Willis, in the basal ganglia, and on the collateral vessels (anterior and posterior choroidal arteries) in a 3:1:1 distribution. Aneurysms of the major arteries of the basal arterial circle occur as a result of the high velocity blood BACKGROUND: We report the efficacy and safety of surgical treatment with the orbitozygomatic transsylvian approach and the rarity of accompanying subarachnoid hemorrhage, Terson syndrome, and ruptured giant basilar tip aneurysm in a patient affected by moyamoya disease with a good outcome.

METHODS: A 40-year-old man with a history of exertional headache from childhood and hypertension diagnosed 5 years previously was referred to us with the presenting symptom of headache. After 1 day, he developed left hemiplegia, decreased level of consciousness, and blindness in the left eye. Imaging studies revealed a subarachnoid hemorrhage and giant basilar tip aneurysm. He underwent surgical clipping of the aneurysm using the orbitozygomatic transsylvian approach.

RESULTS: With obvious improvement in strength and consciousness, the patient was discharged. Early diagnosis and treatment of the other possible complications of moyamoya disease were recommended.

CONCLUSIONS: The rarity and different manifestations of moyamoya disease require individualized decision making with regard to cerebrovascular complications. Individualized decision making and the cooperation of medical teams are the mainstays of treatment.

flow, secondary to the arterial stenosis. Modification of the blood flow after revascularization often leads to spontaneous regression and disappearance of these aneurysms (6, II, I5, I9).

The prognosis for aneurysms associated with moyamoya disease is poor compared with typical cerebral aneurysms. Furthermore, surgical management of these lesions is complicated by the presence of essential delicate collateral vasculature. The prevalence of intracranial aneurysms in this disease is around 10%. During the last 40 years, more than 150 cases of aneurysm associated with moyamoya disease have been reported in the adult population, whereas such association in childhood is very rare with only 8 cases in children in the literature. However, most of these aneurysms were less than 25 mm in diameter, the size defined for a giant aneurysm. Aneurysms in moyamoya disease are classified into 2 groups: those involving the arteries of the circle of Willis (major artery aneurysm, 56%) and those arising from peripheral vessels (choroidal,

moyamoya vessels, and meningeal vessels, 44%) (7, 8, 12, 20).

We report a rare case of moyamoya disease with a Hunt & Hess grade 4 ruptured giant basilar artery aneurysm with Terson syndrome and hemiplegia that was treated using an orbitozygomatic subtemporal transsylvian approach with excellent recovery.

CASE REPORT

A 40-year-old man with a history of exertional headache from childhood and hypertension (HTN) diagnosed 5 years previously, was admitted to another hospital with the presenting symptom of headache. After I day, he had developed left hemiplegia, decreased level of consciousness (Glasgow Coma Score IO/I5), and blindness in the left eye.

The computed tomography (CT) scan on admission revealed a subarachnoid hemorrhage (SAH) located predominantly in the prepontine and proximal sylvian cisterns (Figure 1A), intraventricular hemorrhage



Figure 1. Axial brain computed tomography scan of the patient on the first day shows thick subarachnoid hemorrhage in the prepontine, ambiens, crural, and proximal sylvian cisterns (A). Axial (B) and sagittal (C) T2-weighted magnetic resonance imaging 16 days after subarachnoid hemorrhage shows a mass effect of the partially thrombosed giant aneurysm on pons.

(IVH) in all 4 ventricles, and communicating hydrocephalus. Magnetic resonance imaging revealed a mass effect of the partially thrombosed giant aneurysm on the pons (Figure 1B, C). Given these circumstances, external ventricular drainage (EVD) was started. It was removed 7 days later and the patient was referred to us for further investigation and treatment, 2 weeks after first admission.

Digital subtraction angiography (Figure 2A, B) and CT angiography (Figure 2C, D, F) revealed the tip of a basilar artery giant aneurysm, with maximum dimensions of 27×15 mm in the non-thrombosed section, with a narrow neck (4 mm), antero-inferior projection, and typical features of moyamoya disease, including severe stenosis of the supraclinoidal carotid arteries, the proximal middle cerebral artery, and anterior cerebral artery, with collateral vessels from the external carotid arteries (Figure 3). In the Suzuki classification of moyamoya disease, the patient corresponded to stage 5. On the third hospitalization day in our center, antibiotic therapy was started because of fever and meningeal signs suggestive of a diagnosis of meningitis. Acinetobacter meningitis was confirmed in a cerebrospinal fluid culture and the patient was treated with intravenous meropenem. During hospitalization in our center, rebleeding occurred, making surgery inevitable after the patient recovered.

After the SAH was absorbed and the patient recovered from the meningitis, he underwent left side ventriculoperitoneal shunting because of persistent communicating hydrocephalus. We chose the left side for shunting because we planned right side orbitozygomatic craniotomy.

A prolonged course of rehabilitation and physiotherapy took place to prepare him for the mainstay of treatment. After general anesthesia in the supine-bump position with the head turned 30° to the left side, we performed orbitozygomatic craniotomy. The dura was opened, with preservation of the dural branches and leptomeningeal anastomosis. The sylvian fissure was dissected and the carotid artery, posterior communicating artery, and the posterior cerebral artery were exposed. We did not encounter any problem with collateral vessels. Using a combined transsylvian and subtemporal trajectory, the basilar tip aneurysm, with anteroinferior projection, was seen. The neck of the aneurysm was carefully dissected from the basilar trunk, perforating the vessels and both posterior cerebral arteries. The neck of the aneurysm was clipped with a standard straight 7-mm Yasargil aneurysm clip, from lateral to medial direction, without temporary occlusion (Figure 4). Then, the aneurysm was opened to decompress the brain stem and, after checking for successful clipping and for hemostasis, the dura was closed. The bone flap and zygoma were fixed with titanium miniplates and screws. The postoperative period was uneventful except for transient right occulomotor paresis, which resolved after 2 weeks. The patient was followed for left vitreous hemorrhage.

DISCUSSION

This patient presented with a variety of conditions and complications, with teaching value in the field of neurovascular surgery. The onset of rupture of a giant basilar artery bifurcation aneurysm in a young patient, with a history of HTN, IVH, acute hydrocephalus, Terson syndrome, SAH Hunt & Hess grade 4 neurological grading, a mass effect of such a giant aneurysm on the brainstem, represented the first challenge. Managing the acute hydrocephalus with EVD, postsurgical Acinetobacter meningitis, discovery of moyamoya Suzuki stage 5 on angiography, chronic hydrocephalus, which prompted ventriculoperitoneal shunting presented a second group of management challenges. Further challenges included rand stabilization of the patient, avoiding an endovascular and/or early aneurysm obliteration procedure, a sudden rebleeding accident, and the use of a combined subtemporal orbitozygomatic approach for moyamoya disease accompanying a giant aneurysm of the tip of the basilar artery.

Acute hydrocephalus and IVH are often observed after aneurysm rupture, particularly in poor grade patients and those with thick subarachnoid blood on CT scan. EVD is recommended, particularly when the patient has depressed consciousness. Several surgeons have observed that EVD increases the risk of aneurysm rebleeding or impairs the natural mechanisms that arrest aneurysm rupture (14, 16, 17).

Abrupt intracranial (i.e., subarachnoid) bleeding is an uncommon yet interesting mechanism of vitreous hemorrhage, which is called Terson syndrome. Between 10% and 40% of all patients with SAH have an associated vitreous hemorrhage. Bilateral vitreous hemorrhage often occurs in younger individuals and is a poor prognostic indicator of cerebrovascular accidents. Several studies have suggested that patients with both subarachnoid and vitreous bleeding are 4.8 times more likely to die compared with patients with SAH alone (3, 14, 16).

Percutaneous ventriculostomy is an important adjunct used for the management of hydrocephalus, IVH, and intracranial HTN. This mode of treatment could be compromised by the increased risk of catheter-related cerebrospinal fluid infection. Ventriculostomy infection remains the main morbidity associated with



Figure 2. Digital subtraction angiography on day 20: anteroposterior (A) and lateral (B) views; computed tomography angiography on day 24. Coronal (C) and sagittal (D, E) reconstructions show a partially thrombosed giant basilar tip aneurysm with antero-inferior projection and left side deviation.

this procedure despite the use of prophylactic antibiotic medications, meticulous external drainage care, and valve-regulated systems. An infection rate of 0% to 27% has been reported, with a risk of up to 100% by the 11th day of monitoring. This case was transferred to our ward 6 days after discontinuing EVD. Secondary to



Figure 3. Preoperative digital subtraction angiography of the carotid. The anteroposterior (A) and lateral (B) views show typical features of moyamoya disease with relatively good collateral circulation.

fever on the admission day and cerebrospinal fluid analysis, he received intravenous antibiotic medications.

CASE REPORT

Chronic hydrocephalus is observed in 20% of patients after SAH (16). Half of the patients with acute clinical hydrocephalus eventually require a ventriculoperitoneal shunt. Factors associated with hydrocephalus included ventricular size and IVH on admission, poor clinical grade, preexisting HTN, increased size, pneumonia, aneurysm and meningitis. The need for a permanent shunt can be reduced by EVD (including long, tunneled catheters) and serial lumbar punctures. In our patient, we did serial lumbar punctures for a long period of time. However, because of continuing hydrocephalus, despite this management, we finally placed the shunt.

Aneurysms associated with moyamoya disease represent a unique challenge for cerebrovascular surgeons. General considerations for these patients should be integrated into the operative technique and preoperative planning. First, protection of transdural and leptomeningeal anastomosis is important in planning the craniotomy flap. This can be done by preserving as many dural branches as possible during exposure, as well as minimizing disruption of the rich cortical collateral circulation. Image guidance allowed us to minimize the size of the craniotomy and the cortical incision in our patient, hence theoretically decreasing the disruption of the collateral circulation. These collateral pathways play an important role in moyamoya disease. It is also important to minimize brain retraction during surgery because tolerance to ischemia and hemodynamic reserve capacity are poor (1-5, 9, 10).

To avoid the course of the middle meningeal artery, we opened the dura as far as possible from the emerging point of the artery. Many surgeons advocate subpial uncal resection in the transsylvian approach and others insist on resecting the inferior temporal gyrus with the fusiform and parahippocampal gyri in the subtemporal approach for better exposure. We avoided all of the above cortical incisions and resections to minimize disruption of the collateral circulation.

The timing of aneurysm obliteration for posterior circulation aneurysms is less well defined. Much of the early information



Figure 4. Postoperative digital subtraction angiography 3 months later. The anteroposterior (A) and lateral (B) views show complete clipping of the aneurysm with good bilateral posterior cerebral artery circulation.

favoring delayed surgery came from referral centers and delayed surgery may be preferable for complex lesions, such as giant aneurysms and aneurysms oriented in the basilar artery bifurcation.

Radical bone removal, cranial base approaches, and ventricular drainage may be useful to limit retraction. Similarly, transient interruption of cerebral blood flow, as well as perioperative hypercapnia, hypocapnia, hypotension, and hypovolemia must be controlled meticulously in these patients. Another concern when setting up aneurysm surgery in patients with moyamoya disease is the extent of development of the moyamoya vessels around the aneurysm itself, especially in the case of basilar apex aneurysms. If the parent vessel serves an important collateral route and preservation of the parent artery is thought to be impossible, preoperative bypass surgery or combined bypass and aneurysm surgery should be considered. Because of poor patient grade and the general condition of our patient as listed below, we considered late surgery rather than early surgery for clipping the aneurvsm:

- 1) Hunt & Hess grade 4 SAH
- 2) Suzuki stage 5 moyamoya disease
- 3) Tardy referral of the patient
- 4) Postsurgical meningitis and antibiotic therapy

- 5) Poor prognosis due to SAH combined with viterous hemorrhage
- 6) Giant aneurysm of the posterior circulation with antero-inferior projection
- 7) The young age of the patient influenced decision making, according to the Le Roux et al. algorithm, for patients with a poor grade aneurysm
- 8) Spontanous regression of a giant aneurysm of the posterior circulation after revascularization surgery, such as encephalo-duro-arterio-synangiosis, is not common (13).

The pterional transsylvian approach is one of the most commonly used approaches for aneurysms of the anterior circulation. Exposure can be further enhanced by drilling the pterion and the bony ridges over the floor of the frontal fossa. When combined with a pterional transsylvian approach, orbitozygomatic osteotomy can greatly increase the amount of exposure to the tip of the basilar artery.

After surgical treatment was complete and the patient was discharged, we followed him for further probable interventions. Because of excellent extracranial to intracranial collaterals, we convinced him that he may be protected from additional ischemia. We obtained a perfusion single-photon emission CT scan, with and without acetazolamide, to evaluate his reserve capacity. He did not show any signs of poor reserve and we decided to only follow him, without bypass surgery. One could argue that another reason to revascularize him would be to decrease the size of the moyamoya lenticulostriate vessels, which pose a risk of hemorrhage. Nevertheless, because of scalp incision on both sides, which would probably not afford access to superficial temporal arteries, we decided to postpone bypass surgery until a symptomatic condition arises.

CASE REPORT

According to our protocol, we usually follow up with magnetic resonance angiography once a year, and if new symptoms develop, then we obtain an angiogram. However, no new symptoms have developed in this patient after 2 years. Although several studies prefer antiplatelet or anticoagulation therapy for patients with moyamoya, we worry about that leading to hemorrhage and do not prescribe them.

CONCLUSIONS

We report the efficacy of surgical management of a ruptured giant basilar tip aneurysm using the orbitozygomatic transsylvian approach in a 40-year-old patient with moyamoya disease. The rarity and different manifestations of moyamoya disease require individualized decision making with regard to its cerebrovascular complications. A literature review shows that the neurovascular surgeons' armamentarium is still expanding to cope with the dreadful situations that the disease may cause. The medical team must use all of their experience to manage such difficult patients from the first step to ultimate recovery through follow-up.

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