CASE REPORT

External carotid stenting for symptomatic stenosis in a patient with patent EDAS for Moyamoya disease

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SUMMARY

Background Moyamoya disease is characterized by progressive narrowing of the internal carotid artery (ICA). Symptomatic patients typically undergo cerebrovascular intervention via extracranial–intracranial (EC–IC) bypass, most often with the use of the superficial temporal artery. This case of Moyamoya disease is of particular interest as the patient presented with a unilateral atherosclerotic external carotid artery (ECA) stenosis after EC–IC bypass that eliminated the benefit of his original surgery, resulting in a symptomatic presentation.

Clinical presentation A 53-year-old man presenting with Moyamoya disease and known left ICA occlusion had received a bilateral encephaloduroarteriosynangiosis (EDAS) bypass 10 years previously. He re-presented complaining of right-sided tingling, weakness, and numbness radiating up the arm. CT angiography indicated significant stenosis of the left ECA. ECA angioplasty and stenting with a distal protection device resulted in resolution of his symptoms.

Conclusions This case illustrates that a patient presenting with Moyamoya disease and concurrent symptomatic ECA stenosis post-EDAS can be effectively and safely treated with ECA stenting.

BACKGROUND

Moyamoya is a disease typically requiring surgical intervention through direct or indirect extracranial–intracranial (EC–IC) bypass. Although the etiology of Moyamoya disease has not been entirely elucidated, it is believed to be a heritable genetic condition. It is idiopathic and the pathogenesis involves progressive narrowing of the supraclinoid section of the internal carotid artery (ICA), resulting in ischemic cerebral tissue secondary to vessel occlusion. As a result of ICA narrowing, the intracerebral vessels will form insubstantial and porous collaterals in an attempt to increase perfusion. These collaterals present a hemorrhagic risk in addition to the aforementioned ischemic risk, predisposing the patient to a cerebrovascular accident. Treatment involves direct (superficial temporal artery–middle cerebral artery; STA–MCA) or indirect encephaloduroarteriosynangiosis (EDAS) EC–IC bypass to create substantial anastomoses, thereby increasing perfusion. These surgical approaches require that the external carotid artery (ECA) be patent in order to ensure adequate blood flow through the bypass. Thus, when a patient presents with ECA stenosis after EC–IC bypass, the benefit of the original treatment is reduced or eliminated. A search of the currently available literature showed that there have been no reported cases of

individuals with Moyamoya disease receiving EDAS and subsequently presenting with symptomatic ECA stenosis. This case is unique and provides an insight into the viable treatment options for patients who present similarly.

CASE PRESENTATION

The patient was a 53-year-old man with Moyamoya disease who had presented 10 years previously with bilateral ICA stenosis and a known left ICA occlusion with Moyamoya features. He originally received bilateral EDAS in order to increase perfusion and had been asymptomatic since that time on aspirin. Approximately 1 month prior to presentation the patient reported intermittent episodes of tingling, numbness, and weakness in his right arm. The numbness progressed up his right arm in a non-dermatomal distribution. He also struggled with fine motor skills. Neurological examination demonstrated 4+/5 weakness of the right hand intrinsics, extensors, and grip, with difficulty in fine motor movement of the fingers.

INVESTIGATIONS

A CT angiogram suggested >70% proximal stenosis of the left ECA which was thought to be atherosclerotic in nature. [Figure 1](#) shows pretreatment imaging of the left head, illustrating decreased perfusion through the pre-established EC–IC bypass.

Figure 1 Lateral angiogram of the left head illustrating patency of the extracranial–intracranial bypass with decreased perfusion through collaterals.

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Figure 2 Anterior-posterior (A) and lateral (B) angiogram of the left common carotid artery, confirming 70% stenosis of the external carotid artery.

The angiogram also confirmed a 70% stenosis of the left ECA origin (figure 2A, B).

TREATMENT

The patient underwent ECA angioplasty and stenting with a distal protection device in order to increase vessel patency and flow while on aspirin and clopidogrel (figure 3A–C).

OUTCOME AND FOLLOW-UP

The patient remains asymptomatic 1, 3, and 6 months after surgery with complete resolution of original symptoms.

DISCUSSION

Carotid endarterectomy and angioplasty with stenting are routinely performed for ICA stenosis secondary to atherosclerosis.

Typically, these interventions are indicated when a patient presents with symptoms indicative of ischemic pathology in the presence of severe vessel stenosis. In general, the relative risks and benefits of endarterectomy and stenting are mostly similar, with both serving as viable means of treating carotid stenosis.¹ The primary concern in these patients is the lack of blood flow through the ICA resulting in a nidus for thromboembolic events as well as a source for decreased cerebral perfusion. ECA patency and flow, however, become critical for cerebral perfusion in individuals who rely on EC–IC collaterals. This is the case for a patient with Moyamoya disease, who depends on such collaterals to maintain cerebral perfusion. Under these circumstances, the ECA acts as their ICA, and any narrowing or occlusion could result in significantly decreased cerebral perfusion. Retrospective review has further established the role of

Figure 3 Anterior-posterior (A) and lateral (B) imaging of the left common carotid artery post-angioplasty and stenting. Stenosis has been alleviated. Imaging of the left head (C) illustrates increased perfusion through the extracranial-intracranial bypass after treatment.

Table 1 Reports of external carotid artery stenosis

Study ID	Indication	Disease	Treatment	Outcome/follow-up
Reid <i>et al</i> ³	3 patients with ICA occlusion and ipsilateral ECA stenosis	Atherosclerosis	PTAS for 2 patients, endoluminal graft for 1 patient	All 3 patients asymptomatic with no restenosis at follow-up
Eisenberg <i>et al</i> ⁴	Occluded left ICA with ipsilateral ECA stenosis	Atherosclerosis	PTAS	Asymptomatic with no restenosis at 6 months post-intervention
Schiller <i>et al</i> ⁵	Bilateral ECA stenosis with jaw claudication/neck pain	Atherosclerosis	PTAS with subsequent endarterectomy	Bilateral restenosis at 6 months post-PTAS—redilated at this time; return of symptoms and restenosis at 11 months post-redilation; asymptomatic 3 years post-op
Adel <i>et al</i> ⁶	Bilateral ICA occlusion with right ECA stenosis (90%)	Atherosclerosis	PTAS	Asymptomatic with no restenosis at 20 months post-intervention
Kawamata <i>et al</i> ⁷	7 patients with ICA occlusion and ipsilateral ECA stenosis	Atherosclerosis	Endarterectomy	All asymptomatic with no restenosis at follow-up (range 18–53 months, mean 35.6 months)
Ko <i>et al</i> ⁸	ICA occlusion with ipsilateral ECA stenosis	Atherosclerosis	PTAS	Asymptomatic with no restenosis at >1 year post-intervention
Turtzo <i>et al</i> ⁹	ICA occlusion with ipsilateral ECA stenosis	Atherosclerosis	PTAS	Mild ECA restenosis at 10 months, asymptomatic at 10 months and 1.5 years post-intervention
Xu <i>et al</i> ¹⁰	12 patients with ICA occlusion and ipsilateral ECA stenosis	Atherosclerosis	PTAS	2 of 12 patients with restenosis at ≥5 months post-intervention
Oku <i>et al</i> ¹¹	ICA occlusion with ipsilateral ECA stenosis	Atherosclerotic	PTAS	Asymptomatic at 2 months post-intervention
Kouvelos <i>et al</i> ¹²	Bilateral ICA occlusion with bilateral ECA stenosis	Atherosclerosis	PTAS	Asymptomatic with no restenosis at 1 year post-intervention
Fokkema <i>et al</i> ¹³	27 patients with ICA occlusion and ipsilateral ECA stenosis	Atherosclerosis	Endarterectomy	9 patients dead at follow-up; At 3 years post-op 83% were asymptomatic with 80% of operated arteries patent; 5 patients developed ≥50% restenosis
Lee and Ahn ¹⁴	Bilateral ICA occlusion with bilateral ECA stenosis	Possible Moyamoya or atherosclerosis	PTAS	Asymptomatic at follow-up

ICA, internal carotid artery; ECA, external carotid artery; PTAS, percutaneous transluminal angioplasty and stenting.

ECA patency in individuals with ICA occlusion, with data indicating that ECA stenosis of >70% acts as an independent risk factor for symptomatic presentation.² Under these circumstances, endarterectomy or stenting of the ECA is clearly indicated.

Table 1 shows reports and reviews in which patients received ECA intervention, including endarterectomy or stenting.^{3–14} The majority of patients receiving surgery or angioplasty had concurrent symptomatic ipsilateral ICA occlusion from underlying atherosclerotic disease. Presentation included signs and symptoms of ischemic stroke, transient ischemic attack, and amaurosis fugax. Xu *et al* provide a retrospective analysis of individuals with symptomatic ICA occlusion and ECA stenosis (most with >70%) who were successfully treated with ECA angioplasty and stenting.¹⁰ Similarly, Fokkema *et al*¹³ performed a retrospective review of patients with ICA/ECA occlusion receiving ECA endarterectomy, deeming the procedure safe and effective for resolving neurological symptoms. Adel *et al*,⁶ Eisenberg *et al*,⁴ and Reid *et al*³ provide case reports of individuals with symptomatic ICA occlusion with improvement of symptoms after ECA stenting. These reports support either ECA stenting or endarterectomy in the face of concurrent ICA occlusion as a viable means of managing ipsilateral ICA occlusion. The reports by Ko *et al*,⁸ Kawamata *et al*,⁷ and Oku *et al*¹¹ are particularly relevant as they reference patients who received ECA intervention as a preventative before undergoing STA–MCA bypass. While our patient had different circumstances, the similarity in pathophysiological presentation and successful treatment of these patients supported our therapeutic decision-making.

Lee and Ahn¹⁴ published a report of a patient with symptomatic Moyamoya disease with no prior surgical treatment. He was found to have proximal ECA stenosis and was treated successfully by ECA angioplasty/stenting. In this patient, the etiology of the ECA stenosis was unclear.

Literature expanding upon the true involvement of extracranial arteries in Moyamoya disease is sparse and often limited by a lack of conclusive data. Häckel *et al*¹⁵ provide a case of a patient with Moyamoya disease and suspected involvement of the extracranial vertebral arteries ‘diagnosed’ with angiography which revealed ‘Moyamoya-vessels’ in carotid territories supplied by the vertebrobasilar system. Weber *et al*¹⁶ demonstrated involvement of multiple peripheral vessels in a patient with Moyamoya disease. This was supported by duplex ultrasound and MR angiography showing concentric wall thickening and diffuse luminal narrowing in the absence of systemic inflammation. Yamada *et al*¹⁷ published a report indicating the presence of renal artery stenosis in 7 of 86 patients with Moyamoya disease, a number found to be ‘considerably higher than that in the corresponding general population’. Ikeda¹⁸ provided data supporting systemic involvement through biopsy of extracranial vessels (pulmonary, renal, and pancreatic) in patients with Moyamoya disease showing significant intimal thickening relative to age with histopathology similar to that seen in classic Moyamoya disease. Li *et al*¹⁹ further demonstrated this Moyamoya-like histopathology in extracranial arteries by biopsy of the STA in 15 patients with Moyamoya disease. Aoyagi *et al*²⁰ provided compelling evidence for extracranial involvement in a study of 17 patients with Moyamoya disease aged <20 years who had Moyamoya-like intimal thickening in the

STA while a control group of seven patients aged <20 years had no intimal thickening. Hoshimaru and Kikuchi²¹ reviewed angiograms of 66 patients with Moyamoya disease and found stenosis within the distal ECA branches (superficial temporal, middle meningeal, and occipital arteries) in 13 (20%), although tissue samples were not collected to confirm a definitive etiology. On the other hand, Komiyama *et al*²² reviewed 39 angiograms and concluded that Moyamoya occlusion does not occur outside the intracranial arteries, but they conceded that stenosis of short segments of the ECA system may develop infrequently, probably due to differing susceptibilities from those causing ICA stenosis. The limited evidence available seems to indicate that Moyamoya-like changes may indeed occur outside the intracranial arteries, including within the ECA system, but it is not conclusive.

In our patient, however, the stenosis occurred at the origin of the ECA. Even the extensive report by Hoshimaru and Kikuchi only described distal ECA branch stenosis. Furthermore, our patient was symptomatic. Most of the reports supporting systemic Moyamoya involvement simply described small asymptomatic lesions or Moyamoya-like histopathological changes within the extracranial vessels found on autopsy, which is distinctly different from our patient's symptomatic stenosis. Importantly, the patient with Moyamoya disease described by Lee and Ahn had symptomatic proximal ECA stenosis but the etiology of his steno-occlusion was unclear and unsubstantiated, providing little support to the idea of proximal ECA involvement in Moyamoya disease. Our patient also had several risk factors for atherosclerosis, including age, a long history of cigarette smoking, and significant dyslipidemia. Given these significant risk factors and his symptomatic presentation, it is most likely that the etiology of his stenosis was primarily atherosclerosis, perhaps exacerbated by hemodynamic changes secondary to his past EDAS. Without a tissue biopsy, Moyamoya-like changes cannot be conclusively ruled out but it is unlikely when

framed within the available literature and clinical presentation. In patients with Moyamoya disease managed by EC-IC bypass, the status of the ECA origin is vital to continued adequacy of collateral circulation and stenosis of any etiology can be disabling. Our patient presented under unique circumstances—namely, post-bilateral EDAS with delayed symptomatic atherosclerotic stenosis of the ECA origin. The left ECA stenosis made definitive treatment necessary. In the setting of his Moyamoya disease, obstruction of flow for any period of time could have induced permanent ischemia. We therefore successfully performed angioplasty/stenting to forgo the need for temporary clamping. In patients who have undergone bypass, the ECA effectively becomes their ICA, and stenotic pathology should be addressed with that in mind.

Contributors

Competing interests None.

Patient consent Obtained.

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Learning points

- Patients with bypassed Moyamoya disease who develop late recurrence of ischemic symptoms require evaluation of their external carotid artery (ECA), with treatment of atherosclerotic stenosis as indicated.
- ECA patency and flow becomes critical for cerebral perfusion in individuals who rely on extracranial–intracranial collaterals. This is the case for a patient with Moyamoya disease who depends on such collaterals to maintain cerebral perfusion.
- Literature on the involvement extracranial vessels in Moyamoya disease, especially ECA vessels, is limited. It is likely that Moyamoya-like changes exist in vessels outside the cranium, but few reported cases have resulted in symptomatic presentation due to these extracranial changes.
- The endovascular treatment of this unique patient, who presented post-bilateral EDAS with delayed symptomatic atherosclerotic stenosis of the ECA origin, can be extrapolated to similarly complicated cases, adding an element of depth and complexity to the literature currently available on carotid and cerebral arteriopathies.
- A patient presenting with Moyamoya disease and concurrent symptomatic ECA stenosis post-EDAS can be effectively and safely treated with ECA stenting.

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