Spontaneous epidural hematoma in a child with sickle cell disease

Authors

Engoba Moyen [1, 2]; Judicaël Kambourou [1,2]; Hugues Brieux Ekouelé-Mbaki[1], Caryne Mboutol Mandavo [1] ,Régis Moyikoua[1], Lydie Ocini N'Golet[1], Annie Rachelle Okoko[1,2], Gaston Ekouya-Bowassa[1,2]; Georges Moyen[1, 2.],Jean Louis Nkoua[1],

Affiliation

[1]Faculty of Health Sciences, Marien Ngouabi University of Brazzaville, Congo

[2]Paediatric Intensive Cares Unit, University Hospital of Brazzaville, Congo

Corresponding Author:

Doctor Engoba MOYEN

Email: engoba_m@yahoo.fr

Tel: +242055569305

Abstract

Children living with sickle cell disease often suffer life-threatening challenges including some uncommon neurovascular complications. For instance, epidural hematoma – collection of blood between the skull and the dura mater most commonly from a torn middle meningeal artery – is usually associated with trauma but a few cases of spontaneous epidural hematoma have been described in the litterature as complications of sickle cell disease.

In this issue, We report a case of spontaneous cerebral epidural hematoma observed in a 13-year-old boy with homozygous sickle cell disease, previously treated as severe malaria neuro-anemia form. Patient presented focal motor deficit and impaired consciousness. A magnetic resonance imaging (MRI) was performed and revealed the diagnosis. The clinical evolution was fatal due to an infection one week after the neuro-surgery.

Key words: Epidural hematoma, spontaneous, sickle cell disease, child.

Introduction

Epidural hematoma (EDH) is a collection of blood that forms between the inner surface of the skull and outer layer of the dura, which is called the endosteal layer. It is classically associated with head trauma[1-3]. The spontaneous epidural hematoma without trauma is rarely reported in literature [4-7] and usually occured in young patients between 10 and 40 years old. However, physiopathologic mechanisms of these hematoma are not completely elucidated [8].

Sickle cell disease (SCD) is the most common inherited hemoglobinopathy in the world. Estimates state that in Congo 1% of the population has homozygous sickle cell disease. It is responsible for a high mortality due to its most frequent and documented acute complications, including neurovascular complications [9-15]. Till date no case of spontaneous epidural hematoma has been reported in Congo.

In this issue, we report a case of spontaneous cerebral epidural hematoma observed in a 13-year-old boy with homozygous sickle cell disease.

Case report

A 13-year-old male, only son of a 44-year-old father who is a sergeant in the army and a 35-year-old unemployed mother was admitted to the Pediatric Intensive Care Unit of the Brazzaville University Hospital in April 2017 for a seizure.

The child is a known case of sickle cell disease since his 6 years of age with history of multiple previous vaso-occlusive crises, no previous blood transfusion, good developmental milestones and recommended childhood and adolescent immunisations. He is in junior high school.

Eight days earlier, he complained of chills followed 12 hours later by dizziness, progressive onset of occipitofrontal headaches severe in the morning, vomiting and photophobia. There was no history of any head trauma. Twenty-four hours later, fever at 38.5°C, moderate bone pain and asthenia were added to her symptoms. Initially, Ibuprofen was administered and assuming it was malaria, an intramuscular artemisinin treatment was administered at home at a dose of 2.6 mg/d for two days. The occurrence of unilateral tonic-clonic seizures with sudden onset impaired of consciousness prompted consultation and admission.

On admission, the patient presented with pallor and Glasgow Coma Score at 8/15. Pupils were bilaterally reactive. Signs of meningeal irritation (Kernig'sign, Brudzinsky'sign or neck stiffness) were

absent. His vitals were: temperature 38ºC, pulse 150 beats per minute and respiratory cycle 38 cycles per minute. There was a right fronto-temporal swelling; the liver span was enlarged at 16 cm, the soft liver edge being just palpable at the right costal margin. The weight was 30Kg for a height of 1.45m and a body mass index of 14.28Kg / m^2. The nutritional status was good.

Complete blood count showed leukocytosis 22,6.10^3/mm^3 with neutrophilia 10,2.10^3 , lymphocytes 11300/mm3, and anemia with hemoglobin 5.9g/dl for hematocrit 21.7% and mean corpuscular volume 87.9Fl. Platelets 160000/mm^3, C-reactive-protein 339 mg / l and serum creatinine 6.6 mg/L. Cerebrospinal fluid analysis was normal and hemoculture negative.

The diagnostic hypothesis of combined severe malarial anemia and cerebral malaria associated with bone vasculo-occlusive crisis was postulated. Treatment administered consisted of artesunate at 2.4 mg/kg/day and transfusion of packed red blood cells. At 72 hours of treatment, apyrexia was noted, but impaired consciousness persisted. On neurological examination, there was flattening of the nasolabial fold, hemiparesis and exaggeration of left tendon reflexes.

A different diagnostic hypothesis of severe malaria associated with brain abscess or severe malaria with cerebrovascular accident as its comorbidity was postulated. Magnetic resonance imaging of brain demonstrated heterogenous biconvex lens-shaped configuration in the extra-axial space, occupying the frontoparietal region and causing a mass effect on the homolateral ventricle with cingulate herniation under the falx cerebri. These findings were consistent with an epidural hematoma. The prothrombin rate was 83%, INR of 1.2 and the kaolin clotting time 29.1 seconds. We found a spontaneous epidural hematoma in a 13-year-old homozygous sickle cell patient admitted for mixed vaso-occlusive crisis (bone and anemia).). The decision was made to perform a craniotomy by opening a bone flap above the lesion which enabled discovery of a voluminous right epidural hematoma. There was no active per-operative bleeding, either at the level of the bones or the vessels of the dura mater. The evacuation of the hematoma was followed by suspension of the dura mater and then the repositioning of the bone flap. The postoperative awakening of the child took place 18 hours later. However, 48 hours later a fever appeared. Cerebrospinal fluid and blood cultures remained sterile. CRP was elevated to 160mg/l. Empiric antibiotic therapy with third-generation

cephalosporin at 100 mg/kg/day associated with metronidazole 30 mg/kg/day and ciprofloxacin 20 mg/kg/day was administered. Yet, the child died one week after the surgery with signs of infection.

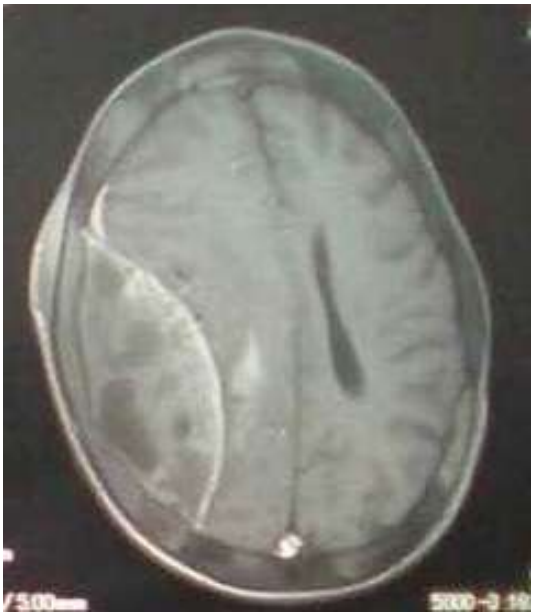

Figure 1: heterogeneous extra-axial biconvex lesion, in both T1 sequence

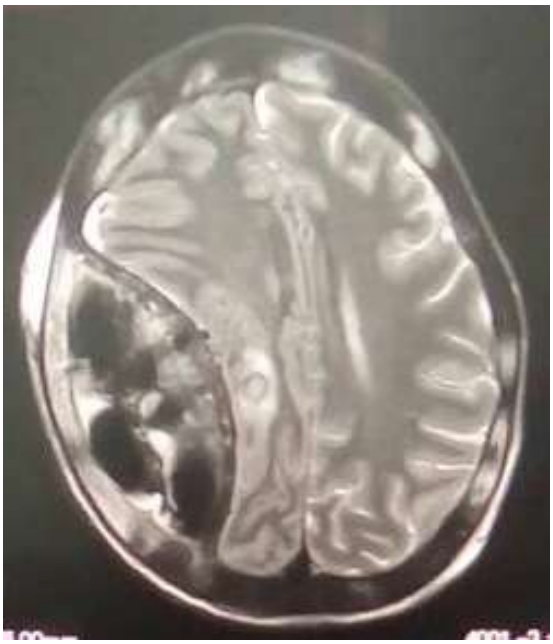

Figure 2: heterogeneous extra-axial biconvex lesion, in both T2 sequence

Discussion

Spontaneous EDHs are infrequent [4-7]. The neurovascular complications in children with sickle cell disease are well known [9, 14], both their frequencies and varieties. However, since areas with a high prevalence of homozygous sickle cell disease are those in which brain imaging is the least available and the most difficult to access, it is likely that the frequency of spontaneous EDH is underestimated.

Mechanisms for the appearance of spontaneous EDH are not well understood in children in general but also during sickle cell disease. Contributing factors have been identified in children [16]: elevation of blood pressure, blood transfusion, use of steroidal or non-steroidal anti-inflammatory drugs, bone infarction responsible for thrombosis of diploic veins with secondary epidural hematoma [17], a bleeding disorder and infection [4,5].

Similarly to others [7], we did not find in this case any history of trauma or bleeding disorder. Since an angiography was not

performed, a vascular cause could not be formally excluded.

Various teams [4, 7, 18] reported diversity in clinical manifestations. These, depending on the topography of the hematoma, could be an alteration of the higher mental functions for the frontal forms, a lateral homonymous hemianopsia in the occipital forms, signs of intracranial hypertension in the frontal and occipital forms, or hemiplegia in the parietal forms. It may be, as in this case, headaches and impaired consciousness. More rarely, parietal swelling has been reported on the side of the hematoma [4]. But in all cases, the diagnosis is made with brain imaging, including brain CT and magnetic resonance imaging, revealing signal intensities with spontaneous biconvex lens-shaped lesion. These diagnostic methods specify the characteristics of the hematoma: location, extent, impact on the brain and particularly mass effect on the ventricles [8].

Treatment and evolution

In some cases, because of the possibility of resorption of the hematoma, observation alone is advised [6]. Neurosurgical treatment is necessary when the hematoma is of great abundance: this is the choice we made for our case [4,18]. When the diagnosis is early and the management is carried out with a suitable technical platform, the evolution is often favorable [5, 6,19]. However, cases of adverse evolution have been reported [18]. In our case, the death of the child could be explained by the conjunction of several factors: the delayed diagnosis, the recourse to unsuitable therapy such as anti-inflammatory drugs, the volume of the hematoma and its impact on the brain manifested by signs of intracranial hypertension; the presence of a brain herniation below the falx cerebri, infection; failure to identify causative organisms consequently the possible unsuitable nature of the empiric antibiotic therapy, and especially the severity of this infection testified by a multi-organ failure in the context of sickle cell disease [20].

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