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Resolution of bilateral moyamoya associated collateral vessel aneurysms: Rationale for endovascular versus surgical intervention

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Abstract

Background: Management of aneurysms associated with deep collateral vessels in moyamoya disease is challenging both from an endovascular and a surgical standpoint. Difficulties with access or localization, and compromise of the collateral circulation with subsequent ischemia are the primary concerns, making direct obliteration potentially unfeasible or risky. Alternatively, superficial temporal artery–middle cerebral artery bypass is another potential strategy for resolution of these aneurysms.

Case Description: Presented are the findings and management for a patient with moyamoya disease and bilateral deep collateral vessel aneurysms, successfully treated with endovascular obliteration following a right-sided hemorrhage and subsequently with bypass for an unruptured but growing contralateral aneurysm.

Conclusions: A rationale and approach to management is outlined, as derived from review of the current literature and the illustrative case with bilateral collateral vessel aneurysms.

Key Words: Aneurysm, embolization, extracranial-intracranial bypass, moyamoya, surgical revascularization

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INTRODUCTION

Although moyamoya disease (MMD) presents most commonly with symptoms of cerebral ischemia, hemorrhagic presentation is also a well-known phenomenon, more typically seen in adult patients.[6,19,20] Intraparenchymal hemorrhage is typically ascribed to bleeding from fragile basal moya collaterals, but can also occasionally be associated with aneurysmal or pseudoaneurysmal formation affecting deep collateral perforators. Surgical revascularization is the standard approach for treating ischemic symptoms, and data describing the efficacy of both direct and indirect surgical intervention, namely superficial temporal artery–middle cerebral artery (STA-MCA) bypass and encephaloduroarteriosynangiosis (EDAS) or encephaloduroarteriomyosynagiosis (EDAMS), abound in the literature.[3,10,18,21] The same approach for reduction in risk of recurrent hemorrhage is less well established and debated, although the efficacy, particularly for combined bypass, has been reported.[6,8,15] Management of aneurysms arising from deep collateral perforating vessels, however, poses a specific challenge and reports regarding their management are scarce. As with any

aneurysm, definitive and rapid obliteration is desirable to avoid recurrent or incident hemorrhage, but direct surgical obliteration is generally difficult or may risk ischemia through obliteration of the associated collateral vessels. Disappearance and resolution of deep collateral vessel aneurysms in MMD patients treated with bypass or with endovascular strategies has only been rarely reported.

In this report, we discuss a patient with MMD presenting initially with hemorrhage and with bilateral deep collateral vessel aneurysms. We outline our management, including the rationale for an endovascular versus surgical bypass approach to achieve aneurysm obliteration.

CASE REPORT

A 43-year-old female presented to the hospital after being found unresponsive with preceding complaints of headaches and dizziness. Upon arrival, a computed tomographic (CT) scan of the head revealed a deep right-sided hemorrhage in the region of the posterior limb of the internal capsule with extensive intraventricular extension [Figure 1a] and parenchymal extension. Her examination demonstrated dense left hemiparesis, affecting the arm more than the leg. An angiogram demonstrated findings of advanced MMD and a small 5 mm right distal thalamostriate branch aneurysm, which was consistent with the source of the hemorrhage [Figure 1b, c]. It was felt that the distal territory of this perforator branch had already been affected by the hemorrhagic event and, thus, no longer providing indispensable supply to a functional brain area. The aneurysm was accessed using a Marathon microcatheter (Covidien, Irvine, CA, USA) with the help of a Mirage guidewire (Covidien), and embolized with 20% *n*-butyl cyanoacrylate glue (n-BCA) (Cordis-Codman, Raynham, MA, USA) in ethiodol oil [Figure 1d]. This resulted in complete obliteration of the aneurysm along with the feeder vessel [Figure 1e]. No further intervention was pursued at that time given the absence of hemodynamic compromise and successful obliteration of the hemorrhage source. The patient demonstrated gradual neurological improvement and was discharged home with stable left hemiparesis, which improved over the course of a year to the ability to ambulate independently with 3/5 distal and 4/5 proximal left upper extremity strength.

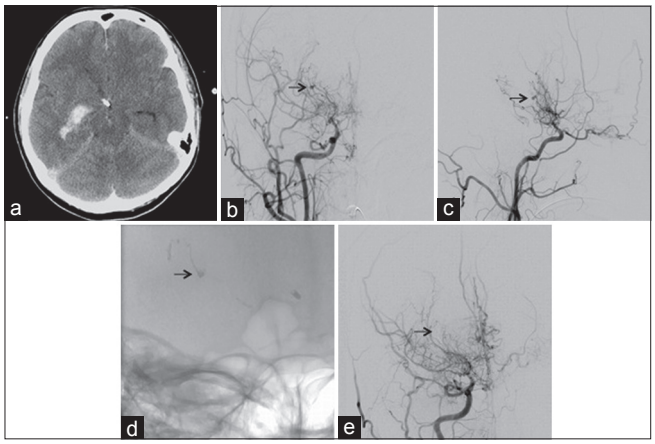

Figure 1: (a) CT scan of the brain demonstrating diffuse subarachnoid hemorrhage with intraventricular extension into the right temporal horn. (b, c) Right internal carotid artery (ICA) digital subtraction angiography (DSA) anterio-posterior (AP) view and lateral view, demonstrating advanced moyamoya disease with right middle cerebral artery (MCA) occlusion and collaterals originating from the ICA partially reconstituting the MCA. There is a 3-mm aneurysm seen on one of the thalamostriate vessels (arrow). (d) Fluoroscopic image of the skull (AP view) post *n*-BCA glue embolization of the thalamostriate aneurysm along with the feeding vessel. The aneurysm is filled with *n*-BCA glue cast (arrow). (e) AP view of right ICA DSA post *n*-BCA embolization. The aneurysm is completely obliterated (arrow: location of obliterated aneurysm)

Follow-up angiography was performed 1 year post embolization. The right-sided aneurysm remained well obliterated. The left internal carotid artery (ICA) injection showed occlusive changes in the supraclinoid ICA with typical moya collaterals, in addition to the finding of a pseudoaneurysm arising from a collateral vessel just lateral to the third ventricle. In retrospect, it was noted that the lesion had likely been present previously but had become substantially more prominent [Figure 2a, b]. Subsequently, embolization was attempted; however, the aneurysm was originating from a deep choroidal branch arising from the left posterior cerebral artery (PCA), which was seen to be directly reconstituting cortical vasculature, including

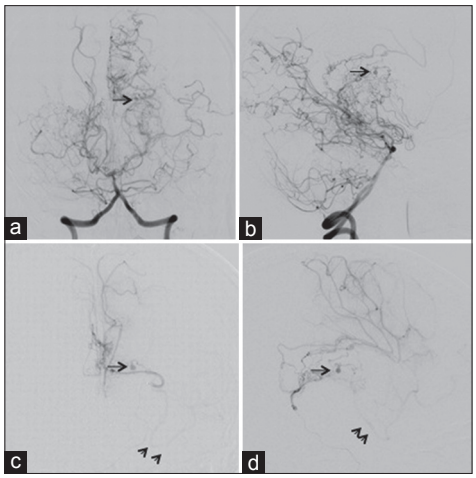

Figure 2: (a, b) Vertebral artery (VA) DSA AP and lateral views demonstrating an unruptured aneurysm (arrow) on a collateral from the left PCA to the left anterior cerebral artery (ACA) and left MCA. (c, d) Selective DSA imaging, AP and lateral views, through a microcatheter introduced through the left PCA. A small 3-mm aneurysm (long arrow) is seen supplied by the posterior choroidal branch of the PCA. There is reconstitution of ACA and MCA (short arrows) from the aneurysm feeding vessels

the distal MCA [Figure 2c, d]. Because of the risk of interruption of this collateral supply to the MCA, the aneurysm was not embolized.

Consequently, a surgical approach was entertained; direct obliteration through surgical trapping was felt to be ill advised given the deep location and the continued concerns regarding placing the territory supplied by the aneurysmal collateral at risk. Direct STA–MCA bypass and EDAS was planned with the specific intention of revascularizing the territory supplied by the collateral arising from the aneurysmal vessel and with the additional premise that subsequent embolization could then be pursued safely if the aneurysm did not resolve with revascularization alone. Intraoperatively, direct STA to MCA bypass was performed over the frontal cortical surface. The STA cut flow intraoperatively was 89 ml/min and the final bypass flow was 45 ml/min, indicating a successful bypass patency with a cut flow index of 0.5.[1] Angiogram in the initial postoperative period showed a patent bypass and stable appearance of the left distal PCA choroidal branch aneurysm [Figure 3a]. A follow-up angiogram was planned 6 months later which revealed that the bypass remained patent. In addition, further vascular contribution from the EDAS, just deep and adjacent to the craniotomy site, to the left frontal lobe was now observed. Furthermore, the deep choroidal aneurysm was no longer seen, indicating complete regression [Figure 3b, c].

DISCUSSION

The incidence of aneurysm formation within the moyamoya adult population is 3-14%, and represents a major potential hemorrhagic risk for these patients.[4,16,24]

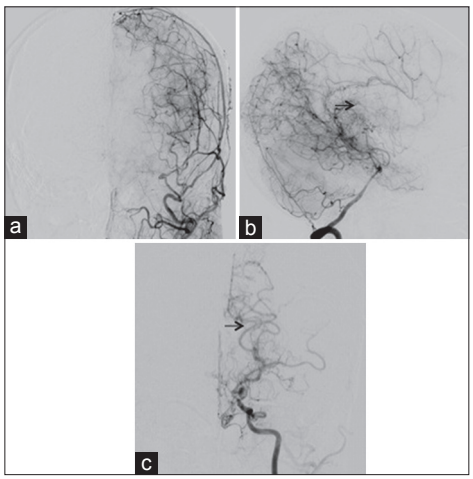

Figure 3: (a) Left external carotid artery DSA (AP view) post superficial temporal artery (STA) to MCA bypass. There is robust filling of the left MCA territories through the bypass graft. (b, c) Lateral VA DSA imaging and left ICA AP 7 months post left STA-MCA bypass showing complete regression of the aneurysm (arrow; site of the obliterated aneurysm)

The formation of aneurysms in MMD is multifactorial and includes features related to hemodynamic stress and pathologic vessel architecture. More than half of these aneurysms can be considered saccular aneurysms developing in association with major arteries that make up the circle of Willis (COW). Others arise from deep perforator collateral branches and are typically considered to be pseudoaneurysms.

The saccular aneurysms are presumed to be indirect sequelae of the stenosis and occlusion encountered in MMD, which results in circulatory changes within the COW and increased hemodynamic stress on associated vessels. These are typically more anatomically accessible for both endovascular and surgical approaches than the perforator pseudoaneurysms, although underlying stenosis of the vasculature can potentially prohibit endovascular access. A study conducted by Yu *et al.* evaluated the endovascular approach to aneurysms in MMD patients. Their cohort included both saccular aneurysms ($n = 10$) and pseudoaneurysms ($n = 3$). Nine of ten saccular aneurysms were successfully embolized with endovascular coiling techniques; one basilar trunk aneurysm associated with a pontine branch could not be safely accessed, in part due to stenosis within the basilar artery inhibiting advancement of the guidewire. They concluded that there is a favorable prognosis with endovascular treatment, but emphasized careful patient selection in regards to the specific site of the aneurysm.[25]

Although a majority of aneurysms are associated with the COW, an estimated 44% arise from either choroidal, meningeal, or moyamoya vessels. Pseudoaneurysms arising from these collateral vessels are subject to high flows, compensating for the lack of flow in the affected ICAs. The presumed pathogenic mechanism involves increased flow force through these much smaller vessels, which lack the muscular strength of larger COW vessels. In MMD, histological changes in the large vessels of the COW include intimal thickening and excessive smooth muscle proliferation, whereas in the moyamoya vessels, fragmented elastic lamina, thinned media, and both excessively thin and thick areas of the intima produce an abnormally shaped characteristic wavy lumen. Microaneurysmal formation is commonly seen in conjunction with these pathologic vessel features, and can thus predispose to pseudoaneurysm formation.[20,22]

Benefits to endovascular management of collateral vessel pseudoaneurysms are related to difficulties in direct surgical access and localization of these aneurysms, which are typically located in deep eloquent areas. Furthermore, manipulation of cerebral vessels in MMD patients involves higher risk in general, due to the increased friability of the vasculature. Although the endovascular approach may provide easier localization,

vessel tortuosity can be prohibitive as encountered in one of three cases attempted for endovascular obliteration by Yu *et al*.[25] Accessibility is not the only consideration for management of these lesions. The endovascular strategy relies primarily on occlusion of the pseudoaneurysm and its feeding vessel with liquid embolic agents such as onyx (ev3, Irvine, CA, USA) or *n*-BCA.[14] Due to the territory supplied distally via the moyamoya collaterals, a proximal embolization to address the aneurysm may prove costly to collateral flow, leading to distal ischemia. This was the situation with the yet unruptured but growing left-sided pseudoaneurysm in the case illustrated in this report. The proximal portion of the artery harboring the aneurysm could be reached endovascularly, but was believed to be too risky to obliterate given the distal territory supplied. In such cases, superselective Amytal injection has been reported as one method to discern the safety of embolization.[7] We refrained from Amytal testing, as it was felt that any sacrifice of major collateral circulation would increase the risk of subsequent, if not acute, stroke. On the other hand, endovascular treatment of the right-sided aneurysm presenting with hemorrhage posed no significant risk of further deficit, as the distal territory had already been affected by the hemorrhage.

Another strategy for deep pseudoaneurysms is an indirect surgical approach involving distal revascularization, aimed at reducing the hemodynamic stress through the deep collaterals. However, aneurysm resolution in MMD patients following surgical STA-MCA bypass, with or without EDAS, has only rarely been reported [Table 1], and our case represents the first report of an unruptured pseudoaneurysm treated in this manner. A surgical bypass approach (both direct and indirect) is generally considered the primary method of management to slow progression of the disease, alleviate ischemia, and prevent future hemorrhagic events.[18,20] However, it has only infrequently been described as a treatment option for existing pseudoaneurysms. Ni *et al*. recently described two cases in which a combined direct and indirect surgical bypass was followed by resolution of ruptured distal artery pseudoaneurysms [Table 1]. Both cases showed resolution at their first post discharge imaging, ranging from 5 to 10 months.[16] Kuroda *et al*. reported three cases in which there was resolution of pseudoaneurysms arising from the thalamoperforating, posterior choroidal, and anterior choroidal vessels in moyamoya patients following STA–MCA bypass with EDAMS.[13] Follow-up imaging showed resolution, ranging from 1 to 3 months postoperatively [Table 1]. Both studies report a favorable outcome not only in terms of prevention of disease progression and symptomatic relief, but also for definitive treatment of the aneurysms.[13,16] As in the reported cases, we opted for a combined direct and indirect bypass strategy in treating our patient; the rationale for this approach is based on the STA–MCA bypass providing an immediate revascularization to quickly reduce hemodynamic stress on the pseudoaneurysm. The addition of an EDAS, over time, stimulates further ongoing superficial to deep angiogenesis to optimize revascularization.[3,10,18,20]

Although our case and others have demonstrated revascularization as a successful strategy for treatment of deep collateral vessel aneurysms, it is important

Table 1: Summary of published cases of surgical revascularization for treatment of moyamoya disease associated collateral vessel aneurysms

Study	Age (years), sex	Presentation	Location of aneurysm	Treatment	Angiographic outcome
Current study	43, F	Incidental, with angiographic growth	Collateral artery arising from choroidal branch of L PCA	L STA–MCA+EDAS	Angiogram at 6 months showed complete resolution
Ni *et al.*, 2012[16]	46, M	Hemorrhage	Moyamoya collateral arising from R M1	R STA–MCA+EDMS	Angiogram at 6 months demonstrated resolution
	29, F	Hemorrhage	Collateral artery arising from the R AChA artery	R STA–MCA+EDMS	Angiogram at 10 months demonstrated resolution
Kuroda *et al.*, 2001[13]	60, F	Hemorrhage	Thalamoperforating artery arising from the R PcoA artery	R STA–MCA+EDMAS	Angiogram at 3 months demonstrated resolution
	33, F	Hemorrhage	Collateral artery arising from the R PChA	R STA–MCA+EDMAS	Angiogram at 1 month demonstrated resolution
	50, F	Hemorrhage	Collateral artery arising from the R AChA.	R STA–MCA+EDMAS	Angiogram at 3 months showed resolution
Otawara *et al.*, 2007[17]	60, M	Hemorrhage	Plexal segment of the R AChA	Bilateral STA–MCA+EDAMS	Angiogram at 10 years showed persistence of aneurysm without change in size

AChA: Anterior choroidal artery, EDAMS: Encephaloduroarteriomyosynagiosis, EDAS: Encephaloduroarteriosynangiosis, EDMS: Encephaloduromyosynagiosis, L: Left, PCA: Posterior cerebral artery, PChA: Posterior choroidal artery, PcoA: Posterior communicating artery, R: Right, STA–MCA: Superficial temporal artery-middle cerebral artery bypass. Not included are five collateral vessel aneurysms treated by revascularization reported by Kawaguchi *et al.*,[9] as no detail of individual cases or outcome has been reported

to note that this strategy is not universally effective. Otawara *et al*. have reported one case in which a microaneurysm persisted for 10 years despite surgical revascularization and ultimately led to hemorrhage.[17] Follow-up short-term angiography to confirm obliteration is, therefore, mandatory, and based on existing literature, should be expected by 6-12 months postoperatively. Failure of obliteration by this timeframe warrants pursuit of endovascular embolization, which can be performed with a presumed lower risk of ischemia given the surgical revascularization of the territory at risk.

The possibility of spontaneous resolution without the aid of intervention must also be considered. In non-MMD patients, reports of spontaneous resolution of deep pseudoaneurysms are rare and only postulated in a limited number of case reports.[12,23] However, multiple instances of spontaneous resolution of ruptured collateral vessel aneurysms in MMD patients have been reported.[9] Yu *et al*. described spontaneous resolution of an aneurysm after failed endovascular intervention: A ruptured perforating artery pseudoaneurysm arising from the MCA disappeared on 1-week angiogram and the patient suffered no further hemorrhagic events over 2-year follow-up, although it is unclear if follow-up angiography was performed to confirm sustained resolution of the aneurysm.[25] Grabel *et al*. reported regression (but not complete disappearance) of a ruptured lenticulostriate pseudoaneurysm on 3-week angiogram.[5] Kodama *et al*. reported spontaneous resolution of collateral vessel ruptured pseudoaneurysms in three patients, based on repeat angiography ranging from 42 days to 11 months post ictus.[11] Arai *et al*. reported one case of ruptured thalamoperforator vessel pseudoaneurysm, which disappeared without re-rupture on 1-year angiogram.[2] All these reports, however, involve aneurysms which were discovered post-rupture and were resolved by the time of follow-up angiography. In our case, persistence and increased prominence of the pseudoaneurysm over the course of a year prior to treatment would strongly indicate that ultimate resolution was in fact a result of revascularization rather than a spontaneous phenomenon.

CONCLUSION

When considering treatment of collateral vessel aneurysms in MMD, the efficacy, timeframe and potential risk for ischemia or recurrent/incident hemorrhage must be considered in weighing endovascular and surgical approaches. Although bypass procedures are already considered a primary approach for prevention of future ischemic events in MMD patients, our case adds to the small number of reported cases that support its efficacy for treatment of collateral vessel pseudoaneurysms as well. The lag time to resolution of aneurysm post bypass, however, tends to favor the more immediate obliteration, which can be achieved with endovascular embolization in the setting of hemorrhage, as long as this can be performed without incurring additional deficit. Although spontaneous resolution following rupture has been reported in a small number of cases, it occurs unpredictably, and conservative management incurs the risk of re-hemorrhage.

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