

## CASE REPORT

# Imaging of disease progression in a case of idiopathic moyamoya

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Moyamoya is a rare cerebrovascular disease characterised by progressive stenosis of the terminal portion of the internal carotid artery and its main branches. We report on the progression of the disease in a 3-year-old boy, confirmed to have idiopathic moyamoya disease, over 6 ensuing years, using serial magnetic resonance imaging with MR angiography. Comparison is also made with conventional angiography at the last visit. Characteristic imaging appearances of asymmetric narrowing of the internal carotid arteries (especially the supraclinoid portion) with numerous collateral vessels around the brainstem, especially within the ambient and quadrigeminal plate cisterns, are demonstrated with progression.

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Moyamoya is a rare cerebrovascular arteriopathy characterised by progressive stenosis of the terminal portion of the internal carotid artery (ICA) and its main branches, and is associated with cerebral ischaemia. The moyamoya vessels are the fragile collaterals formed in compensation for this progressive stenosis of the ICA at the base of the brain (at the level of the Circle of Willis). These collaterals of the thalamostriate and lenticulostriate arteries give the characteristic appearances on MR or conventional angiography. Aetiology can be varied, and numerous causes can result in secondary moyamoya-like collateralisation. Idiopathic or primary moyamoya is rare; diagnosis is usually made with MRI and after exclusion of all possible secondary causes. We present progressive imaging of a 3-year-old boy confirmed to have idiopathic moyamoya disease, over 6 years using serial MRI with MR angiography, and compare with conventional angiography at the last visit.

## Case report

A 3-year-old boy presented initially in 2006, with alternating and resolving hemiplegias, seizures and headaches. MR images (Figs 1a - c) demonstrated numerous collateral vessels around the brainstem, especially within the ambient and quadrigeminal plate cisterns. Also demonstrated was asymmetry and narrowing of the supra- and infraclinoid portions of the internal carotid arteries (ICAs) with significant stenosis of the supraclinoid portion (Figs 1d and 1e). A diagnosis of moyamoya disease was made after clinical and laboratory exclusion of secondary causes and with the use of imaging.

Over the next 6 years (2006 to 2012), the patient was followed up regularly as an outpatient, utilising serial routine MRI studies to monitor progression of the disease. Documented deterioration of cognitive function as well as significant learning difficulties was maintained.

Progression of the disease from 2006 - 2012 was demonstrated over serial MR examinations, which clearly demonstrated progressive collateral vessel formation around the brainstem as well as progressive narrowing of the ICAs bilaterally. No dilatation of the anterior or middle cerebral arteries was ever demonstrated; in fact, these vessels appeared smaller than on initial presentation.

In 2012, owing to worsening cognitive function and learning difficulties, repeat MR imaging (Fig. 2) was performed as well as conventional angiography (Fig. 3) to plan for possible surgical intervention. The characteristic 'puff/spiral of smoke' appearance of the collateral vessels at the level of the Circle of Willis was noted on conventional angiography. Marked narrowing of the infraclinoid portion of the ICAs bilaterally as well as complete stenosis of the supraclinoid portions of the ICAs bilaterally was also noted.

Currently, the patient is stable and awaiting neurosurgical intervention in the form of a pial synangiosis.

## Discussion

Moyamoya syndrome is a cerebral arteriopathy characterised by bilateral progressive stenosis of the terminal portions of the ICAs.<sup>[1]</sup> Collateral vessels (most commonly the lenticulostriate and thalamoperforator arteries) hypertrophy to compensate for the stenosis. The angiographic appearance of these collaterals gave rise to the term moyamoya and refers to 'hazy like a puff of smoke drifting in the air'.<sup>[2]</sup> Many disorders are associated with the moyamoya syndrome, and almost any slowly progressive intracranial vascular occlusion results in a

similar appearance. For this reason, moyamoya syndrome is seen by many to be a radiographic syndrome rather than a specific disease. The term moyamoya disease or primary moyamoya, however, is reserved for cases where no underlying or secondary cause can be identified, as in our patient.<sup>[3]</sup> An inherited form has been described in Japanese patients, where 10% of cases are familial.<sup>[4]</sup>

Disorders associated with secondary moyamoya include Down syndrome, neurofibromatosis type 1, tuberous sclerosis and sickle cell disease. An underlying cause is more likely when the occlusive changes are unilateral, and can usually be identified.<sup>[3]</sup> Causes of moyamoya must be actively and rigorously investigated for. Most patients present during childhood with varying neurological signs and symptoms, as in our patient. Approximately 70% of reported cases occur in patients <20 years of age, and 50% occur in children <10 years. Children usually have recurrent transient ischaemic attacks (TIAs) with progressive neurological impairment, while adult patients more often present with intracranial haemorrhage.<sup>[4]</sup> Our patient was noted to have alternating resolving hemiplegias (TIAs), seizures and headaches.

CT and MR are useful for detecting regions of cerebral infarction or ischaemia owing to the vasculopathy. MR is more sensitive than CT for detecting the enlarged moyamoya collaterals, which are seen as signal voids in the basal ganglia. MRA also demonstrates narrowing of affected intracranial vessels, as in our patient (Figs 1 and 2).

If intravenous contrast is administered, intense enhancement of the basal nuclei can occur – either owing to infarction and breakdown of the blood-brain barrier or from visualisation of the collateral vessels coursing through them. Curvilinear enhancement may be seen coursing radially through the centrum semiovale from the lateral ventricles to the cortex, probably representing dilated deep medullary veins. Branching, pial enhancement may also be seen, probably representing dilated capillaries. Perfusion imaging (SPECT or MR) is useful for identifying regions of relative ischaemia at risk for infarction.<sup>[5]</sup>

Conventional angiographic evaluation is nearly always included to define the extent of the disease and adequacy of collateral flow. The typical angiographic appearance is narrowing of the supraclinoid ICA, proximal anterior cerebral artery and proximal middle cerebral artery. The posterior circulation is rarely involved until late in the course of the disease. Lenticulostriate and thalamoperforator arterial hypertrophy is well appreciated on angiography. Evaluation of associated vascular abnormalities, such as saccular aneurysms, dissecting aneurysm and arteriovenous malformations is necessary at angiography.<sup>[6]</sup> None of these complications were noted in our case (Fig. 4). If left untreated, intellectual function and motor deficits worsen over time.

Indirect bypassing procedures are preferred for good outcome and relative simplicity v. direct bypassing procedures. Pial synangiosis (placing a branch of the external carotid artery in apposition to the pia-arachnoid) is an example of an indirect bypassing procedure that can be used.<sup>[7]</sup> The pial synangiosis procedure is planned for our patient to try to improve blood flow to the cerebral parenchyma.

## Conclusion

Moyamoya is a rare cerebral arteriopathy, especially in its idiopathic form. Secondary causes, with a moyamoya-like appearance, are more common. CT, MRI and conventional angiography can all be used to image the disease and its progression, and to assist planning for the various bypass treatment procedures that are possible.

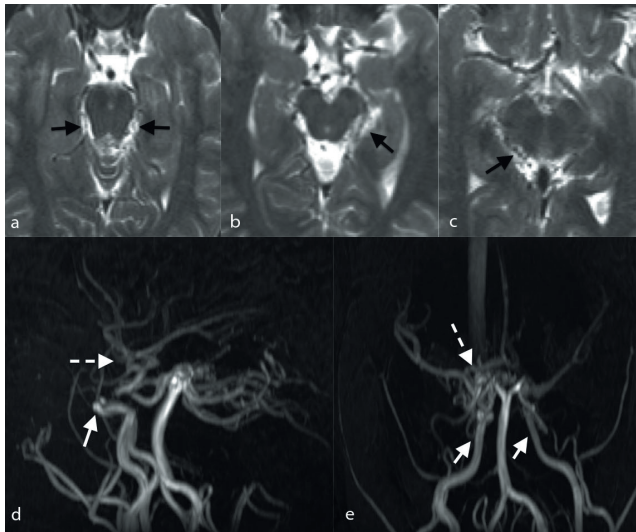


Fig. 1. MRI and MRA imaging diagnosis of moyamoya at the initial presentation of a 3-year-old boy: T2W axial images (a - c) of the brain, at the level of the Circle of Willis, show numerous collateral vessels, seen as multiple flow voids (black arrows), around the brainstem, especially within the ambient and quadrigeminal plate cisterns. MRA images with sagittal (d) and coronal (e) views show asymmetrical narrowing of the supra- and infraclinoid internal carotid arteries (white arrows) as well as the collateral vessel formation (dashed white arrows).

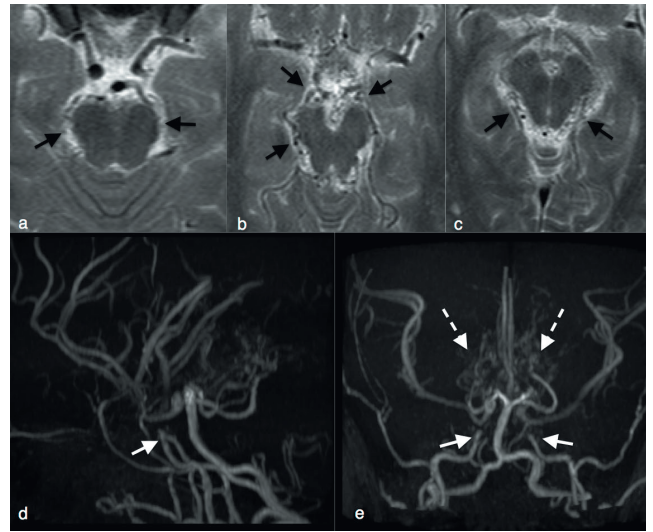


Fig. 2. Follow-up MRI and MRA imaging of the patient in Fig. 1 after a 6-year period: T2W axial images (a - c) show the markedly increased number of collateral vessels around the brainstem at the level of the Circle of Willis (black arrows). MRA on sagittal (d) and coronal (e) views shows increased stenosis of the supra- and infraclinoid portion of both internal carotid arteries (white arrows) and more numerous collateral vessels (dashed white arrows) compared with Fig. 1.

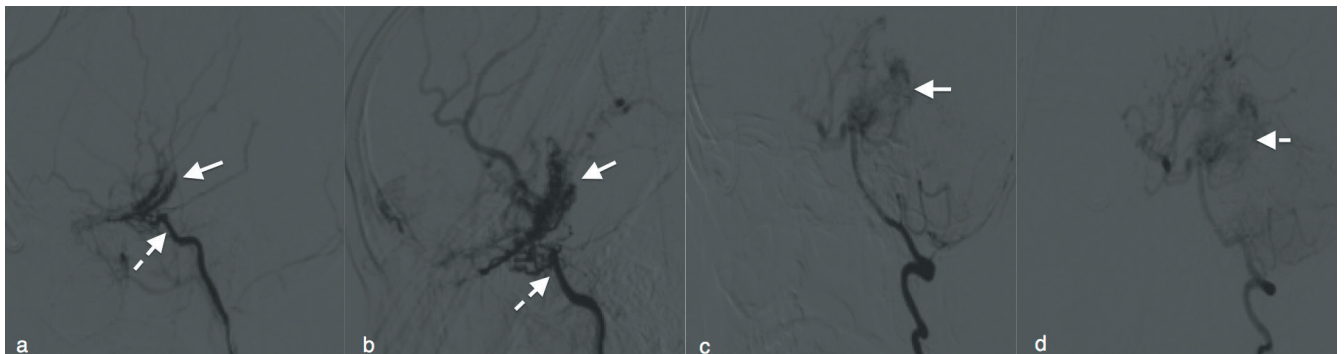


Fig. 3. Conventional angiogram images with lateral views of the cerebral arteries. Note the characteristic 'puff/spiral of smoke' appearance of the collateral vessels at the level of the Circle of Willis (white arrows). Also noted is the marked narrowing and stenosis of the supraclinoid portion of bilateral internal carotid arteries (dashed white arrows). a = left internal carotid artery; b = right internal carotid artery; c = left vertebral artery; d = right vertebral artery.

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