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Challenges in Managing an Ischemic Stroke in a Child with Homozygous SS Sickle Cell Disease Associated with Cerebral Palsy: A Case Report

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Abstract

Introduction: Ischemic stroke is one of the most frequent complications of sickle cell disease (SCD) and may be associated with cognitive impairment. These two neurological manifestations can coexist in the same patient. **Objective:** To report a case of ischemic stroke and cognitive delay in a child with homozygous SS sickle cell disease. **Case Report:** We describe a 5-year and 8-month-old boy with homozygous SS sickle cell disease and epilepsy, non-compliant with his antiepileptic treatment, admitted for seizures that had progressively worsened over two months. A diagnosis of complicated major sickle cell syndrome was made. **Discussion:** The management of neurovascular, epileptic, and cognitive complications in this patient with cerebral palsy and sickle cell disease required evaluating the risk

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of severe anemia and stratifying the likelihood of stroke recurrence. **Conclusion:** Managing major sickle cell syndrome associated with neurovascular, epileptic, and cognitive complications in a child with cerebral palsy requires adherence to current evidence-based recommendations.

Keywords: Stroke, Sickle cell anemia, Exchange transfusion, Transcranial Doppler, Côte d'Ivoire

Introduction

Sickle cell disease (SCD) is an autosomal recessive hemoglobinopathy caused by a point mutation at the sixth codon of the β-globin gene. It is common among individuals of African and Caribbean descent but also occurs in Mediterranean, Middle Eastern, and Indian populations. Children with SCD have a high risk of cerebrovascular complications, including ischemic and hemorrhagic strokes, with a prevalence of approximately 14% before the age of 20. Cerebral ischemia in SCD results from "vaso-occlusion" due to deformation and rigidity of sickled red blood cells under hypoxic conditions. These abnormal cells obstruct small cerebral vessels, impair oxygen delivery, and may lead to cerebral infarctions and cognitive impairment. We report a case of ischemic stroke in a child with known sickle cell disease.

Review of the Literature

Epidemiology and Burden

Globally, SCD affects an estimated 300,000 newborns annually, with more than 75% born in sub-Saharan Africa (Makani et al., 2020; Tshilolo et al., 2019). Limited access to early diagnosis and comprehensive care contributes to high rates of complications and premature mortality among affected children. Ischemic stroke is among the most severe complications of SCD. In children with homozygous sickle cell anemia, the annual incidence is approximately 0.61–1.0% between ages 2 and 9 years, and the cumulative risk can reach 11% by age 20 (Ohene-Frempong et al., 1998; Dowling & Kirkham, 2021). Silent cerebral infarcts, often detected only by MRI, occur in up to 40% of pediatric patients and are associated with cognitive impairment and poor academic performance (Prussien et al., 2019; Ware et al., 2022).

Pathophysiology

The pathophysiology of stroke in SCD is primarily due to progressive intracranial vasculopathy. Repeated episodes of sickling lead to endothelial damage, inflammation, and intimal hyperplasia, resulting in stenosis and occlusion of large cerebral arteries, especially the internal carotid and middle cerebral arteries (Bernaudin et al., 2020). This process leads to hemodynamic insufficiency and infarction, particularly in watershed regions of the brain.

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Prevention and Management

The introduction of **transcranial Doppler (TCD)** ultrasound has revolutionized stroke prevention in SCD. It allows the identification of children at high risk for stroke based on increased cerebral blood flow velocities. The **STOP trial** demonstrated that chronic transfusion therapy could reduce the risk of first stroke by over 90% in high-risk children (Adams et al., 1998). In addition, **hydroxyurea therapy** has proven effective in reducing the incidence of vaso-occlusive crises and may offer partial protection against stroke when transfusion is not feasible (Ware et al., 2016; Bernaudin et al., 2020). However, in many African countries, TCD screening programs, blood transfusion services, and access to hydroxyurea remain limited, resulting in delayed diagnosis and suboptimal management (Ndeezi et al., 2016; Diallo & Guindo, 2021; Komba et al., 2023).

SCD and Cerebral Palsy: A Diagnostic Challenge

Cerebral palsy (CP) encompasses a group of permanent disorders of movement and posture caused by early non-progressive brain injury. Its coexistence with SCD is rare but clinically challenging. Baseline neurological deficits in CP can mask the onset of new strokes, delaying recognition and treatment. Only a few case reports describe the overlap between CP and SCD, and data remain scarce, particularly in African contexts (Hogan et al., 2018; Byiringiro et al., 2022).

Case Report

A 5-year and 8-month-old right-handed boy, not attending school, was diagnosed with homozygous SS sickle cell disease at 6 months of age but had no regular follow-up. In November 2022, he presented with an infarction in the right superficial sylvian artery territory, resulting in left-sided motor sequelae. He also had right structural focal epilepsy since age 4 years and 10 months, treated with carbamazepine 150 mg every 12 hours, with poor compliance.

He was admitted for recurrent left-sided tonic seizures with secondary generalization. There were no preceding signs of vaso-occlusive crisis. The neurological examination, performed one week after symptom onset, revealed a conscious, reactive, hemodynamically stable child with predominantly left-sided flaccid–spastic tetraparesis, focal seizures secondarily generalized during the assessment, impaired gait and posture, and delayed language development.

Brain CT showed a right-sided post-ischemic infarction with diffuse cortical atrophy. EEG revealed right hemispheric dysfunction without paroxysmal abnormalities.

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Laboratory tests revealed microcytic hypochromic anemia (Hb 5.7 g/dL), leukocytosis (16,000/mm³), and CRP at 12 mg/L; other results were unremarkable.

The patient received two simple red blood cell transfusions followed by three sessions of exchange transfusion, spaced 48–72 hours apart, performed in day hospital with support from a charitable organization due to financial constraints. Each session cost approximately 300,000 CFA francs (≈457 euros), usually scheduled every four months. Adjunctive therapy included hydroxyurea (500 mg, three times per week), folic acid (10 mg, twice daily), carbamazepine (150 mg, twice daily), diazepam (5 mg rectally), baclofen (0.3 mg/kg/day), and niflumic acid (400 mg daily). Clinical outcomes were favorable, with disappearance of seizures, reduced spasticity and pain, and improved mobility without decubitus complications. Follow-up transcranial Doppler ultrasound performed 21 days after treatment showed normal flow velocities (<170 cm/s) and HbS <30%.

Discussion

This case illustrates the complexity of managing children with sickle cell disease complicated by stroke, cerebral palsy, and epilepsy. Such clinical presentations are increasingly common in sub-Saharan Africa and underscore the challenges related to early detection, preventive treatment, and rehabilitation capacity. Sickle cell disease remains the most prevalent genetic disorder in sub-Saharan Africa and a leading cause of pediatric stroke. The prevalence of stroke in children with SCD ranges from 5–11% before the age of 20, with even higher cumulative risks in resource-limited settings (Tshilolo et al., 2019). Sequelae often include cerebral palsy–like motor disability and epilepsy, which worsen functional and social outcomes. The neurological findings in our patient were consistent with secondary generalized focal epilepsy and bilateral spastic pyramidal syndrome. According to Kirkham et al. (2021), clinical features of stroke in SCD patients resemble those in non-sickle cell patients, sometimes presenting initially with seizures, as in this case. Early screening using transcranial Doppler (TCD) ultrasound is the most validated strategy to identify children at high risk of stroke. Stroke risk is defined by mean velocities in the arteries of the circle of Willis: values ≥200 cm/s indicate a high risk (>20% within 12 months and >50% within 30 months) (Mazumdar et al., 2007; Kirkham et al., 2021). In our patient, post-transfusion and hydroxyurea velocities were <170 cm/s. If exchange transfusion is delayed, a simple transfusion should be administered promptly to raise hemoglobin levels to about 10 g/dL. Both treatments were used in this case. Optimal care for children with SCD and stroke in Africa involves:

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1. **Prevention** (TCD screening, hydroxyurea therapy),
2. **Acute and chronic management** (safe transfusions, hydroxyurea, antiepileptics), and
3. **Functional rehabilitation** (physiotherapy and community-based care).

Hydroxyurea plays a key role in both primary and secondary stroke prevention. Dose-escalation protocols provide greater clinical benefits than fixed-dose regimens (Abdullahi et al., 2022; John et al., 2020). Clinical trials such as SPRING and REACH confirmed hydroxyurea's effectiveness in reducing vaso-occlusive crises, anemia, and stroke risk (Tshilolo et al., 2019). Post-stroke motor disabilities are common, but access to rehabilitation services remains limited in sub-Saharan Africa. Community-based programs involving families and caregivers have proven effective in improving functional independence (Kakooza-Mwesige et al., 2017). Strengthening physiotherapy infrastructure and implementing early rehabilitation programs are therefore essential. Post-stroke epilepsy is also frequent, with prevalence twice as high in children with SCD compared with the general population (Lagunju et al., 2014). Lack of access to regular antiepileptic treatment due to low income remains a major challenge (WHO, 2019).

Finally, establishing national registries and implementing dedicated health coverage programs represent crucial steps toward reducing mortality and improving quality of life for children with SCD complicated by stroke, motor sequelae, and epilepsy, particularly in Côte d'Ivoire (Grosse et al., 2011).

Conclusion

The coexistence of stroke and sickle cell disease is common and presents a major therapeutic challenge in sub-Saharan Africa, particularly in Côte d'Ivoire, where care remains costly and limited.
Improving management requires:

- systematic TCD screening from the age of 2 years;
- early initiation of hydroxyurea or chronic transfusions;
- better access to antiepileptic drugs; and
- early functional rehabilitation and psychological support programs.

Integrating sickle cell disease care into national universal health coverage schemes would greatly improve survival and quality of life for affected children.

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