

Ruptured Aneurysm of the Accessory Middle Cerebral Artery Associated with Moyamoya Disease – A Case Report

Cheng-Chi Lee, MD; Zhuo-Hao Liu, MD; Shih-Ming Jung¹, MD; Tao-Chieh Yang, MD

The accessory middle cerebral artery can provide collateral blood supply in moyamoya disease. We report a case of unilateral moyamoya disease which demonstrates the anatomy of the right accessory middle cerebral artery and a ruptured peripheral aneurysm on the artery. Our patient was a 56-year-old woman who initially suffered from headache and lethargy. Right caudate nucleus hemorrhage with intraventricular extension and spontaneous subarachnoid hemorrhage were found on brain computed tomography. A ruptured peripheral accessory middle cerebral artery aneurysm associated with unilateral moyamoya disease was diagnosed on cerebral angiography. Surgical intervention to excise the peripheral accessory middle cerebral artery aneurysm assisted by frameless navigation guidance to reduce the risk of damage to collateral vessels was done successfully. Histopathology of excised tissue showed this anomaly was a pseudoaneurysm. The management of an aneurysm in moyamoya disease should be modified based on its location and collateral vessels. Prevention of aneurysm bleeding and preservation of collateral vessels during craniotomy are the critical when managing hemorrhagic moyamoya disease. This case suggests that surgical intervention for ruptured intracranial aneurysms is safe with the use of frameless navigation guidance to minimize collateral vessel injuries. (*Chang Gung Med J* 2011;34:541-7)

Key words: accessory middle cerebral artery, moyamoya disease, ruptured cerebral aneurysm

Moyamoya disease is a chronic occlusive cerebrovascular disorder of unknown etiology. It is characterized by progressive steno-occlusive changes in the terminal portions of the intracranial internal carotid arteries and the circle of Willis, along with concomitant development of fine networks of collateral vessels at the skull base which collateralize vessels distal to the occlusions and appear to serve as a source of supplemental blood flow to ischemic regions of the brain.^(1,2) Unilateral involvement with development of moyamoya vessels is called ‘unilateral’ moyamoya disease. Moyamoya disease is commonly accompanied by intracranial aneurysms.⁽³⁾ The accessory middle cerebral artery (MCA) is a varia-

tion of middle cerebral artery branching, and its incidence has been reported to be 0.3-4.0% in angiographic studies.⁽¹⁾ The accessory MCA can provide collateral blood supply in moyamoya disease.⁽⁴⁾ To the best of our knowledge, this type of accessory middle cerebral artery aneurysm associated with moyamoya disease is extremely rare. We report a rare case of a ruptured peripheral right accessory MCA aneurysm with unilateral moyamoya disease.

CASE REPORT

A 56-year-old Taiwanese woman without a history of systemic medical disease was sent to our

From the Department of Neurosurgery; ¹Department of Pathology, Chang Gung Memorial Hospital at Linkou, Chang Gung University College of Medicine, Taoyuan, Taiwan.

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Correspondence to: Dr. Tao-Chieh Yang, Department of Neurosurgery, Chang Gung Memorial Hospital at Linkou, 5, Fusing St., Gueishan Township, Taoyuan County 333, Taiwan (R.O.C.) Tel: 886-3-3281200 ext. 2119; Fax: 886-3-3285818;

E-mail: jade5048@yahoo.com.tw

emergency room after the sudden onset of headache, dizziness, nausea, and lethargy. The patient had a head injury with a right occipital contusion hemorrhage about 20 years before. Neurological examination on admission revealed an unsteady gait without focal limb weakness. A brain computed tomography (CT) scan demonstrated a diffuse, thin subarachnoid hemorrhage in the basal cistern, hemorrhage in the right caudate nucleus with intraventricular extension, an enlarged ventricle suggestive of acute hydrocephalus and one small, old stroke in the right anterior frontal area. (Fig. 1A, B). Bilateral frontal external ventricular drains were placed. The patient's symptoms and discomfort were relieved after intracranial pressure was controlled by cerebrospinal fluid drainage. Cerebral digital subtraction angiography

(DSA) was performed 7 days after the intracerebral hemorrhage, revealing moyamoya-type vasculopathy in the right hemisphere, including distal supraclinoid internal carotid artery stenosis, right anterior cerebral artery occlusion, proximal MCA stenosis (Fig. 2A, B) and a lobular aneurysm in the right accessory MCA arising from the left anterior cerebral artery. (Fig. 2C, D). Surgical intervention was undertaken to prevent aneurysm rebleeding. We performed a right frontal craniotomy using transfrontal-transventricular approach with the aid of frameless CT-guided stereotactic navigation (Stryker system, Kalamazoo, MI, U.S.A.) (Fig. 1C). After identifying the peripheral right accessory MCA aneurysm and parent artery (accessory MCA artery), excision of the lobular aneurysm with transection of the distal segment of

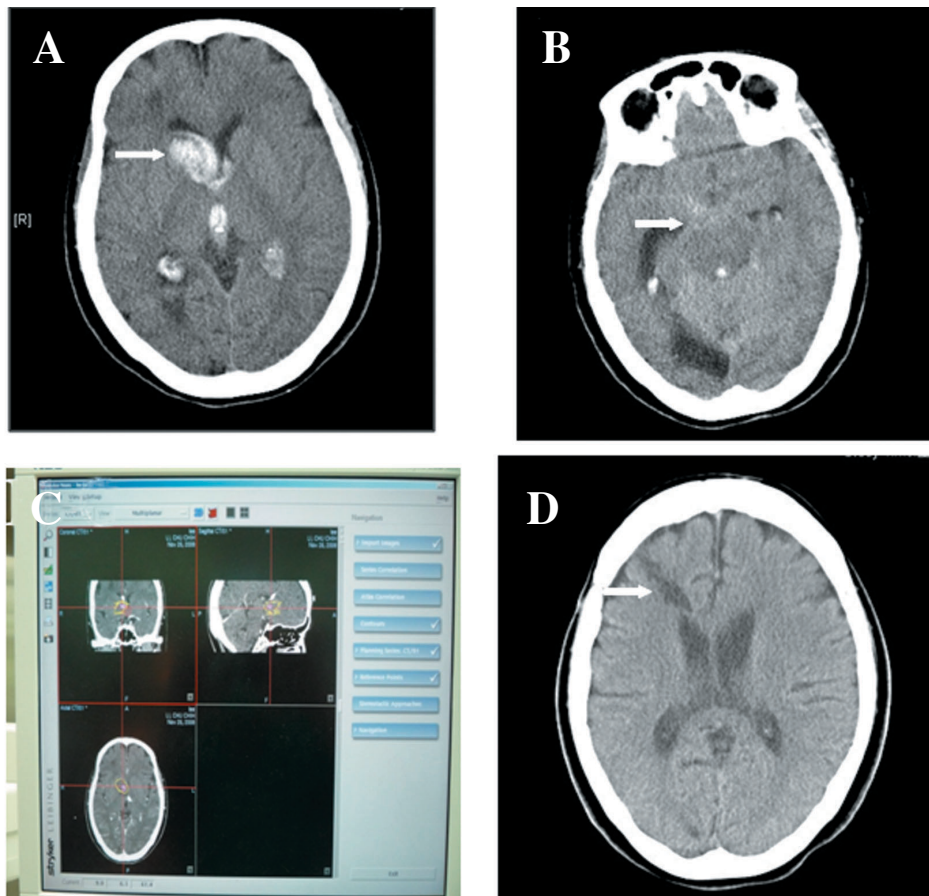


Fig. 1 Serial brain CT studies. (A) Brain CT shows a right caudate nucleus hemorrhage with intraventricular hemorrhage and one small stroke in the right anterior frontal area. (B) Brain CT shows a diffuse thin subarachnoid hemorrhage in the basal cistern. (C) Frameless stereotactic navigator system for aneurysm localization. (D) Right frontal lobe encephalomalacia is seen 3 months postoperatively via a transfrontal transventricular approach. Abbreviation used: CT: computed tomography.

the aneurysm involving the accessory MCA artery was performed. The postoperative course was uneventful, and the patient tolerated the procedure well without any new neurological deficit. Pathology of the excised tissue showed the anomaly was a pseudoaneurysm (Fig. 3A, B). The patient was discharged 2 weeks after the operation and returned to usual daily activity. Cerebral DSA demonstrated complete excision of the aneurysm and accessory MCA transection from the aneurysm site 7 days after the operation (Fig. 3C, D).

DISCUSSION

Cerebral aneurysm associated with moyamoya disease was first reported in 1965.⁽⁵⁾ Aneurysm and

pseudoaneurysm formation have been described in numerous locations in patients with this condition. The incidence of intracranial aneurysm in patients with moyamoya disease ranges from 3% to 14%.^(6,7) Aneurysms in moyamoya disease are thought to arise from the increased hemodynamic stress applied to the fragile collateral blood supply.⁽⁸⁾ Kawaguchi et al. reported that the distribution of these aneurysms on the circle of Willis, the basal ganglia and the collateral vessels was 3:1:1.⁽³⁾ Aneurysms in patients with moyamoya disease are most often located on the bifurcation of the basilar artery and on the cavernous segment of the internal carotid artery. The aneurysms are encountered more often in the posterior circulation than in the anterior circulation.^(3,6) Cases with aneurysms on the collateral vessels or in the basal

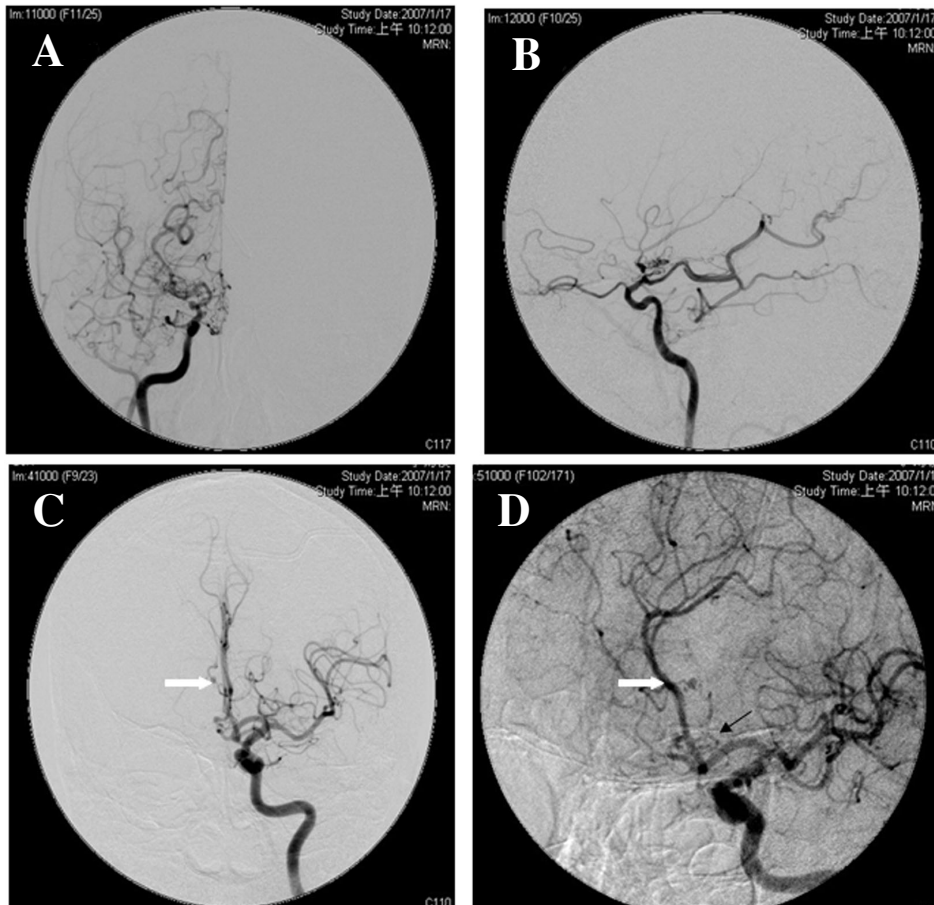


Fig. 2 Preoperative cerebral angiography. Right internal carotid artery (ICA) angiography in the anteriorposterior (A), and lateral views (B) show moyamoya-type vasculopathy in the right hemisphere, distal supraclinoid ICA stenosis, right anterior cerebral artery occlusion, and proximal middle cerebral artery stenosis. Left internal carotid angiography in Cand D shows a lobular aneurysm (white arrow) in the right accessory middle cerebral artery arising from the left anterior cerebral artery (black arrow).

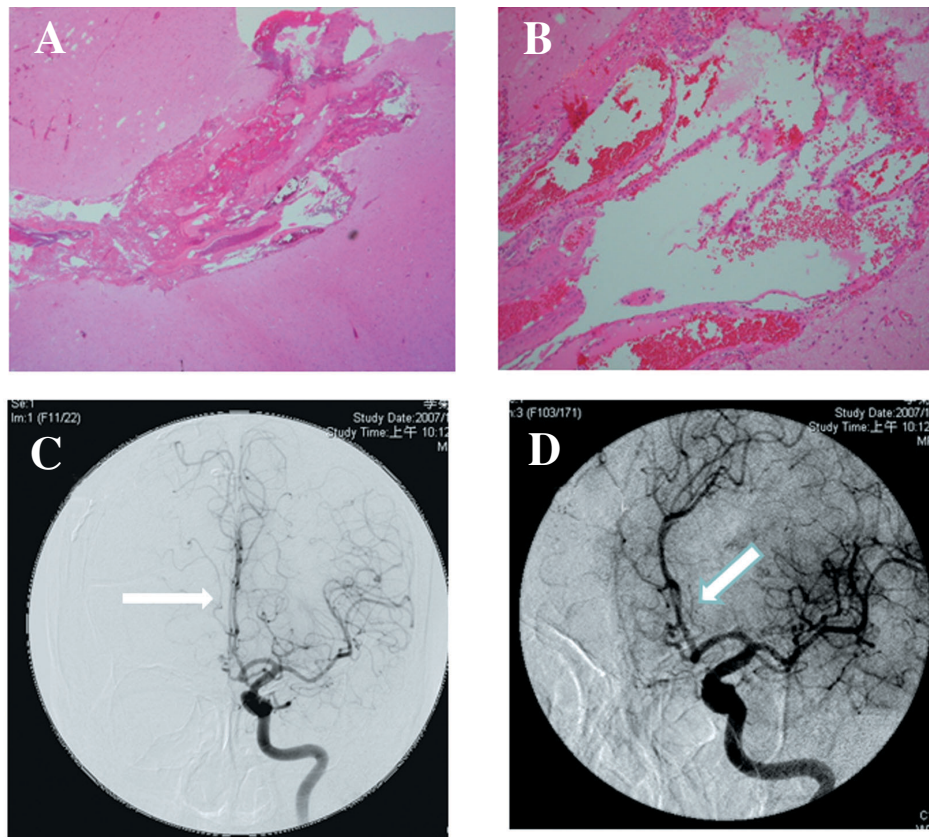


Fig. 3 Pathology and follow-up angiography. Photomicrographs of the resected neurosurgical histopathological specimen (A, B) show only adventitia without an internal elastic membrane and muscle layer, indicating a pseudoaneurysm. (A, hematoxylin and eosin original magnification $\times 8$; B, original magnification $\times 20$) Post-operative left internal carotid artery cerebral angiography (C, D) shows complete excision of the aneurysm and accessory middle cerebral artery transection from the aneurysm site (white arrow).

ganglia tend to have poorer clinical grades compared with those with aneurysms around the circle of Willis.⁽³⁾

In 1962, Crompton et al proposed using the term accessory middle cerebral artery (MCA) to describe the anomalous vessel originating from the horizontal portion of the anterior cerebral artery.⁽⁹⁾ An association of the accessory middle cerebral artery with moyamoya disease was reported by Komiyama et al.⁽¹⁾ This vessel can provide collateral blood supply in moyamoya disease.⁽⁴⁾ The accessory MCA has been reported as an incidental anatomic finding or in coexistence with cerebral aneurysms.⁽⁴⁾ Aneurysms rarely occur at the origin of the accessory MCA. An aneurysm in this vessel was first reported in 1977.⁽¹⁰⁾ There are only ten previous reports of accessory MCA aneurysms.⁽¹¹⁾ Most of them arose at the junc-

tion of the accessory MCA and the A1 segment, and none were associated with moyamoya disease. The collateral arteries most commonly involved with intracranial aneurysms associated with moyamoya disease are the anterior choroidal artery and posterior choroidal artery.⁽³⁾ In our patient, the accessory MCA provided collateral blood supply and there was peripheral aneurysm formation in this collateral artery.

The natural history of peripheral artery aneurysm in moyamoya disease remains to be elucidated. In most reported cases, peripheral artery aneurysms were considered to be pseudoaneurysms.^(7,12) The bleeding rate of aneurysms associated with moyamoya disease has been estimated to be 87.1% in adults.⁽¹³⁾ Other reports also indicated that aneurysmal rupture may recur within a few

months after the initial bleeding episode.^(14,15) Intracranial hemorrhage is the major catastrophic event in the natural course of moyamoya disease, and the outcome of patients with rebleeding is very poor.⁽¹⁶⁾ The rate of recurrent hemorrhage in untreated aneurysm in moyamoya disease is approximately 30%, and therefore definitive treatment is necessary to avoid aneurysm rebleeding to reduce morbidity and mortality in these already compromised patients.⁽⁸⁾

Definitive treatment of aneurysms associated with moyamoya disease can involve both surgical and endovascular methods. Surgical approaches are difficult because of interference by abundant fragile collateral vessels which easily bleed. The collateral pathways play an important role in moyamoya disease, and cannot be severely sacrificed.⁽¹⁷⁾ These aneurysms are thought to be pseudoaneurysms, which cannot be safely clipped. Sacrificing the distal parent artery is necessary to prevent intra-operative rupture and post-operative rebleeding. It is also important to minimize brain retraction, because the tolerance of brain tissue to ischemia and the hemodynamic reserve capacity are poor in patients with moyamoya disease.⁽³⁾ Excessive brain retraction may cause cerebral flow disturbance, resulting in postoperative complications such as cerebral infarction and intracerebral hemorrhage. The use of stereotactic guidance with either CT angiography, magnetic resonance imaging, or three-dimensional angiography can aid in the localization of such small, deep lesions.^(1,14) Once the lesion is localized, a direct approach is used to identify the aneurysm with minimal brain manipulation. Surgical intervention with the use of stereotactic navigation can result in less collateral supply damage and minimal brain manipulation during surgery.

An endovascular approach to aneurysms associated with moyamoya disease has also been considered.⁽¹⁸⁻²⁰⁾ The greatest advantage of endovascular treatment is the avoidance of direct invasion of the brain, such as occurs with retraction, and the aneurysm can be approached without affecting the moyamoya vessel. To the best of our knowledge, reported endovascular treatments for cerebral aneurysm associated with moyamoya disease all involved the main trunk of the posterior circulation.⁽¹⁸⁻²⁰⁾ We found no reports of attempts at treatment of distal peripheral aneurysms associated with

moyamoya disease with an endovascular approach in our review of the literature. Endovascular intervention for distal peripheral aneurysms associated with moyamoya disease is often difficult because of the small caliber of the involved parent vessels and often extremely tortuous endovascular vessel route. In addition, the use of an endovascular approach to obliterate this peripheral type of aneurysm by coils or glues often requires sacrifice of a longer segment of the parent vessel than does surgical intervention.

Conclusion

Peripheral accessory MCA aneurysms associated with moyamoya disease are extremely rare. The optimal management of hemorrhagic moyamoya disease associated with this type of aneurysm remains to be determined because of the condition's rarity. Rapid diagnosis and early intervention are important to avoid rebleeding with subsequent morbidity and mortality.

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毛毛樣腦血管病合併(副)中大腦動脈瘤破裂之個案報告

李丞騏 劉倬昊 容世明¹ 楊道杰

在毛毛樣腦血管病變的少數病例中，附屬中大腦動脈（一種罕見的腦部血管變異）可以提供因毛毛樣病變導致血流供應相對減少的大腦區域足夠的側枝循環。在我們病人身上除了診斷出有單側毛毛樣腦血管病變、同側附屬中大腦動脈變異之外，還發現在此附屬中大腦動脈上有破裂之動脈瘤。一位 56 歲女性病患因頭痛及意識改變被送到急診，腦部電腦斷層發現右側基底核出血、腦室內出血及蜘蛛膜下腔出血。腦部血管攝影證實在右側附屬中大腦動脈上有破裂之動脈瘤，並且意外發現病人之右側大腦半球之血管有毛毛樣血管病變。我們藉著術中定位導航系統的輔助，在沒有傷害到側枝循環血管的情形下成功的摘除此動脈瘤。最後病理診斷確定此一血管病灶為偽動脈瘤。最後的結論是：對於毛毛樣腦血管病變合併有顱內動脈瘤的處理方式，端視此動脈瘤以及其側枝循環位置之差異而有所改變。尤其在面對出血性毛毛樣腦血管病變的病人，最重要而且不變的是要如何在術中保留側枝循環血管的完整性以及避免動脈瘤破裂。而利用術中定位導航系統可以讓我們在處理腦動脈瘤的同時，將側枝循環的血管所可能遭受到的損害降到最低。(長庚醫誌 2011;34:541-7)

關鍵詞：附屬中大腦動脈，毛毛樣腦血管病變，腦動脈瘤破裂

長庚醫療財團法人林口長庚紀念醫院 神經外科，病理科；長庚大學 醫學院

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通訊作者：楊道杰醫師，長庚醫療財團法人林口長庚紀念醫院 神經外科。桃園縣333龜山鄉復興街5號。

Tel: (03)3281200轉2119; Fax: (03)3285818; E-mail:jade5048@yahoo.com.tw