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Surgical management of a ruptured posterior choroidal intraventricular aneurysm associated with moyamoya disease

Case Report

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Abstract: Background: prevention of rebleeding is the most important aspect of the management of hemorrhagic moyamoya disease because rebleeding causes significant morbidity and mortality.Clinical presentation: a 68-year-old female patient presented with intraventricular hemorrhages abutting the atrium of the right ventricle. Cerebral angiography showed internal carotid artery occlusion with moyamoya vessels on the right side and internal carotid artery stenosis with moymoya vessels on the left side. The posterior cerebral artery was enlarged on the right side, and a lateral posterior choroidal intraventricular aneurysm was identified. The aneurysm was successfully excised transcortically using a neuronavigation system to minimize damage to collateral vessels and shorten the surgical corridor. Histopathology revealed a pseudoaneurysm. Three months later, indirect revascularization at the right hemisphere was performed. Conclusion: the management of hemorrhagic moyamoya disease should be modified based on the source of hemorrhage. Because of the rebleeding risk, we recommend early intervention to treat ruptured intracranial aneurysms using minimally invasive surgical techniques.

Keywords: *Moyamoya disease • Aneurysm • Neuronavigation*

1. Introduction

Cerebral aneurysms associated with moyamoya disease are uncommon. Aneurysms are thought to arise in collateral vascular structures in moyamoya disease, like perforator and choroidal artery because of altered hemodynamics and the inherent vascular fragility [1] The cause of hemorrhage is reportedly moyamoya vessel failure from hemodynamic stress. Therefore, the rate of rebleeding is higher, especially in the presence of an aneurysm [2]. Conversely, the surgical management of aneurysms is complicated by the presence of an important and delicate collateral vasculature.

We report a rare case of a patient with moyamoya disease who had a ruptured aneurysm originating from the distal right lateral posterior choroidal artery, which we managed by a frameless stereotactically guided trapping and excision method. In addition, we present a review of the literature on moyamoya-related cerebral aneurysms and intraventricular aneurysms.

2. Case presentation

A 68-year-old female patient was admitted to our hospital with severe headache and conscious disturbance. Head computed tomography (CT) revealed intraventricular hemorrhage and acute hydrocephalus (Figure 1).

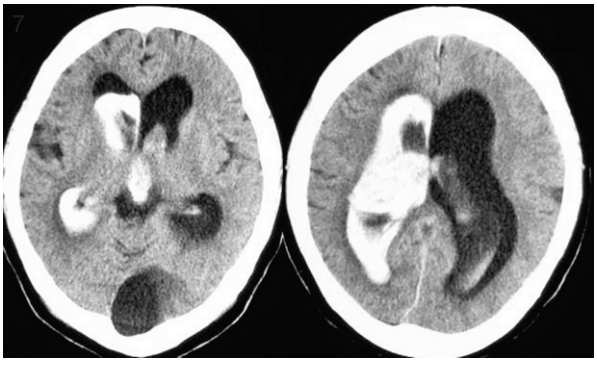

Figure 1. Head CT showinf intraventricular hemorrhage and acute hydrocephalus.

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Extraventricular drainage was established, and cerebral angiography was performed. The right distal internal carotid artery was completely occluded above the origin of the ophthalmic artery. The left A1 segment was also nearly occluded. Ethmoidal moyamoya vessels were found on the right side. Transdural anastomosis was not found. Right vertebral artery cerebral angiography showed an aneurysm on the lateral posterior choroidal artery. The aneurysm was identified next to the lateral wall of the atrium by CT angiogram (Figure 2).

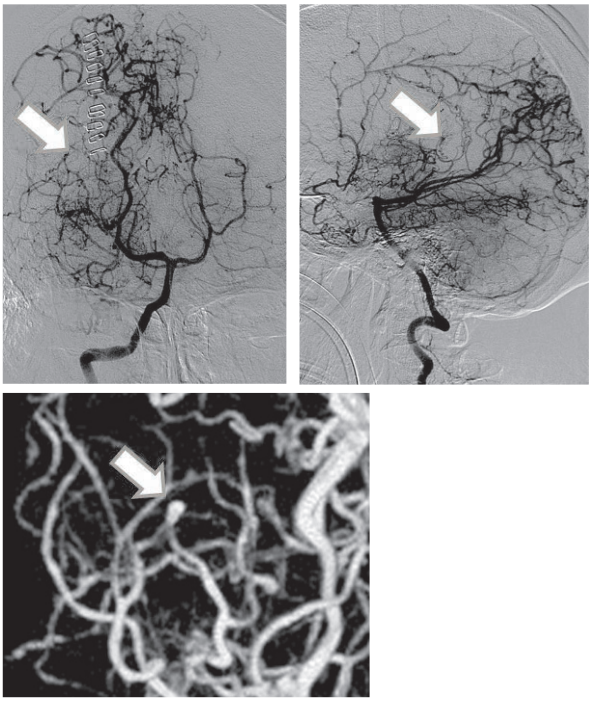

Figure 2. Right vertebral angiogram reveals the aneurysm on the right lateral posterior choroidal artery (arrow).

We performed surgical resection of the aneurysm the next day using a neuronavigation system. Temporal craniotomy preserved the frontal branch of the superior temporal artery, which could possibly be used as a donor artery for future revascularization. A corticotomy was created in the superior temporal sulcus. After reaching the right ventricle, the intraventricular hematoma was evacuated. The aneurysm was exposed on the lateral wall of the atrium (Figure 3).

We trapped the aneurysm on the parent artery and performed aneurysmectomy because this lesion was suspected to be a pseudoaneurysm. Postoperative right vertebral arteriography showed no aneurysm on the lateral posterior choroidal artery and no change in collateral flow. Diffusion-weighted images showed no major infarction. Gross specimen examination revealed that the rupture site included a freshly attached thrombus with no arterial wall. Histopathology showed a pseudoaneurysm with a small fragmented arterial wall composed of thickened fibrocellular intima (Figure 4). Three months later, the patient was alert and had no motor paresis. Direct superficial temporal artery–middle cerebral artery anastomosis was attempted, but no recipient artery was found. Therefore, we performed encephalo-duro-arterio-myo-synangiosis. The patient had no additional focal deficits after the procedure.

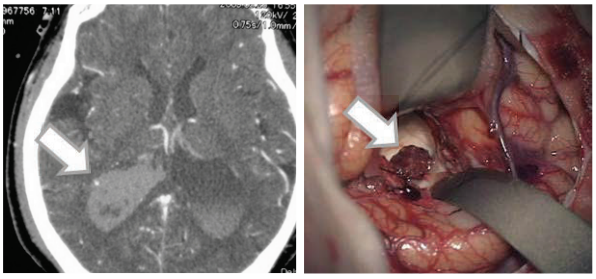

Figure 3. CT angiogram reveals the aneurysm is identified beside the lateral wall of the atrium arrow.
Fig 3-2 Intraoperative Finding. The aneurysm (arrowhead) exists beside the lateral wall of the atrium (asterisk), opening the superior temporal sulcus.

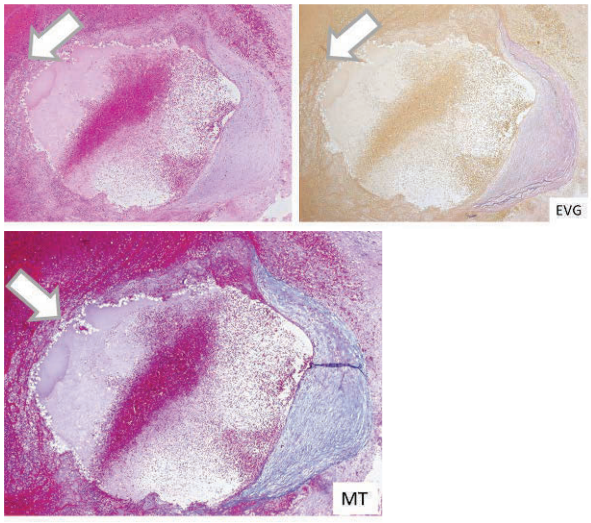

Figure 4. Histopathological specimen The rupture sites are attached by fresh thrombus, and showed no arterial wall (arrowhead). Only the small fragmented arterial wall, which is composed of thickended fibrocellular intima, was found (arrow).

3. Discussion

Cerebral aneurysms associated with moyamoya disease generally occur around the Circle of Willis, in the basal ganglia, and on the collateral vessels. In a series of 81 patients with moyamoya disease, 15% had cerebral aneurysms; 56% of those were present in the Circle of Willis, and a significant minority were found in the basal ganglia (18%) and collateral vessels (22%) [2].

There are few reports in which the aneurysms were histologically analyzed. To the best of our knowledge, only seven such cases of peripheral artery aneurysms associated with moyamoya disease [3-6] have been reported. Most peripheral or collateral artery aneurysms in moyamoya disease are pseudoaneurysms [4,7-9], as in our case. Cerebral angiography could not distinguish among these pathological findings. Collateral vessels in moyamoya disease have a weakened intima media with segmentation of the internal elastic lamina and fibrosis [10]. The attenuated arterial wall acquires a predisposition for microaneurysm formation or rupture under hemodynamic stress [10].

The natural history of peripheral artery aneurysms in moyamoya disease is unclear. For moyamoya-related aneurysms that arise from choroidal arteries, some authors recommend a conservative approach because spontaneous regression may occur [2,3]. Conversely, some reports have indicated that aneurysmal rupture may be repeated earlier than previously thought [3,5,11]. Iwama et al. [12] analyzed 46 patients who developed rebleeding associated with hemorrhagic moyamoya disease and concluded that tiny aneurysms at the periphery of collateral vessels may have caused rebleeding within a few months resulting from weakened vessel wall failure under unusually increased hemodynamic stress. Furthermore, the prognosis of rebleeding in patients with hemorrhagic moyamoya disease is extremely poor [3,5,12,13]: rebleeding has been associated with a mortality rate of approximately 25% compared with the first episode of bleeding (5% mortality rate) [13].

To prevent early rebleeding from an aneurysm, direct surgical intervention such as aneurysmectomy or clipping could be a treatment option. However, these procedures may carry a risk of additional brain damage because aneurysms of collateral vessels are usually deeply located. Ali et al. [14] suggested three considerations in the surgical planning for aneurysms in moyamoya disease. First, transdural and leptomeningeal anastomosis must be preserved, and a donor for the bypass surgery (i.e., STA) should be presented in planning the craniotomy flap. Second, the degree of moyamoya vessel development around the aneurysm itself should be considered. Because collateral circulation is important in patients with moyamoya disease, surgical injury of these vessels can produce additional ischemic brain damage [15]. Finally, the indication and timing of the bypass surgery should be taken into consideration. If the parent vessel serves as 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. However, direct revascularization in the acute phase might carry a risk of aneurysmal re-rupture. In this situation, surgical resection of the aneurysm should be conducted. Indeed, surgical revascularization could reduce preexisting collateral vessels or hemodynamic stress on those vessels and may reduce hemorrhagic risk [15]. Surgical revascularization might effectively prevent later rebleeding and additional aneurysm formation. Therefore, we performed revascularization to prevent rebleeding and to reduce the hemodynamic stress in the chronic stage, although the operation turned into indirect revascularization as a result.

Neuronavigational techniques help minimize surgical damage caused by direct surgery. Ali et al. [14] reported a similar case in which they also performed neuronavigation. In our case, the transcortical route via the superior temporal sulcus was the shortest surgical corridor to the aneurysm in the neuronavigation system. On cerebral angiography, the collateral vessels were relatively rough around the superior temporal sulcus. There are other surgical routes to reach the atrium of the lateral ventricle via a transcortical route [16]. Our patient developed leptomeningeal anastomosis from the posterior pericallosal artery and leptomeningeal anastomosis from the posterior cerebral artery. The transcortical route would not only have involved a long distance to reach the lesion, but also would have risked injury to the important collateral circulation.

Endovascular treatment for such aneurysms has recently been reported [17]. Eight collateral aneurysms in patients with hemorrhagic moyamoya disease were treated by endovascular embolization [17]; seven of these eight aneurysms were successfully and completely occluded, but one failed because of a small artery. The lateral posterior choroidal artery in our patient was also too small for an endovascular approach.

Treatment of peripheral artery aneurysms associated with moyamoya disease remains controversial. Surgical resection with a neuronavigation system could be a therapeutic option to prevent rebleeding in the acute phase.

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