Paediatric Moyamoya Vasculopathy with Anterior and Posterior Circulation Involvement: A Case Report

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Abstract

A 14-year-old boy presented with recurrent episodes of transient left-sided limb weakness over eight months. MRI showed predominantly right parieto-occipital atrophy and signal changes compatible with previous ischaemic injury, without restricted diffusion. Time-of-flight magnetic resonance angiography demonstrated severe steno-occlusive changes involving the anterior and posterior circulations, with basal collateral vessels consistent with moyamoya vasculopathy. ANA testing showed nucleolar 2+ reactivity on immunofluorescence with a negative immunoblot; these findings alone neither establish nor exclude an associated autoimmune disorder. The parents opted for conservative management. The child remained clinically stable during five days of inpatient observation and was discharged with advice for regular follow-up. The case illustrates the contribution of MRI and angiography to evaluating recurrent transient neurological symptoms in children and the need to interpret vascular and laboratory findings in their clinical context.

Keywords: Moyamoya vasculopathy; Paediatric; Transient ischaemic attack; Magnetic resonance angiography

Introduction

Moyamoya is a progressive cerebrovascular disorder characterized by stenosis or occlusion of the terminal internal carotid arteries and their proximal branches, with compensatory collateral vessels that produce the characteristic puff-of-smoke appearance on angiography [1,3,10]. Children may present with transient ischaemic attacks, seizures, headaches or cognitive difficulties. Moyamoya syndrome describes a similar vascular pattern associated with other conditions. Assessment of both the vascular findings and potential associated disorders is therefore important [10].

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Case Report

A 14-year-old boy presented to the Emergency Department at North Bengal Medical College and Hospital with recurrent episodes of transient weakness of the left upper and lower limbs over eight months. Each episode began suddenly and resolved within one to two hours. During episodes, he had difficulty standing or walking, fell and required assistance to get up. There was no associated loss of consciousness, seizure activity, vomiting, tongue biting, or bladder or bowel incontinence. No visual disturbances, dizziness, headaches or convulsions were reported.

He was the first child of non-consanguineous parents and had been born by normal vaginal delivery in a hospital. Blood pressure was 100/60 mmHg and pulse rate 70 beats/min; anthropometric parameters were appropriate for age. Muscle tone was normal and power was 5/5 in all limbs. Knee and ankle reflexes were brisk bilaterally. The plantar response was flexor on the right and absent on the left. No significant sensory deficit or systemic abnormality was detected. The clinical account also noted cognitive decline and poor scholastic performance.

MRI showed predominantly right parieto-occipital atrophy in the MCA and PCA territories. No restricted diffusion was present. Gyriform high signal on FLAIR images along the posterior parietal and occipital convexity was interpreted as sequelae of previous infarction (Figure 1).

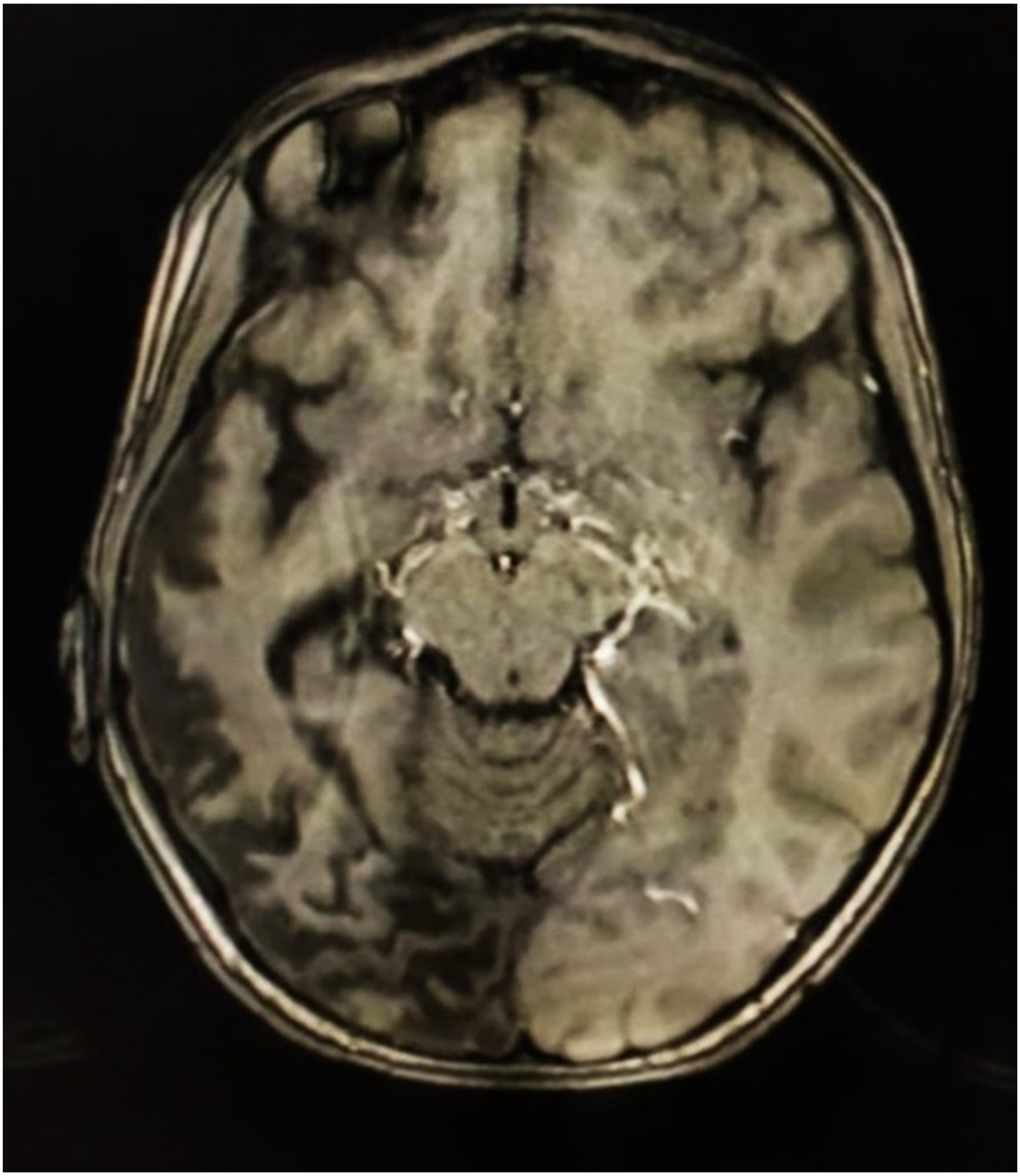


Figure 1. MRI and TOF-MRA showing predominantly right parieto-occipital atrophy in the MCA and PCA territories, with multiple small collateral vessels and reduced visualization of the major intracranial arteries. The side- and segment-specific findings are described in the text.

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Angiographic Findings

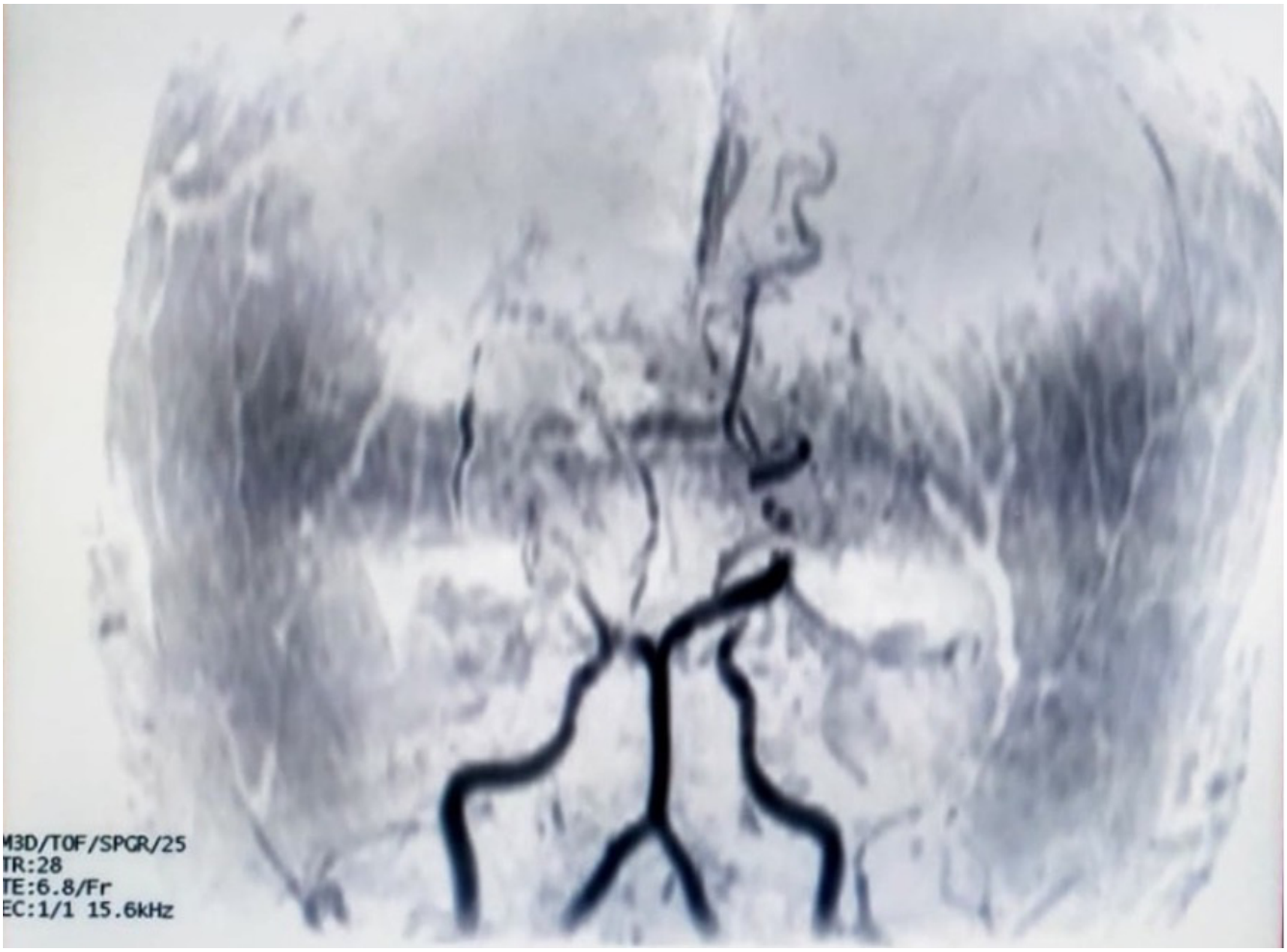


Figure 2. Coronal maximum-intensity-projection TOF-MRA showing severe steno-occlusive changes with multiple serpiginous basal collateral vessels, producing the characteristic puff-of-smoke appearance. Visualization differs by side and arterial segment as described in the text.

TOF-MRA demonstrated severe steno-occlusive changes with extensive basal collateral vessels. On the left, the supraclinoid ICA, ACA and MCA were not visualized. On the right, a small terminal ICA segment and a short M1 segment were visualized; the report described both ACAs as arising from the right terminal ICA segment. The left PCA was visible to P4 but had reduced calibre. The right PCA was not visualized beyond P1, and the right posterior communicating artery was not visualized. Non-visualization on TOF-MRA does not by itself establish anatomical absence.

The basal collateral network had the characteristic puff-of-smoke appearance. A working diagnosis of moyamoya vasculopathy was made [10].

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Laboratory Findings and Clinical Course

On the third day of admission, ANA testing showed nucleolar 2+ reactivity on immunofluorescence, while the ANA immunoblot was negative. These laboratory findings were considered alongside the clinical assessment.

Treatment with aspirin and statins was reported. The child was monitored for further neurological symptoms and remained clinically stable during five days of inpatient observation. After discussion of management options, the parents opted for conservative treatment. Regular follow-up was advised at discharge.

The sensory and motor systems were monitored during hospitalization, without further deterioration. At discharge, the general and systemic examinations were normal, with no significant change in motor function. The family was counselled to avoid situations provoking hyperventilation, crying or straining, because associated reductions in carbon dioxide may precipitate transient ischaemic symptoms [7,8,9].

Discussion

This child presented with recurrent transient left-sided weakness and imaging evidence of chronic predominantly right-sided ischaemic injury. The reported TOF-MRA findings involved both anterior and posterior circulations. Residual visualization varied by side and arterial segment, alongside an extensive collateral network. Moyamoya is associated with genetic susceptibility, including RNF213, and may also occur with associated systemic conditions [5,10].

A positive ANA immunofluorescence result does not by itself establish an autoimmune disease, and a negative immunoblot only addresses the antigen specificities included in that assay [11]. The combination reported in this case cannot be used to exclude all autoimmune disorders. Imaging was consistent with moyamoya vasculopathy; distinguishing idiopathic moyamoya disease from moyamoya syndrome requires assessment for associated conditions [10].

Antiplatelet treatment may be considered for ischaemic moyamoya disease, while revascularization is an important treatment option for symptomatic patients [7]. The short inpatient course in this case does not establish the long-term effectiveness of medical treatment or a specific benefit from statins.

Conclusion

Moyamoya vasculopathy should be considered in children with recurrent transient focal neurological symptoms. MRI and angiographic imaging help characterize ischaemic injury and the distribution of vascular changes. Assessment for associated conditions and individualized management are required; a negative ANA immunoblot alone does not establish idiopathic moyamoya disease [7,10,11].

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Declarations

Consent for publication: The authors state that written informed consent was obtained and patient anonymity was ensured. They report that the study was conducted in accordance with the Declaration of Helsinki and applicable local and international standards.

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