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Case Report

# A Kiss of Death from a Puff of Smoke: Moyamoya due to Sickle Cell Neuro-vasculopathy

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## Abstract

Moyamoya syndrome (MMS) is a neuro-vascular complication of sickle cell disease which is characterized by progressive narrowing of the carotid arteries and the subsequent development of collateral circulation. MMS is relatively uncommon but carries significant risk of morbidity and mortality as it is associated with an increased risk of cerebrovascular events [1]. The management of MMS is difficult, particularly in patients with sickle cell disease (SCD). Further research is needed to evaluate the efficacy of available treatment modalities in improving outcomes for patients with SCD. We report the first documented paediatric case of Moyamoya syndrome in Barbados in a patient with a history of homozygous sickle cell disease (HbSS) who developed recurrent haemorrhagic cerebrovascular events which resulted in the patient's demise.

***Keywords:*** *Moyamoya; Sickle Cell Disease; Cerebrovascular Events*

## Introduction

Moyamoya syndrome (MMS) is a neuro-vascular complication of sickle cell disease which is characterized by progressive narrowing of the carotid arteries and the subsequent development of collateral circulation.

## Case Report

An 11-year old, Afro-Caribbean girl with a history of sickle cell disease (SCD) with recurrent cerebrovascular events presented to the emergency department with acute onset, severe right- sided headache and seizure activity from the morning of presentation. Within a few minutes of onset of the headache, the patient vomited and began having a generalized tonic-clonic seizure which lasted a few minutes. After the initial seizure, she was noted to be poorly responsive and continued to have intermittent seizures with only brief periods of remission over an hour and was brought to the emergency department via private transport.

There was no reported visual disturbance or limb weakness. There was no antecedent history of fever, cough or other coryzal symptoms, diarrhea, change in speech or behavior, trauma, falls or toxic ingestions.

On arrival at the emergency department, she was actively seizing, and her GCS was noted to be 3/15 with increased tone noted throughout. The seizures continued despite two (2) doses of IV benzodiazepines and a phenytoin loading. The decision was then made to intubate the patient as per APLS status epilepticus protocol. The seizures aborted with rapid sequence induction agents. She was ventilated and admitted to the paediatric intensive care unit (PICU) for further management.

## Past medical history

The patient was one of a dichorionic, diamniotic twin gestation, born via caesarean section at 33 weeks with Apgar scores $9^1$, $9^5$. She had a brief stay in the neonatal ICU for five days. She was diagnosed with sickle cell disease at age 6 months and her non-identical twin sister was Hb AA.

Her past medical history was significant for a transient ischaemic attack at age 6 years, left internal carotid artery stroke at age 7 years and haemorrhagic stroke at age 10 years. She was diagnosed with Moyamoya based on neuroimaging done when she was 9 years (Figure 1). She subsequently commenced hydroxyurea and exchange transfusions were initiated but the patient subsequently defaulted from the transfusion programme.

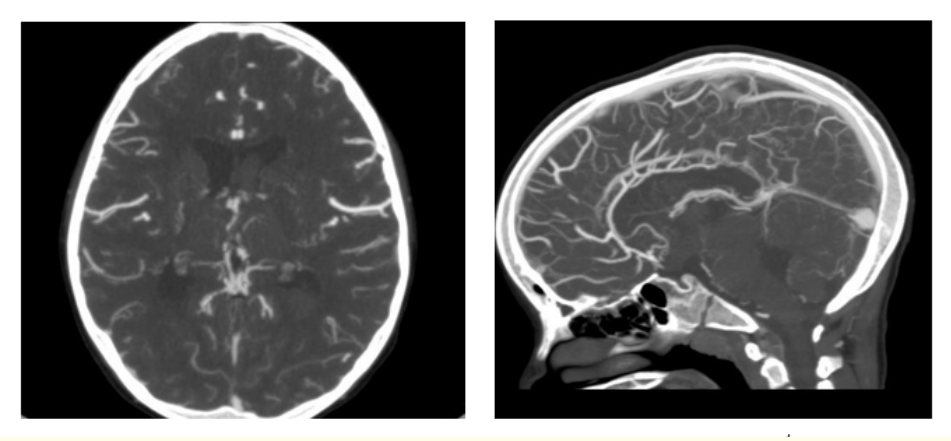

***Figure 1:*** *CT brain with contrast demonstrating Moyamoya syndrome.*

During this current admission, a CT brain with contrast revealed an acute haemorrhage in all the ventricles, more so in the lateral ventricles. There was extension into the periventricular white matter adjacent to the posterior horn of the right lateral ventricle. There was mild dilatation of both lateral ventricles with a mild shift of the midline to the left (Figure 2).

A neurosurgical consultation was requested, and it was deemed that the intracranial haemorrhage was too extensive and the risks of surgery outweighed the benefits for this patient. No neurosurgical intervention was offered as it was deemed her prognosis was extremely guarded and it was unlikely to change the outcome for this patient.

Within 12 hours of admission, the patient developed fluid refractory hypotension and commenced inotrope support. Her pupils were noted to be fixed and dilated at 7mm bilaterally. Given the acute neurological change, cerebral oedema versus herniation was suspected. Repeat neuroimaging showed marginal extension of the intracranial haemorrhage with trans-tentorial herniation noted. By day 2 of admission, she developed central diabetes insipidus and progressive renal impairment with multiorgan failure and subsequently demised.

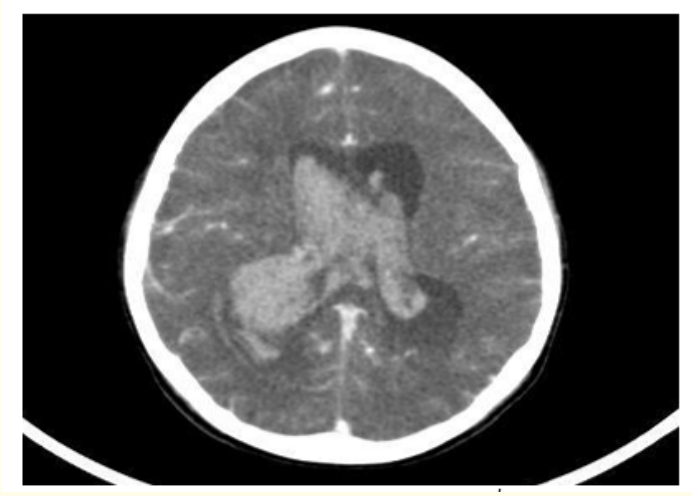

***Figure 2:*** *Intracranial haemorrhage.*

## Discussion

This 11-year-old child had severe, progressive neuro-vasculopathy as a complication of sickle cell disease. This led to multiple ischaemic cerebrovascular events in the past and subsequent development of moyamoya syndrome, which further predisposed the patient to recurrent intracranial haemorrhages.

Moyamoya disease refers to the idiopathic form whereas moyamoya syndrome (MMS) refers to the development of collateral cerebral circulation in children with an underlying clinical condition. MMD is a rare, progressive vascular disease that occurs as a result of progressive narrowing of cerebral vasculature leading to transient ischaemic attacks or strokes. In response to cerebral ischemia, there is the development of collateral vessels in the region of the basal ganglia, which give rise to the characteristic angiographic appearance of "puff of smoke", moyamoya [2].

Pediatric MMS usually presents with cerebral ischemia in up to 80% of cases, rather than hemorrhage which is more common in adults [3]. Patients present with seizures, headaches, transient ischaemic events and cognitive decline.

Conventional cerebral angiography is the gold standard for diagnosis of MMD. Magnetic resonance angiography is also useful and has a specificity of up to 100% for moyamoya. Currently, the only available medical treatment proven to be beneficial to stabilize or reverse MMS has been reported in patients with sickle cell disease who receive chronic exchange transfusions [4]. Antiplatelet agents and calcium channel blockers are used as adjuncts. Revascularization procedures can be undertaken to prevent further ischaemic injury, but these procedures should be undertaken early prior to disease progression [5,6]. Stroke recurrence risk decreases after patients have been treated with surgical revascularization procedures [7,8].

It is important to screen all patients with sickle cell disease with transcranial doppler ultrasonography as they are at increased risk of ischaemic neurovascular complications. MMS should be considered in any patient with SCD who presents with recurrent cerebrovascular events, as untreated moyamoya may progress slowly with intermittent transient events or have rapid progression with devastating permanent cognitive impairments or haemorrhagic events associated with significant morbidity and mortality, as occurred with this patient.

## Conclusion

Moyamoya syndrome is a relatively infrequent complication of sickle cell disease but should be considered in any patient who presents with recurrent cerebrovascular events. Conventional cerebral angiography remains the gold standard for diagnosis of Moyamoya syndrome. Management of MMS is difficult and further research is required to evaluate neurologic outcomes of patients who have had either medical or surgical interventions. Without treatment, the risk of intracranial haemorrhage increases significantly which could result in poor outcomes, as occurred in this case.

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