Diagnosis and Management of Complex Cases

MOYAMOYA DISEASE: Lessons from a Fatal Intracerebral Haemorrhage in a Young Patient Who Did Not Undergo Revascularisation

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ABSTRACT

This report describes a fatal case of moyamoya disease to illustrate the consequences of not undertaking surgical revascularisation.

A 13-year-old boy with no previous specialist follow-up was admitted with status epilepticus (Glasgow Coma Scale score 7). His history revealed progressive moyamoya disease diagnosed six years earlier, complicated by two ischaemic strokes, and managed solely with aspirin. Emergency non-contrast brain computed tomography demonstrated a massive intracerebral haemorrhage with intraventricular extension. Despite intensive care management including endotracheal intubation and external ventricular drainage, the clinical course was rapidly fatal. This tragic outcome highlights the catastrophic natural history of moyamoya disease in the absence of surgical revascularisation. It underscores the critical need for early detection, systematic referral to expert centres, and structured multidisciplinary management to prevent lethal complications. Surgical revascularisation remains the cornerstone disease-modifying therapy.

Keywords: *Moyamoya disease; Intracerebral haemorrhage; Child; Surgical revascularisation; Death; Case report.*

INTRODUCTION

Moyamoya disease is a rare, progressive cerebrovascular disorder characterised by chronic occlusive stenosis of the internal carotid arteries and their proximal branches, associated with the development of a collateral vascular network with a characteristic "puff-of-smoke" appearance ("moyamoya") [1].

Its natural course is punctuated by ischaemic and/or haemorrhagic cerebrovascular events that may be fatal, with an annual stroke risk of up to 10% under medical management alone [2].

Surgical revascularisation, via direct bypass and/or indirect synangiosis procedures, is the only treatment shown to stabilise the disease and significantly reduce recurrence [3,4].

We report a tragic case illustrating the consequences of the absence of specialised neurosurgical management, emphasising the imperative need for early referral to expert centres.

CASE REPORT

A 13-year-old boy with no previous follow-up in our department was admitted to the emergency department with status epilepticus and a Glasgow Coma Scale score of 7. History obtained from the family revealed a diagnosis of progressive moyamoya disease established six years earlier and subsequently complicated by two ischaemic strokes. The patient had been treated only with antiplatelet-dose aspirin and had not undergone specialist neurosurgical follow-up or assessment for revascularisation.

An urgent non-contrast brain CT scan demonstrated a massive intracerebral haemorrhage with rupture into the ventricular system (intraventricular haemorrhage) and upstream ventricular enlargement (Figure 1). Despite intensive resuscitation including orotracheal intubation and placement of an external ventricular drain, the clinical course rapidly deteriorated, and the patient died within 48 hours.

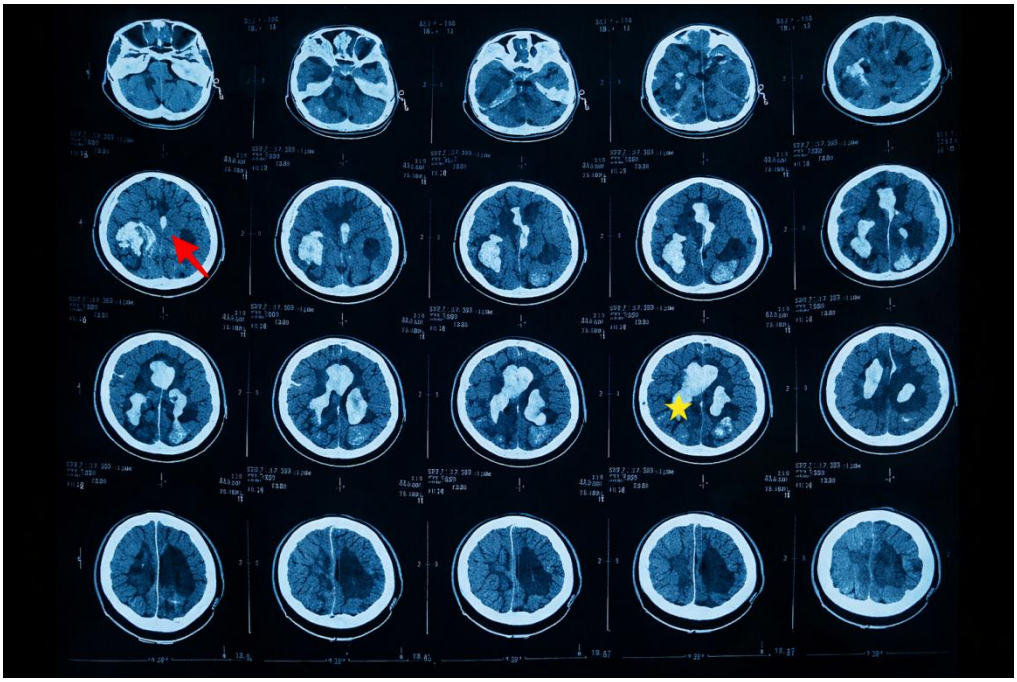

Figure 1: *Non-contrast axial brain CT showing a massive intracerebral haemorrhage (asterisk) with intraventricular extension and ventricular dilatation (arrows).*

DISCUSSION

This fatal case highlights several key issues in the management of moyamoya disease.

Natural history and prognosis without revascularisation

Our observation illustrates the poor prognosis of forms not treated surgically. Under medical management alone (antiplatelet therapy), the annual risk of cerebrovascular events (ischaemic or haemorrhagic stroke) remains high, with substantial cumulative morbidity over time [2,5].

Central role of surgical revascularisation

Revascularisation techniques (direct superficial temporal artery–middle cerebral artery bypass and/or indirect synangiosis procedures) aim to restore stable cerebral perfusion. Contemporary evidence indicates that these procedures reduce stroke risk by more than 70%, markedly improving prognosis [4,6]. In symptomatic disease, intervention within a few months of diagnosis is generally recommended [3,7].

Failures along the care pathway

This case underscores the consequences of a lack of referral to an expert centre with neurovascular expertise. Persistence with medical therapy alone in a symptomatic child with documented progressive disease represents an avoidable adverse prognostic factor [2,3].

Need for structured multidisciplinary management

Optimal care requires close clinical and radiological follow-up (annual MRI/vascular imaging) and multidisciplinary, consensus-based decision-making involving neurologists, neuroradiologists, and neurosurgeons within dedicated care pathways, as emphasised in recent recommendations [3,7].

Recent Recommendations (2025)

An expert consensus published in 2025 provides updated recommendations for the diagnosis and management of moyamoya disease, emphasising: (i) diagnostic confirmation using appropriate vascular imaging and, where available, haemodynamic assessment tools; (ii) an individualised therapeutic strategy; and (iii) management in specialised centres with standardised peri-operative measures [9].

Recent publications also highlight important developments in paediatric management, including optimisation of imaging, personalisation of revascularisation strategies, and improvements in peri-operative protocols, further supporting the value of expert-centre care pathways [10].

Revascularisation remains the reference treatment in symptomatic patients with evidence of haemodynamic insufficiency, consistent with European Stroke Organisation (ESO) materials, which stress that these procedures should be undertaken in reference centres by experienced teams [11].

Finally, recent evidence underscores the importance of anticipating complications, particularly cerebral hyperperfusion syndrome following revascularisation. Its incidence was reappraised in a systematic review and meta-analysis (published in 2026 with a 2025 DOI), supporting structured post-operative monitoring [8].

Key Messages

- Moyamoya should be suspected in children with recurrent TIAs/strokes and promptly confirmed using appropriate vascular imaging.
- Early referral to an expert centre (neurology, neurosurgery, neuroradiology) is essential for haemodynamic assessment and multidisciplinary decision-making.

- Revascularisation is central when indicated; medical therapy is adjunctive, and peri-operative surveillance is crucial.

CONCLUSION

This potentially preventable death underscores the imperative need for specialised and proactive management of moyamoya disease. It supports early detection and timely referral to expert centres, recognition of surgical revascularisation as the only treatment that modifies vital prognosis, and the organisation of national care pathways for this rare disorder. Such pathways should ensure structured, multidisciplinary follow-up in order to prevent outcomes as tragic as that reported here.

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