

Intracranial Aneurysm in Sickle Cell Disease: Case Report of a Fatal Outcome

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Case Report

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Abstract

Background: Intracranial aneurysms are an under-recognized but increasingly reported cerebrovascular complication of sickle cell disease (SCD). Ruptured aneurysms in SCD patients often occur at a young age and are associated with poor outcomes, particularly when definitive treatment is delayed or unavailable.

Case Presentation: We report a 21-year-old African male with homozygous sickle cell disease (HbSS) who presented to a tertiary hospital in Ghana with a 16-hour history of sudden thunderclap headache and progressive reduction in consciousness. On admission, he had a GCS of 9/15 with nuchal rigidity. Non-contrast CT scan revealed acute subarachnoid hemorrhage with intraventricular extension and mild hydrocephalus. Cerebral angiography confirmed a ruptured anterior communicating artery aneurysm.

Urgent endovascular coiling was recommended following neurosurgical and hematology review. However, due to limited resources and financial constraint, the patient could not undergo the procedure. He was managed conservatively with analgesia, nimodipine, seizure prophylaxis, and supportive intensive care. On day 5 of admission, his neurological status deteriorated (GCS 6/15), requiring intubation and mechanical ventilation. He succumbed on day 7 of admission despite maximal supportive therapy.

Conclusion: This case highlights the devastating consequence of ruptured intracranial aneurysm in a young patient with SCD when timely endovascular intervention is not undertaken. It underscores the urgent need for improved access to neuro-interventional services, strengthened health financing mechanisms, and context-specific guidelines for managing cerebrovascular complications in SCD within resource-limited settings.

INTRODUCTION

Sickle cell disease (SCD) is one of the most common inherited hemoglobinopathies worldwide, with a particularly high prevalence among individuals of African descent.⁹ While vaso-occlusive crises and ischemic strokes are well-documented neurological complications, hemorrhagic cerebrovascular events such as aneurysmal subarachnoid hemorrhage (aSAH) are less common but associated with significant morbidity and mortality.^{1,8}

Intracranial aneurysms occur at a markedly higher frequency in patients with SCD than in the general population.¹⁰ Recent studies report a prevalence ranging from 4.1% to as high as 17% in screened SCD cohorts, compared to approximately 3% in the general adult population.^{1,2} These aneurysms frequently present at a younger age, are often multiple, and may rupture at smaller sizes.³

The pathogenesis of the aneurysm is believed to involve chronic endothelial damage from recurrent sickling, altered blood rheology, and increased hemodynamic stress secondary to chronic anemia and

elevated cerebral blood flow.^{1,4} Unlike Moyamoya vasculopathy, which is also seen in SCD, intracranial aneurysm formation appears to be particularly linked to the homozygous HbSS genotype.⁴

Despite growing awareness, ruptured intracranial aneurysms in young SCD patients remain particularly challenging to manage, especially in low-resource settings where endovascular coiling or surgical clipping may not be feasible due to financial or infrastructural limitations.

CASE PRESENTATION

A 21-year-old African male with known SCD (genotype HbSS) presented to the Emergency Department of Komfo Anokye Teaching Hospital on 21st April 2026 with a 16-hour history of sudden-onset thunderclap headache associated with reduced level of consciousness. His past neurological history was unremarkable.

On initial assessment, he had a patent airway that was maintainable, was spontaneously breathing with adequate air entry bilaterally. Vital signs showed a blood pressure of 118/61 mmHg, pulse rate of 66 bpm, respiratory rate of 16 cpm, and oxygen saturation of 97% on room air. He was unconscious and reacted to some amount of pressure with a Glasgow Coma Scale (GCS) score of 9/15 (E2M5V2). His pupils were equal in size, round and reactive to light. He was pale and anicteric. Supplemental oxygen was given via intra-nasal canula.

Computed tomogram (CT) of brain showed acute subarachnoid hemorrhage (SAH) with intraventricular extension. Further investigation with angiography showed a ruptured Anterior Communicating Artery (aCoA) aneurysm complicated by acute SAH with intraventricular extension and mild communicating hydrocephalus.

A decision was made in consultation with neurosurgery and hematology specialists to proceed with urgent endovascular coiling of the aneurysm, but this was delayed due to limitation of access and funding. While waiting for definitive care, the patient was started on supportive care with intravenous paracetamol and morphine for analgesia, intravenous phenytoin for seizure prophylaxis, oral nimodipine for vasospasm prevention, syrup lactulose for constipation prevention, and head-of-bed elevation to 30 degrees. He was admitted to the intensive care unit.

On day 5 of admission, the patient experienced further neurological deterioration with a decline in GCS to 6/15 (E1M4V1), necessitating endotracheal intubation and mechanical ventilation. Despite supportive intervention, the patient died on day 7 of admission.

INVESTIGATIONS

Representative neuroimaging is presented in Figs. 1 and 2. Laboratory findings showed a hemoglobin level of 8.2 g/dL and a platelet count of $595 \times 10^3/\mu\text{L}$. Renal and liver function tests were unremarkable.

DISCUSSION

Intracranial aneurysms are increasingly recognized as an important cerebrovascular complication in SCD, occurring at a significantly higher prevalence than in the general population. Recent prospective imaging studies report a prevalence of approximately 17% in SCD cohorts, compared to ~3% in the general adult population.^{1,2} These aneurysms frequently present at a younger age, are often multiple (up to 45–80%), tend to be small and saccular, and show a relatively higher involvement of the posterior circulation.³

The pathogenesis is multifactorial, primarily driven by chronic endothelial injury from recurrent vaso-occlusion, abnormal blood rheology, and increased hemodynamic stress due to chronic anemia and elevated cerebral blood flow.^{1,4} Higher cerebral blood flow has been independently associated with presence of aneurysm in SCD patients.¹

Outcomes of Surgical/Endovascular Intervention versus Conservative Management

Literature suggests that definitive aneurysm securement (endovascular coiling or surgical clipping) significantly improves survival and functional outcomes compared to conservative management in aneurysmal subarachnoid hemorrhage, including in SCD patients.⁵ In general aSAH cohorts, early treatment of aneurysm reduces the risk of re-bleeding and associated mortality.¹¹

In SCD-specific reports, patients who received endovascular coiling or clipping, combined with careful perioperative transfusion strategies and multidisciplinary care (hematology, neurology, and neurocritical care), have achieved favorable outcomes (modified Rankin Scale 0–2 at follow-up) in several documented cases.^{5,6} Endovascular therapy appears safe when preceded by exchange transfusion to reduce HbS levels and optimize oxygenation.

In contrast, conservative management — as applied in our case due to limited resources and financial constraint — is associated with high mortality. Without aneurysm securement, patients remain at risk of re-bleeding, worsening vasospasm, hydrocephalus, and raised intracranial pressure. Our patient's rapid neurological decline from GCS 9/15 to 6/15 and death on day 7 illustrates the poor prognosis of unsecured ruptured aneurysms, even with supportive measures such as nimodipine, analgesia, seizure prophylaxis, and mechanical ventilation.

This case emphasizes the importance of prompt neuroimaging in patients with sickle cell disease who presents with thunderclap headache and the need for increased awareness of aneurysmal subarachnoid hemorrhage as a differential diagnosis. It also highlights major challenges in resource-limited settings, including limited neuro-interventional infrastructure and the heavy economic burden faced by families of SCD patients in low and middle-income countries.

CONCLUSION

Ruptured intracranial aneurysm is a rare but devastating neurological complication in young patients with sickle cell disease. This report describes the fatal outcome of a 21-year-old male with HbSS genotype who could not access endovascular coiling due to financial constraints. While conservative management remains an option in resource-limited settings, it is associated with markedly higher mortality compared to timely interventional treatment.

Improving outcomes requires a multi-pronged approach: early diagnosis, multidisciplinary care with optimized transfusion strategies, development of affordable neurosurgical services, imaging modalities, and policies that reduce catastrophic health expenditure for families affected by SCD. Greater investment in preventive screening and health system strengthening in SCD-endemic regions is essential to prevent similar preventable deaths.

Declarations

Consent for publication

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images.

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Figures

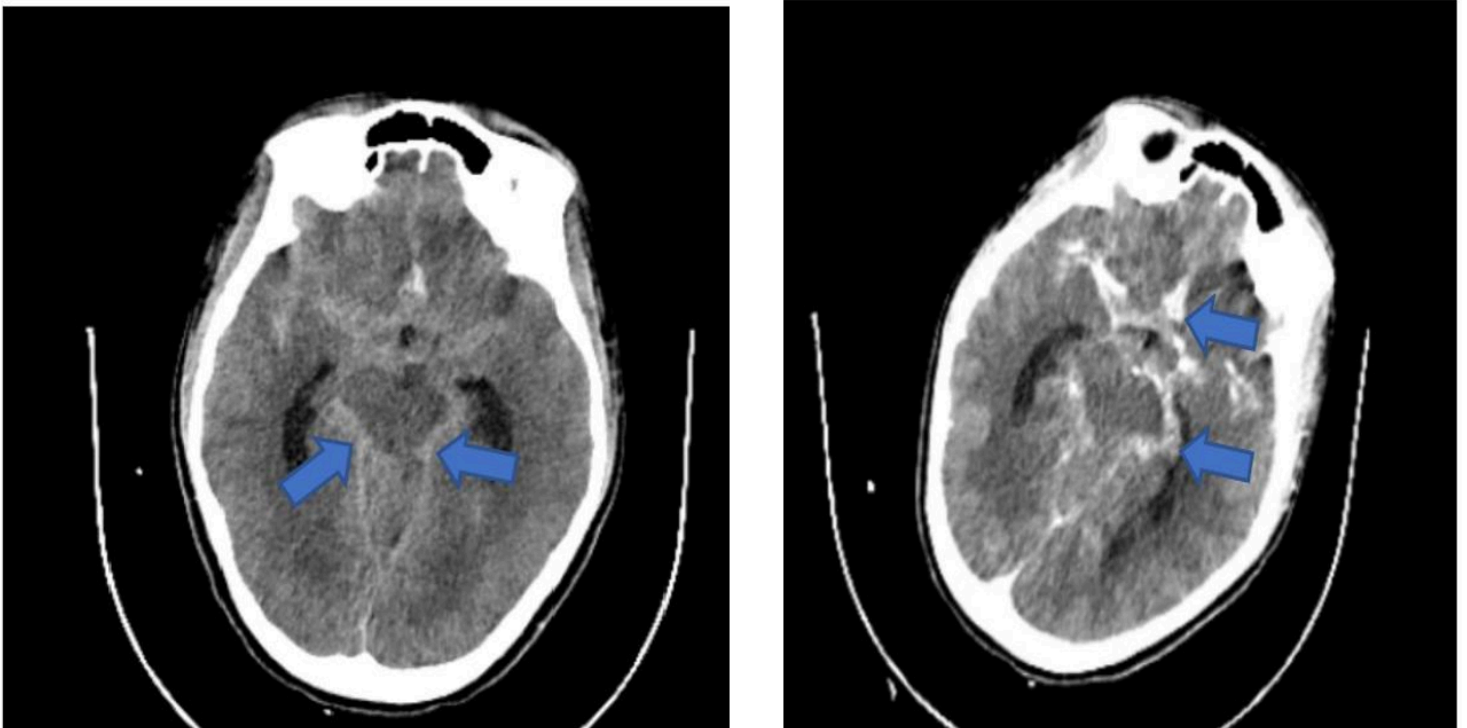


Figure 1

Axial CT of the head. (A) Non-contrast image demonstrating hyperattenuating blood in the basal cisterns consistent with acute subarachnoid haemorrhage. (B) Contrast-enhanced image showing subarachnoid haemorrhage and intraventricular extension.

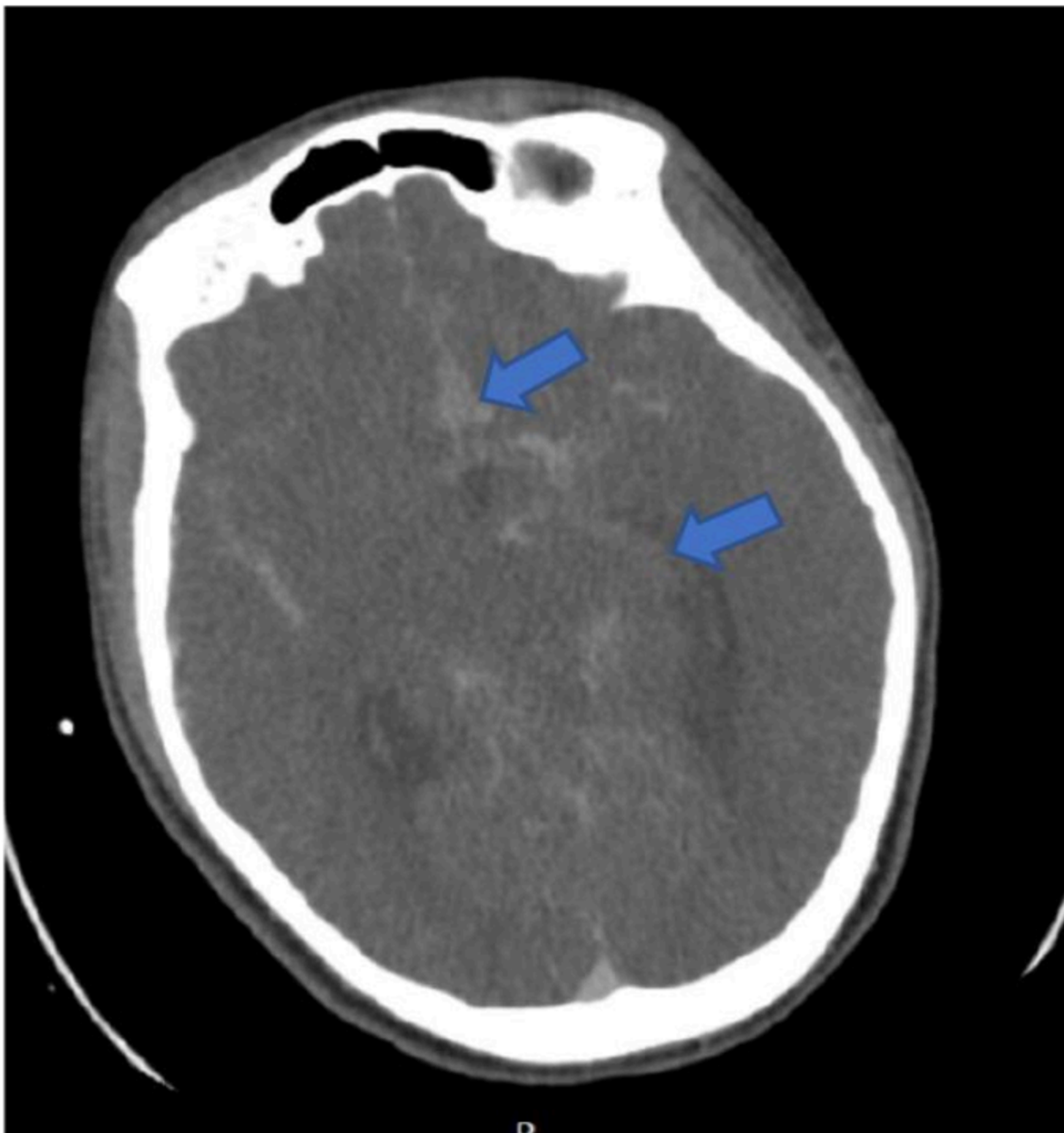


Figure 2

CT angiography demonstrating a ruptured anterior communicating artery (ACoA) aneurysm with associated subarachnoid and intraventricular haemorrhage.