

Ischemic Stroke in Children With Sick Cell Disease: A Report of Three Cases and a Review of the Literature

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Abstract

Sickle cell disease (SCD) is an inherited hemoglobin disorder associated with significant morbidity beginning early in childhood. Neurological complications are among the most severe manifestations of the disease. Stroke remains a major complication, leading to permanent neurological deficits, impaired quality of life, and unfavorable long-term outcomes in affected children.

We report three cases of ischemic stroke occurring in children aged 8-12 years with homozygous SCD (HbSS). All patients presented with acute focal neurological deficits, including hemiparesis and facial paralysis. Brain imaging confirmed cerebral ischemic lesions, while magnetic resonance imaging (MRI) revealed associated Moyamoya syndrome in one patient. Management included blood transfusions, hydroxyurea therapy, and motor rehabilitation. Despite treatment, one patient experienced recurrent strokes, and another died from acute chest syndrome during hospitalization.

These cases highlight the severity of cerebrovascular complications in children with SCD and underscore the importance of early diagnosis, systematic screening, and improved access to specialized diagnostic and therapeutic resources to optimize neurological outcomes in resource-limited countries.

Categories: Hematology, Pediatrics

Keywords: morocco, pediatric stroke, sickle cell disease (scd), transcranial doppler ultrasound, young child

Introduction

Sickle cell disease (SCD) is the most common inherited hemoglobinopathy worldwide and is particularly prevalent in sub-Saharan Africa and the Mediterranean region, including Morocco [1]. In Morocco, SCD appears to be more frequent in certain rural communities, where the high prevalence of consanguineous marriages may contribute to the persistence and transmission of the sickle cell allele. However, the true incidence and prevalence of SCD in Morocco remain difficult to estimate due to the absence of a nationwide systematic newborn screening program and limited epidemiological data, suggesting that the actual burden of the disease may be underestimated.

Despite advances in supportive care, SCD remains associated with substantial morbidity and mortality, particularly among children [2]. Neurological complications, especially ischemic stroke, are among the leading causes of morbidity and mortality in children with homozygous SCD (HbSS) [3]. Without appropriate preventive strategies, including transcranial Doppler (TCD) screening and chronic transfusion therapy, the cumulative risk of stroke in children with SCD may reach 11% by 20 years of age [4].

The underlying pathophysiology involves progressive cerebral vasculopathy driven by chronic hemolysis, vaso-occlusion, endothelial dysfunction, and persistent vascular inflammation [5,6]. Early identification of children at high risk through routine TCD screening, followed by chronic transfusion therapy, has substantially reduced stroke incidence in high-resource settings. However, implementing these preventive strategies remains challenging in low-resource settings, where access to specialized care is often limited [4,5].

We report three cases of ischemic stroke in children with homozygous SCD to illustrate the clinical presentation, neuroimaging findings, management, and outcomes of this life-threatening complication in a resource-limited setting.

Case Presentation

Case 1

An eight-year-old boy born to first-degree consanguineous parents, both carriers of sickle cell trait (HbAS),

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had been followed in our department for homozygous SCD (HbSS) since the age of one year. The diagnosis was established following an episode of dactylitis (hand-foot syndrome), which represented the first clinical manifestation of the disease, and was subsequently confirmed by hemoglobin electrophoresis. The electrophoretic profile revealed a predominance of sickle hemoglobin (HbS) (78.7%), with fetal hemoglobin (HbF) at 18.3% and hemoglobin A2 (HbA2) at 3.0%, confirming the diagnosis of HbSS disease. Initial management consisted of hydroxyurea therapy with regular monitoring of HbS levels.

The patient was admitted for sudden-onset neurological symptoms characterized by weakness of the right upper limb. The motor deficit was associated with cognitive impairment, including difficulties with memory and concentration, as well as speech arrest. There was no history of head trauma, infection, or recent vaso-occlusive crisis preceding the neurological event.

Neurological examination revealed right upper limb monoparesis with preserved deep tendon reflexes, right central facial palsy, and dysarthria. No sensory impairment, sphincter dysfunction, or sensory level suggestive of spinal involvement was observed. These findings were consistent with a focal central neurological deficit suggestive of an acute cerebrovascular event.

Brain computed tomography (CT) demonstrated an acute ischemic infarction involving the left precentral (pre-Rolandic) frontal region, associated with a lacunar infarction affecting the head of the left caudate nucleus and the genu of the ipsilateral internal capsule. Brain magnetic resonance imaging (MRI) confirmed the presence of multiple left supratentorial ischemic infarcts of different ages, including a subacute infarction within the left middle cerebral artery (MCA) territory. Magnetic resonance angiography (MRA) revealed cerebral vasculopathy characterized by collateral vascular networks compatible with Moyamoya syndrome, associated with hypoplasia of the left internal carotid artery (Figure [1](#)).

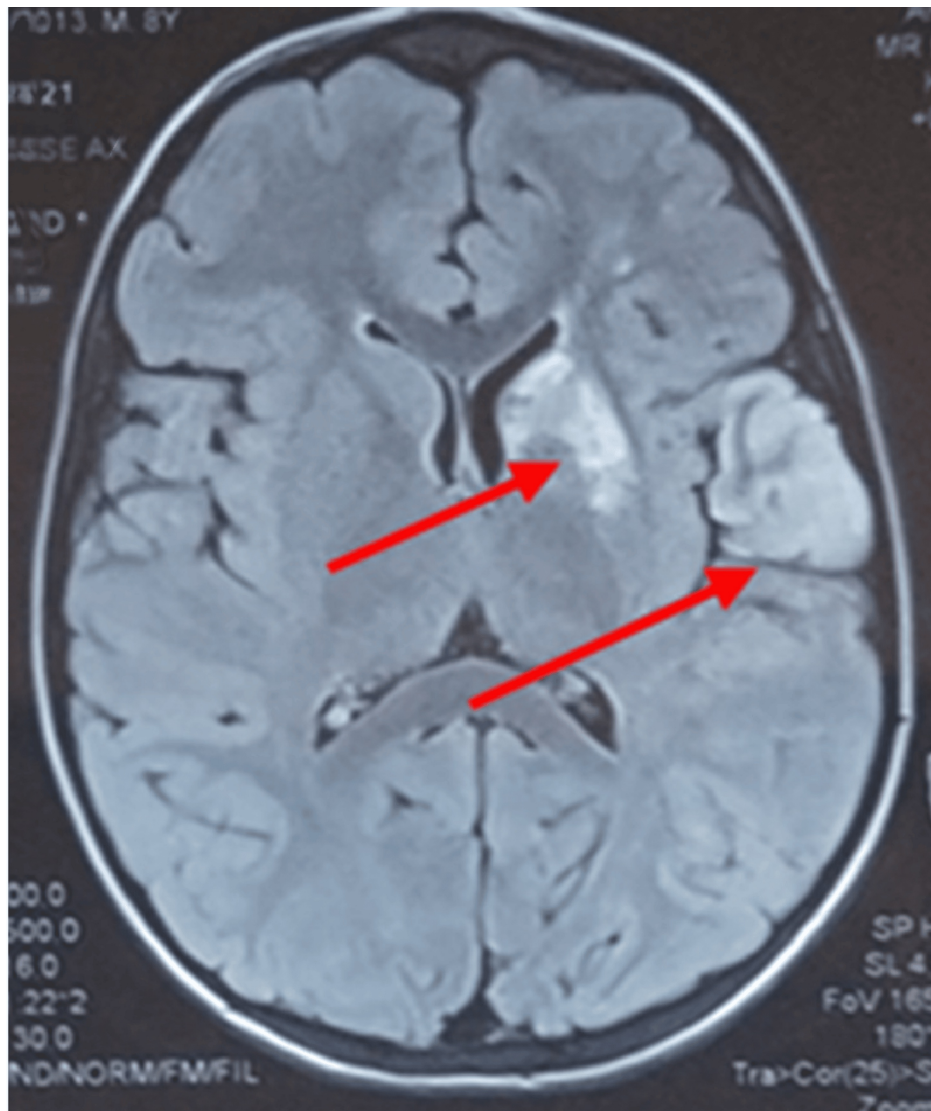


FIGURE 1: Brain MRI (FLAIR sequence) showing supratentorial left ischemic hyperintensities of different ages, with a subacute middle cerebral artery stroke associated with features consistent with Moyamoya syndrome and a hypoplastic internal carotid artery

FLAIR: fluid-attenuated inversion recovery, MRI: magnetic resonance imaging

The patient was managed with packed red blood cell transfusions, hydroxyurea therapy, and intensive physical rehabilitation. Given the occurrence of ischemic stroke in the context of HbSS disease, a chronic monthly transfusion program was initiated to maintain adequate hemoglobin levels and reduce the proportion of circulating sickled hemoglobin while awaiting access to exchange transfusion. During follow-up, the patient remained under close neurological surveillance with monitoring of motor recovery and cognitive outcomes.

Case 2

A 12-year-old boy born to first-degree consanguineous parents, both carriers of sickle cell trait (HbAS), was followed in our department for homozygous SCD (HbSS), diagnosed at the age of nine months. The diagnosis was confirmed by hemoglobin electrophoresis, which demonstrated HbS at 85.1%, HbF at 11.6%, and HbA2 at 3.3%. The family history was notable for a severe burden of sickle cell disease, including two siblings affected by HbSS disease and the death of another sibling at the age of three years due to sickle cell-related complications.

Before the cerebrovascular event, the patient was managed with a chronic scheduled transfusion program combined with hydroxyurea therapy, with regular monitoring of HbS levels as part of routine disease

management.

The patient presented with sudden-onset left-sided motor weakness occurring after an episode of status epilepticus complicated by postictal coma. Following recovery of consciousness, a persistent motor deficit was identified, raising suspicion of an acute cerebrovascular event.

Neurological examination revealed a conscious patient with left pyramidal syndrome associated with left central facial palsy. No sensory deficit or signs of increased intracranial pressure were noted. Brain CT demonstrated an acute ischemic infarction involving the deep territory of the right MCA, explaining the contralateral neurological deficits (imaging not available).

The patient was treated with packed red blood cell transfusions, hydroxyurea therapy, and intensive physical rehabilitation. In view of the high risk of recurrent cerebrovascular events associated with HbSS disease, a chronic monthly transfusion program was initiated. However, despite preventive therapy, the patient experienced two additional ischemic stroke episodes during follow-up, largely related to the unavailability of exchange transfusion, which limited optimal secondary stroke prevention.

Case 3

A 10-year-old boy with homozygous SCD (HbSS) had been followed in our pediatric department. Hemoglobin electrophoresis confirmed the diagnosis, showing HbS at 88.7%, HbF at 8.5%, and HbA2 at 2.8%. The patient had previously experienced severe sickle cell-related complications and was initially hospitalized for acute chest syndrome, a frequent and potentially life-threatening complication of HbSS disease.

The patient had a history of irregular medical follow-up, repeated loss to follow-up, and poor adherence to prescribed therapy, including hydroxyurea treatment.

Seven days after admission, while receiving supportive care for acute chest syndrome, the patient developed sudden-onset right-sided motor weakness. The neurological event occurred in the absence of trauma or other identifiable precipitating factors, raising concern for an acute ischemic stroke.

Neurological examination revealed right central facial palsy associated with a positive Bell’s sign and a right pyramidal syndrome predominantly affecting the lower limb. There were no reported sensory abnormalities or sphincter disturbances. Brain CT revealed multiple left hemispheric ischemic infarcts of different ages, consistent with recurrent cerebrovascular ischemic injury (imaging not available).

The patient received packed red blood cell transfusions, hydroxyurea therapy, and intensive physical rehabilitation and was placed on a chronic monthly transfusion program while awaiting exchange transfusion. Despite supportive management, his clinical course was complicated by worsening severe acute chest syndrome requiring intensive care admission. The patient subsequently deteriorated and died in the intensive care unit.

The clinical, radiological, therapeutic, and outcome characteristics of the three patients are summarized in Table 1.

Variable	Case 1	Case 2	Case 3
Age	8 years	12 years	10 years
Sex	Male	Male	Male
HbS (%)	78.7	85.1	88.7
Initial presentation	Monoparesis	Status epilepticus + hemiparesis	Acute chest syndrome then hemiparesis
Imaging	MRI + Moyamoya	MCA infarction	Multiple infarcts
Treatment	Transfusion + hydroxyurea	Transfusion + hydroxyurea	Transfusion + hydroxyurea
Outcome	Rehabilitation	Two recurrences	Death

TABLE 1: Clinical, radiological, therapeutic, and outcome characteristics of the three patients
MCA: middle cerebral artery, MRI: magnetic resonance imaging

Discussion

Stroke, particularly ischemic stroke, represents the most severe neurological complication of pediatric SCD. Children with homozygous SCD are at the highest risk of ischemic stroke, particularly between 2 and 16 years of age [3]. In our series, all patients were boys with HbSS disease. This male predominance may reflect the characteristics of our small cohort. However, current evidence does not support male sex as an independent risk factor for cerebrovascular complications in SCD.

The neurological manifestations observed in our patients were mainly characterized by motor deficits, central facial palsy, and cognitive impairment. These findings are consistent with previous reports, in which hemiparesis represents the most frequent clinical manifestation of overt ischemic stroke in SCD [7]. However, stroke may occasionally present with subtle or nonspecific symptoms, potentially leading to delayed diagnosis, particularly in children with silent or minimally symptomatic cerebrovascular events [1,7]. Therefore, regular follow-up, including neurological assessment and appropriate cerebrovascular screening, is crucial for the early detection of warning signs and timely intervention in patients with SCD at high risk of severe complications.

Additionally, neuroimaging plays a central role in the diagnosis, characterization, and follow-up of cerebrovascular complications in children with SCD. Brain CT remains an important first-line examination in emergency settings because of its rapid accessibility and ability to exclude intracranial hemorrhage and detect established cerebral infarctions. However, its sensitivity is limited during the early phase of ischemic events and for the detection of small or silent cerebral lesions [5].

Brain MRI, particularly with diffusion-weighted imaging (DWI), is considered the reference modality for the evaluation of ischemic brain injury in children with SCD, allowing early detection of acute cerebral ischemia, characterization of infarct patterns, and identification of chronic or silent cerebral infarctions. In addition, MRI combined with magnetic resonance angiography (MRA) provides detailed assessment of cerebral vasculopathy, including arterial stenosis, occlusion, and collateral vessel formation suggestive of Moyamoya syndrome, which represents a severe progressive vasculopathy associated with a high risk of recurrent stroke and poor neurological outcomes [5]. In our first patient, brain MRI revealed multiple ischemic lesions associated with Moyamoya syndrome, a severe progressive cerebral vasculopathy recognized as a major risk factor for stroke recurrence and poor neurological outcomes.

Stroke prevention remains a major objective in the management of pediatric SCD. International guidelines recommend annual transcranial Doppler (TCD) screening for children with SCD, particularly those with HbSS disease, to identify patients at increased risk of cerebrovascular events [2,5]. TCD is considered the reference screening method for stroke risk assessment. It measures cerebral blood flow velocities in major intracranial arteries, particularly the middle cerebral artery and internal carotid artery. Time-averaged mean maximum velocities ≥ 200 cm/s are considered abnormal and are strongly associated with an increased risk of ischemic stroke [3,4].

In high-income settings, abnormal TCD velocities are managed with regular transfusion therapy to maintain HbS $< 30\%$, which significantly reduces the risk of first stroke [4,5]. However, in low-resource settings where chronic transfusion programs and iron chelation are often unavailable, hydroxyurea (≥ 20 mg/kg/day or maximum tolerated dose) represents an accessible alternative to reduce cerebrovascular risk, particularly in settings where transfusion strategies are difficult to implement [5]. The SPHERE study demonstrated that dose-escalated hydroxyurea therapy was associated with a significant reduction in stroke risk among children with SCD, supporting its role in cerebrovascular prevention strategies [8].

In our three patients, they were managed with urgent transfusion therapy according to available resources. Exchange transfusion, which is recommended as the preferred approach when available, could not be performed in our setting due to limited access to this procedure [5,8]. Therefore, all patients received simple transfusion with packed red blood cells as an emergency measure, aiming to increase hemoglobin levels and reduce the proportion of circulating sickle hemoglobin (HbS). This therapeutic limitation highlights the challenges of managing cerebrovascular complications of sickle cell disease in low-resource settings, where access to exchange transfusion and specialized transfusion programs remains restricted.

This situation represents a major therapeutic challenge and potentially contributes to an increased risk of stroke recurrence and other severe complications. Among these complications, acute chest syndrome remains one of the leading causes of morbidity and mortality in patients with SCD, as illustrated by our third case [9]. Acute chest syndrome may also contribute to cerebral ischemic events through mechanisms involving hypoxemia, systemic inflammation, and increased vaso-occlusive phenomena, thereby exacerbating cerebral vascular injury [10].

Despite advances in screening and therapeutic approaches, stroke remains a major cause of long-term neurological disability and mortality in children with SCD, particularly among patients with severe cerebral vasculopathy, as observed in our second patient [7,10].

Overall, these three cases illustrate that ischemic stroke in children with SCD may occur through different pathways: progression of silent or established vasculopathy despite treatment, insufficient control of disease severity despite preventive strategies, or lack of access/adherence to recommended preventive care. Early identification of high-risk patients and improved access to preventive therapies are essential to optimize neurological outcomes and reduce the burden of cerebrovascular complications in pediatric SCD.

Conclusions

Stroke is a severe and potentially life-threatening complication of pediatric SCD that requires prompt recognition and management. Any focal neurological deficit in a child with SCD should raise suspicion of stroke and prompt urgent neuroimaging and therapeutic intervention. Our observations highlight the challenges associated with the management of pediatric SCD in resource-limited settings, particularly the restricted availability of TCD screening, chronic transfusion programs, and exchange transfusion facilities.

These limitations emphasize the need to strengthen comprehensive sickle cell care systems in low-resource countries through improved screening strategies, wider access to disease-modifying therapies, and the development of specialized multidisciplinary programs. Optimizing the management of cerebrovascular complications remains crucial to improving both neurological outcomes and overall survival in children with SCD.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Karel Weyo, Fabrice Ekomisa Boyele, Noufissa Benajiba, Karima Ryouni, El Alaoui Mounia

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Disclosures

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