

Case Reports & Case Series

Angioplasty and stenting of adult onset Moya-moya disease

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ABSTRACT

Moya moya is a disease that typically affects children and is treated with extra-cranial to intracranial bypass in suitable patients. We present an adult 28-year-old patient with Moya-moya disease who had a stepwise progression of the disease and was treated with angioplasty and stenting of the stenosis bilaterally. This novel treatment method allowed her posterior circulation sufficient time to collateralize the areas in the anterior circulation which had a deficient blood supply. After the procedure, the patient has been asymptomatic for the last 5 years despite developing proximal anterior circulation occlusions bilaterally. This is the first description of angioplasty and stenting in Moya-moya disease. Although this strategy does not treat the underlying cause, it is potentially effective temporizing measure that could allow the brain to develop alternative blood supply or allow a bypass operation time to mature in an urgent setting.

1. Angioplasty and stenting of adult Moya-moya disease

A 28-year-old Caucasian male with a history of depression presented with a sudden onset of left sided numbness and facial droop. He had no diabetes, hypertension or other cardiovascular risk factors, neither did he have any family history of stroke or heart problems. A non-contrast CT scan was unremarkable, but a CT-angiogram revealed a severe right terminal-ICA stenosis (TICA). This was further investigated with a conventional angiogram and a CT-perfusion which confirmed the stenosis causing reduced distal perfusion without infarction (Fig. 1A–F). In view of the unilateral lesion and age group, a diagnosis of Moya-moya syndrome was considered [1], however as the symptoms were acute, urgent angioplasty and stenting was performed with a Gateway 4 × 20 mm balloon and a Wingspan stent 4 × 20 mm (Boston Scientific, Fremont, California) (Fig. 2A–D). This was done with a view towards a superior temporal artery-to-middle cerebral artery (STA-MCA) bypass being performed as a definitive treatment at a later date. The procedure was successful with immediate relief of symptoms and he was discharged home on dual antiplatelets and statins. He was well on follow-up and elected to delay his STA-MCA bypass despite advice against it. A year later he

suffered from 2 generalized tonic-clonic seizures and recurrent TIAs manifesting as transient right hemiplegia. This was due to a new stenosis which had developed in the M1 region of the left MCA (Fig. 3A–D). This was again, but now on the contralateral side, stented open with a Wingspan stent 4 × 20 mm with immediate relief of his symptoms (Fig. 4A–C). The patient has been asymptomatic for the last 5 years after his last intervention, but had poor compliance with medication despite counselling. Serial vascular and perfusion imaging showed a progressive stenosis of the bilateral proximal anterior circulation vasculature. The most recent scans revealed a right T-ICA and left mid M1 occlusion with no perfusion defects (Fig. 5A–D), due to the posterior circulation having had sufficient time to accommodate the ischemia and collateralize the anterior circulation (Fig. 5E–H).

2. Discussion

This is the first description of Moya-moya disease treated with angioplasty and stenting. It was effective in giving the brain time to collateralize and adequately perfuse the at-risk telencephalon via the posterior circulation and the transdural supply to a smaller extent

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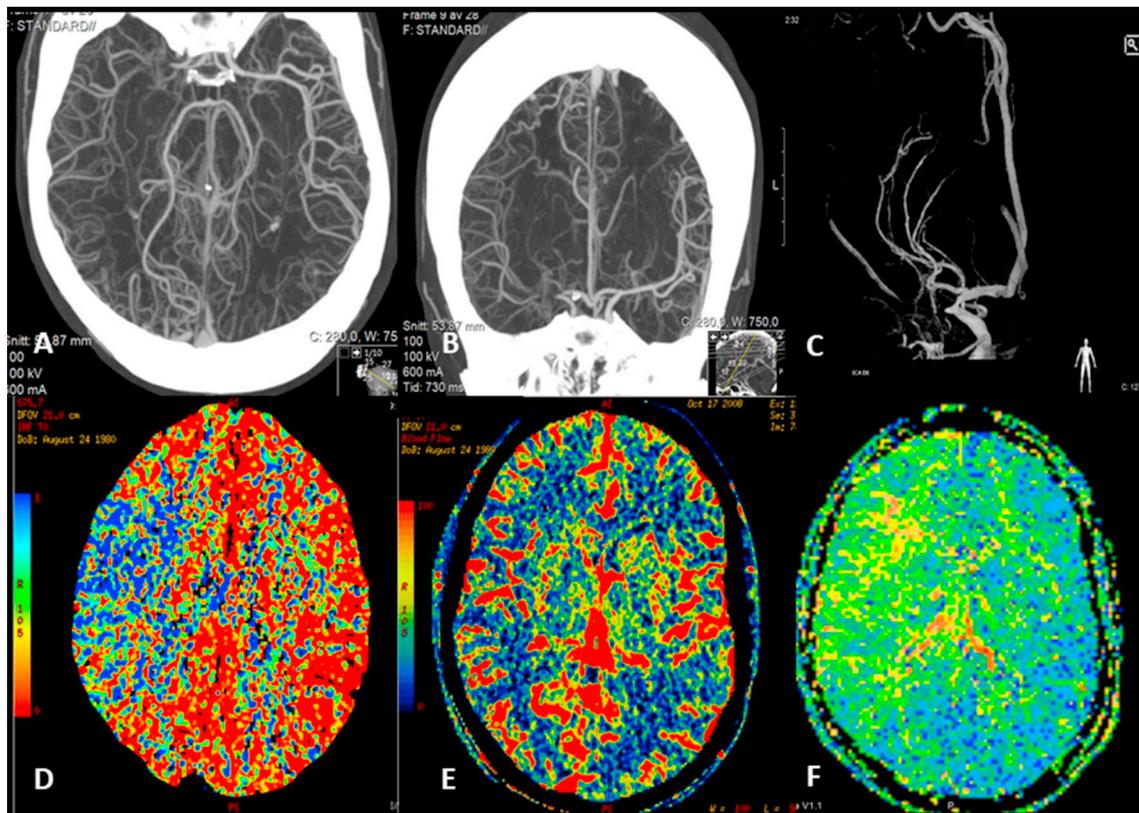


Fig. 1. A - Computed tomography axial MIP projections showing a terminal right ICA tight stenosis with lenticulostriate proliferation, B - Similar findings in coronal view, C - 3-dimensional reconstruction of DSA demonstrating right T-ICA near occlusion, D - CT-perfusion showing prolonged right MCA territory mean transit time, E - the cerebral blood flow is relatively well preserved, F - while the blood volume in the corresponding MCA territory is increased.

(Supplementary Fig. 1).

Moya-moya syndrome is a vascular disease of the cerebral vessels that predisposes patients to stroke from progressive stenosis of the intracranial terminal internal carotid arteries and the proximal MCA. Although it mainly involves the anterior circulation of the brain, in some cases it can affect the posterior circulation especially the basilar and posterior cerebral arteries. Patients with unilateral pathology are typically described to have the Moya-moya syndrome rather than Moya moya disease, even without other associated cardiovascular risk factors to account for the stenosis [1]. In Moya-moya syndrome with initial unilateral disease, the contralateral side is eventually involved in up to 40% of patients [2,3].

The incidence of the disease peaks at approximately 5–10 years of age and again in adults in their mid-40s [4]. There is a female predisposition to the disease with approximately twice as many female patients [4]. Our patient presented outside the typical age-range at the beginning of his disease.

Moya moya typically presents with headache due to dilated transdural blood supply [5], although ischemic symptoms or haemorrhagic complications are commonly seen as well. In rarer scenarios, patients with moya moya may also have more unusual symptoms at presentation such as seizures, or personality changes which may lead to the patient initially presenting to psychiatric specialists [6].

The vessels affected in Moya-moya syndrome show signs of smooth muscle hyperplasia with intramural thrombus formation rather than the typical signs of cerebral vasculature stenosis such as atherosclerotic changes or inflammatory signs on MRI imaging [7,8]. On histological examination the media of the vessel is often thinner, and the elastic lamina has an irregular appearance [8].

Treatment for Moya-moya disease in children is typically by an extracranial to intracranial bypass or encephaloduroarteriosynangiosis however, this is still somewhat controversial in adults [9]. Angioplasty and stenting has not been described in the literature for Moya-moya. In our case it did not provide a definitive treatment and the stenosis re-occurred, however was an effective way to immediately restore flow to the cerebral circulation. In our patient the strategy of angioplasty and stenting provided time to allow the development, via non-sprouting neoangiogenesis, of pial collaterals principally via the posterior circulation. It should be noted that the risk for arterial damage from the stent and dilation is not well understood.

A potential risk of this strategy is the possibility of haemorrhage with the antiplatelet treatment required with an implanted stent, moreover a portion of Moya-moya disease presents with haemorrhage. However, the authors believe that it is the friable vessels which proliferate after the stenosis occurs which are the cause of the increased haemorrhage risk. If the stenting is performed early in the disease, such as this patient, it could potentially prevent the vessel proliferation while the other vascular territories collateralize the ischemic areas.

A small percentage of Moya-moya disease involves the posterior circulation as well rendering this strategy ineffective. Early extracranial-to-intracranial bypass is still the established recommended treatment.

3. Conclusion

Treatment for adult onset Moya-moya disease is still controversial. We show a case of Moya-moya disease where the ischemic onset was successfully delayed with angioplasty and stenting. While not

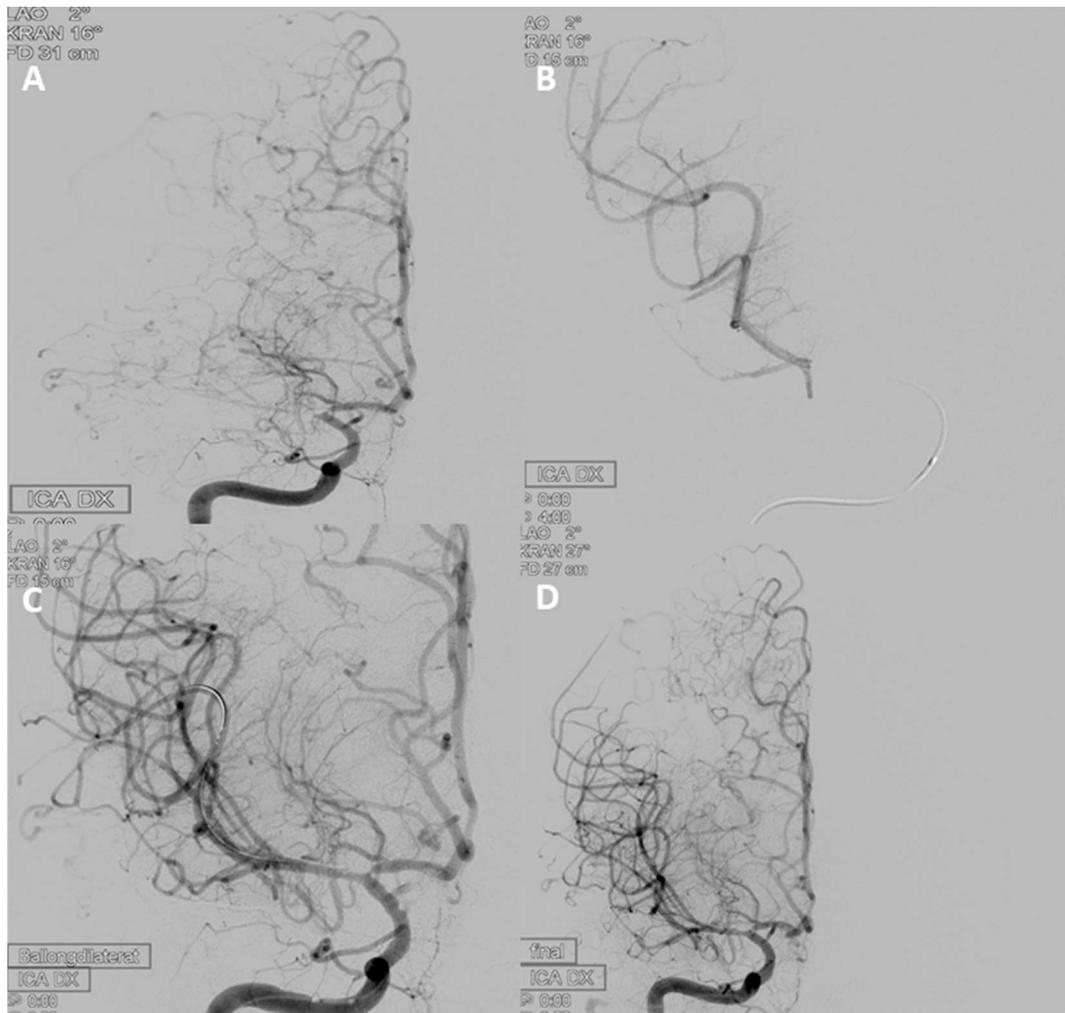


Fig. 2. A - DSA showing a right terminal ICA occlusion, B - Distal angiographic run performed after the occlusion is crossed with a microcatheter, C - After angioplasty of the occlusion with a Gateway 4 × 20 mm balloon, D - After stenting with a 4 × 20 mm Wingspan stent.

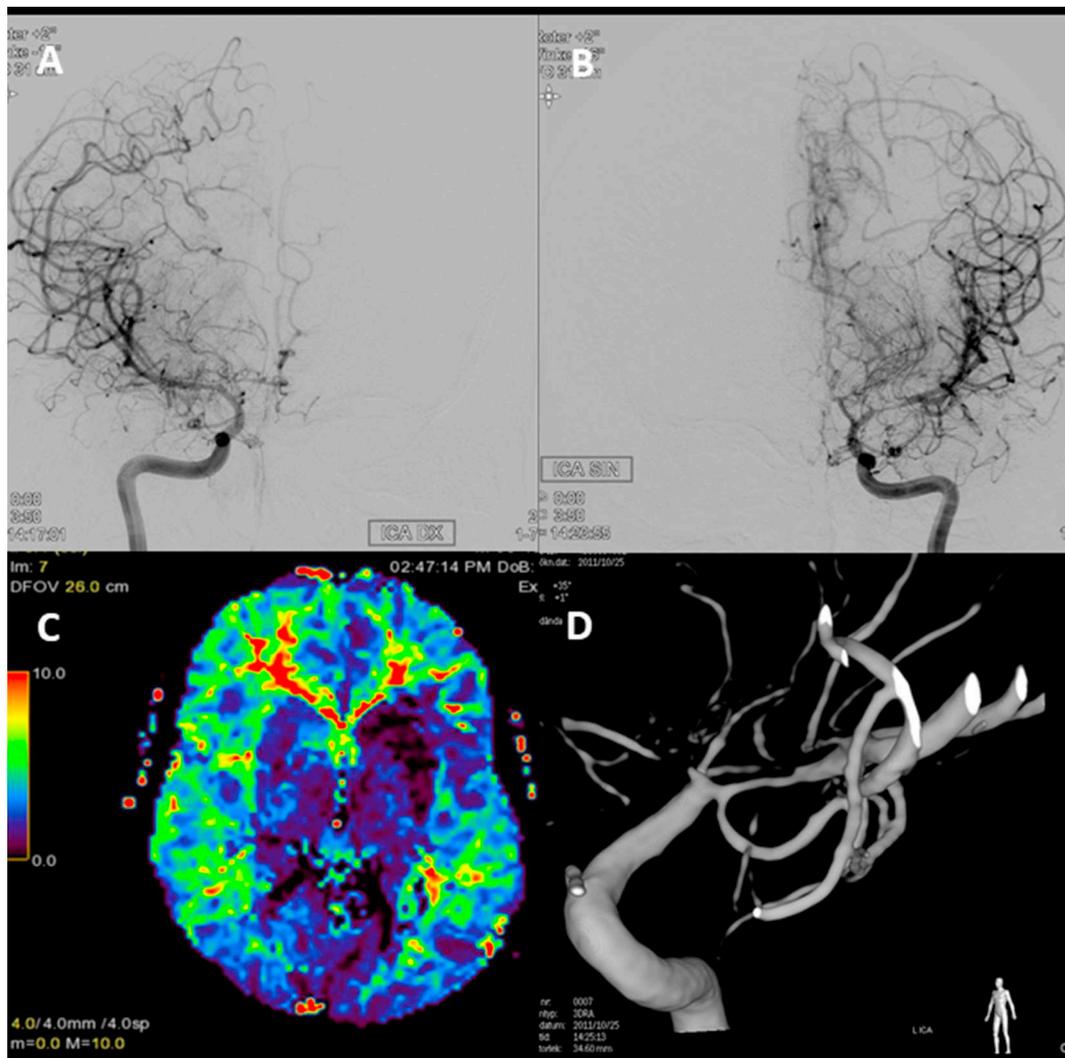


Fig. 3. A – DSA showing the right TICA is still open, B - however the left M1 has now developed a tight stenosis. C - CT-perfusion shows a corresponding reduction in the cerebral blood flow in the region. D - 3-dimensional reconstruction of the angiogram of the stenotic region.



Fig. 4. A – DSA showing the pre-treatment M1 stenosis, B - Angiographic run after angioplasty of the stenosis, C - Final run after stenting with a 4 × 20 mm Wingspan stent with relief of his symptoms.

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