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Challenges in Managing an Ischemic Cerebral Vascular Stroke in a Child with a Homozygote SS Sickle Cell Associated with Cerebral Palsy: A Case Study

Muriel Amon Tanoh
Rosette Boundjike
Nancy Tanya De-Souza
Cedric Valery Kadjo
Mody Kone
Constance Yapo-Ehounoud
Ismaila Diakite
Christian Tanoh
Evelyne Aka Diarra
Berthe Assi
Félix Houphouët Boigny University, Abidjan, Republic of Côte d'Ivoire

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Abstract

Introduction: Ischemic stroke is one of the most frequently observed complications of sickle cell disease. It can also be associated with cognitive impairment. These two neurological expressions of the disease can coexist in the same patient. **Objective:** Report a case of stroke and mental retardation in a child with homozygous SS sickle cell disease. **Observation:** A child aged 5 years and 8 months, homozygous sickle cell anemia, epileptic non-compliant with his antiepileptic treatment, was admitted for epileptic seizures that gradually developed over 2 months. The diagnosis of severe complicated major sickle cell syndrome was established. **Discussion:** The management of neurovascular, epileptic, and cognitive complications of sickle cell disease in our patient with cerebral palsy and major sickle cell

disease required evaluating the risk of occurrence of severe anemia and stratification of the risk of stroke recurrence. **Conclusion:** Management of a major sickle cell syndrome associated with neurovascular, epileptic, and cognitive impact in a child with sickle cell disease and cerebral palsy must comply with recent recommendations.

Keywords: Stroke - Sickle cell anemia - Transfusion exchanges - Transcranial Doppler Ultrasound - Côte d'Ivoire

Introduction

Sickle cell disease is a hemoglobin disease transmitted in an autosomal recessive mode, due to a mutation in the 6th codon of the b-globin chain gene, common in patients of African and Caribbean origin. Children with sickle cell disease have a high risk of developing cerebrovascular complications such as ischemic or hemorrhagic stroke with a frequency of 14% before the age of 20. We will report a case of ischemic stroke in a child with a known sickle cell disease.

Observation

Identity of our patient

This is a child aged 5 years and 8 months, right-handed, not in school. He was known for symptomatic homozygous SS sickle cell disease since the age of 6 months without follow-up, an infarction of the right superficial sylvian artery with left motor sequelae occurred in November 2022, and right structural focal epilepsy since the age of 4 years and 10 months with left focal epileptic seizures treated with Carbamazepine 150 mg every 12 hours, non-compliant with treatment.

Reason for admission

He was admitted for left hemicorporeal tonic epileptic seizures with secondary bilateralization.

Neurological examination

It revealed a secondarily generalized left focal epileptic syndrome, a spastic bilateral pyramidal syndrome, and psychomotor regression evolving for 2 months.

Paraclinical explorations

Brain CT scan revealed a right sequelae infarction associated with diffuse cortical atrophy. The EEG performed revealed right hemispheric brain damage, without paroxysmal abnormalities. It was carried out after transfusion exchanges due to financial difficulties and the lack of social care

within our hospitals in our work context. Biology revealed hypochromic microcytic anemia at 5.7g/dl, leukocytosis at 16,000/mm3, CRP at 12mg/l. The other blood tests were without abnormalities.

Treatment

The patient benefited from a background treatment based on 2 blood transfusions of packed red cells while waiting for the transfusion exchanges to be covered by a social organization helping poor patients. Only after 21 days of hospitalization, he was able to benefit from a transfusion exchange session, the cost of which in Ivory Coast amounts to three hundred thousand CFA francs or 457 euros per session covered by the parents. The adjuvant treatment consisted of the administration of hydroxyurea: 500 mg*3/week, folic acid: 10 mg*2/day, an antiepileptic Carbamazepine: 150 mg*2/day, diazepam, and a muscle relaxant, baclofen, as well as an NSAID, niflumic acid.

Evolution

The evolution was favorable after administration of the treatment with disappearance of epileptic seizures, regression of spasticity and pain.

Monitoring

Control transcranial duplex ultrasound was performed 21 days after the 48-hour inpatient treatment to monitor the risk of recurrence of stroke. It was normal with mean velocities of the carotid arteries, anterior and posterior cerebral arteries <170cm/s and HbS <30%.

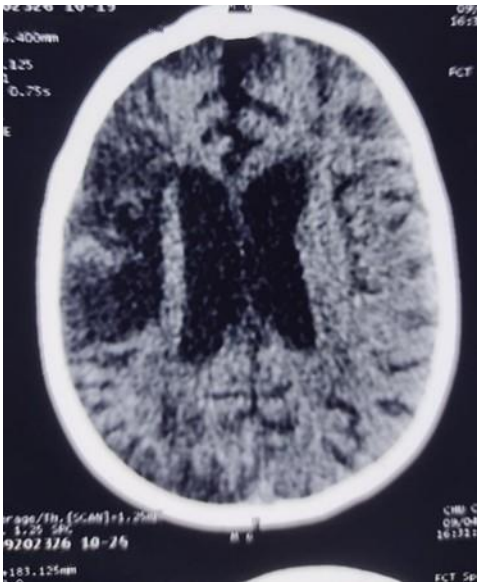

Figure 1: Cranio-encephalic CT scan revealing an old-looking superficial right sylvian infarct associated with diffuse cortical atrophy

Discussion

According to Ohene-Frempong and al., the spontaneous risk of symptomatic stroke is 11% before the age of 20, most often before the age of 10, with a peak between 2 and 5 years of age (Ohene-Frempong et al., 1998). Which is close to the age of our patient which was 5 years and 8 months? The neurological examination revealed secondarily generalized left focal epileptic syndrome, and bilateral spastic pyramidal syndrome. Indeed, Bernaudin and al. found that the clinical signs of stroke in sickle cell patients are like those in non-sickle cell patients. There may be acute focal involvement, including epileptic seizures as in this reported case (Bernaudin et al., 2011). The risk of stroke in sickle cell patients is predicted by measuring the average velocity in the arteries of the polygon of Willis using transcranial Doppler Ultrasound. The risk is said to be high for a velocity ≥ 200 cm/s (more than 20% risk of stroke within 12 months, more than 50% within 30 months) (Bernaudin et al., 2011) (Adams et al., 1992) (Maitreyi et al., 2007). Our patient had a velocity <170 cm/s after transfusion exchanges and hydroxyurea. When the establishment of the transfusion exchange takes a long time, a simple transfusion must be performed without delay to increase the hemoglobin level to around 10 g/dL. The objective being to improve oxygen extraction without increasing viscosity too much and to allow the circulation of non-sickle cell erythrocytes (Kossorotoff et al., 2012). These two treatments were administered to our patient. Likewise, hydroxyurea helps in the primary and secondary prevention of stroke (Sarah et al., 2011) (Ware et al., 2011). It has an alternative place in the transfusion program helping to reactivate the synthesis of fetal hemoglobin (HbF) limiting the growth of the HbS polymer (Winfred et al., 2011). This article reviews the diagnostic considerations and recommendations during the evaluation and management in the acute or long-term phase, following so-called 'silent' stroke in sickle cell patients, taking into account the recent update of the literature but also by the evaluation of the risk of recurrence of strokes in these patients and the risk of occurrence of significant neurocognitive deficits (Moustapha et al., 2021) (Nicolaeu et al., 2023).

Conclusion

Stroke and sickle cell disease are a common association. Care remains difficult to access in sub-Saharan Africa, including Côte d'Ivoire, given the delay and high cost of caring for children with sickle cell disease and cerebral palsy.

Conflict of Interest: The authors reported no conflict of interest.

Data Availability: All data are included in the content of the paper.

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Declaration for Human Participants: This study was approved by the institutional review board of Félix Houphouët Boigny University of Abidjan-Ivory Coast and the principles of the Declaration of Helsinki were followed.

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