

Case Report

Unilateral Papilledema Revealing a Subdural Hematoma in a Sickle Cell Patient

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Abstract: The authors report a case of papilledema revealing a subdural hematoma in a 24-year-old male subject with known SS homozygous sickle cell disease. The patient presented with a sudden unilateral visual acuity loss on the right side and violent headaches that were resistant to level 1 analgesics. The discovery of unilateral papilledema on fundus examination led to a CT scan which revealed a subdural hematoma. The evolution was favorable under corticoid and hypotonizing treatment with the total resorption of the papillary edema, a significant regression of the subdural hematoma after 10 days of treatment and a recovery of the visual acuity. The authors insist on the necessity of performing a fundus examination accompanied by a CT scan in all sickle cell patients in the context of intractable headaches.

Keywords: Headache, papilledema, subdural hematoma, sickle cell disease SS.

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INTRODUCTION

Sickle cell disease is the most common genetic hemoglobin disease in the world and is inherited in an autosomal recessive pattern [1]. In 1910, James Herrick described elongated (sickle-shaped), irregularly deformed red blood cells in a West Indian anemic patient (sickle cell anemia) and his trainee Ernest Irons detected the abnormality [2]. Hemoglobin S is characterized by the substitution of the hydrophilic glutamic acid at position 6 of the β -chain by a hydrophobic valine [2, 3]. The geographical distribution of sickle cell disease reaches its maximum frequency in the sickle cell belt extending from the 15th parallel of north latitude to the 20th parallel of south latitude with 5 to 20% of carriers in West Africa (Gentilini 1993). The World Health Organization estimates that over 5% of the world's population has some type of hemoglobinopathy [4]. Every year, approximately 300,000 children with sickle cell disease are born throughout the world and this number will increase to 400,000 by 2050 [5]. In Niger, the epidemiology of sickle cell disease is not well known [6]. The complications of sickle cell disease are polymorphous and polyvisceral and the eye is not left out. In this work, we present a case of unilateral papilledema which led to the discovery of a subdural hematoma in a sickle cell patient.

OBSERVATION

The patient was a 24-year-old homozygous sickle cell patient known since childhood and followed at the internal medicine department of the National Hospital of Zinder. He was referred to the ophthalmology department of the same hospital for violent headaches that did not respond to ordinary analgesics. He had a history of hospitalization for vaso-occlusive crises, the last of which lasted 17 days, during which he received analgesic and anti-inflammatory treatment (Perfalgan, Trabar, Profenid) and a blood transfusion of two 250 ml of iso rhesus group (O rhesus +) blood after severe anemia at 3.7 g/dl. On admission, the patient was in acceptable general condition with a Glasgow score of 15/15, but reported violent headaches in an atraumatic context. The ophthalmological examination of the day, noted a visual acuity of distance without correction at 10/10 on the right and 7/10 on the left. No oculomotricity disorders were noted. On biomicroscopy, the conjunctiva were icteric bilaterally, the anterior segments normal with normal direct and consensual photo-motor reflexes. The fundus was normal on the right, whereas stage III papilledema (Figure 1A red arrow) with vessel tortuosity (Figure 1A green arrow) and peripapillary hemorrhages were noted on the left (Figure 1A blue arrow). The rest of the fundus examination was normal bilaterally. An

orbitocerebral CT scan was ordered and showed a subdural hematoma in the occipital region (Figure 2A green arrows). The renal workup was normal with urea at 5.1 mmol/l and creatinine at 76 umol/l. Treatment was initiated with acetazolamide 250 mg one tablet per day, corticosteroid therapy with prednisolone 1 mg/Kg per day and potassium supplementation. The evolution

was favorable after ten days of treatment with improvement of the headaches. Visual acuity was 10/10 bilaterally. The fundus showed total resorption of the papilledema of the left eye (Figure 1B) and significant regression of the subdural hematoma (Figure 2B, blue arrow).

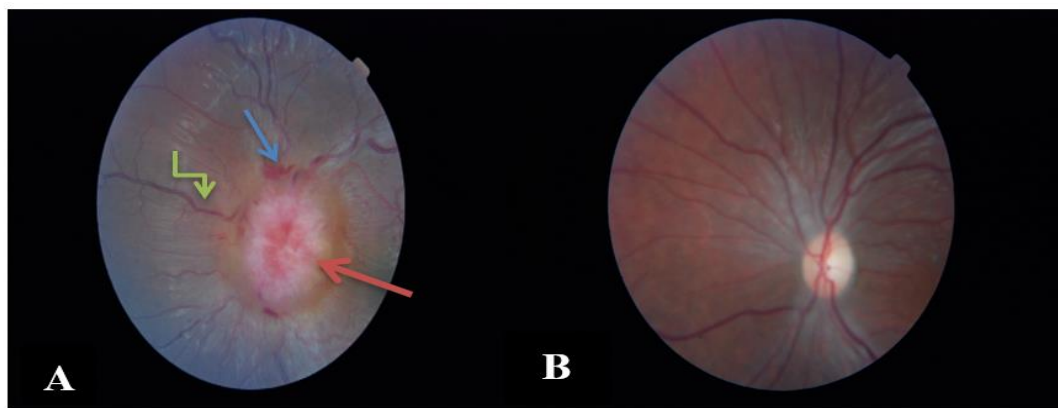


Figure 1: Retinography of the left eye: (A) Champagne cork papilledema with filling of the papillary excavation (red arrow), peripapillary hemorrhages (blue arrow) and vessel tortuosity (green arrow), (B) Normal fundus after 10 days of treatment

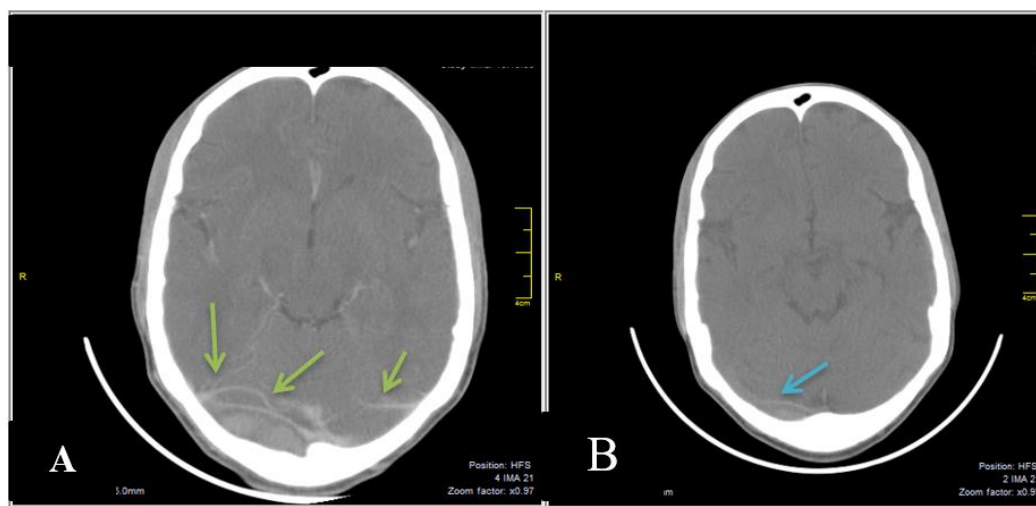


Figure 2: Cerebral CT: (A) Subdural hematoma occipital region (green arrows), (B) Regression of the subdural hematoma after 10 days of treatment (blue arrow)

DISCUSSION

Sickle cell disease is an autosomal recessive disease, resulting from the substitution of a glutamic acid by a valine in position 6 of the β -globin chain. This abnormal hemoglobin HbS polymerizes, during deoxygenation, into long fibers leading to rigidification and erythrocyte deformation at the origin of the two main manifestations: chronic hemolysis (anemia) and vaso-occlusive accidents [5, 7]. The increase in life expectancy of patients suffering from sickle cell disease, will crescendo with its complications in adults [8, 9]. These complications are multisystemic, and at the ocular level they are dominated by retinopathies, much more frequent in case of SC genotype [8, 10, 11], our patient is of SS genotype. Vaso-occlusive accidents

lead to optic nerve damage, causing visual impairment even before the appearance of retinal changes [12]. Clinically, our patient presented with violent headaches that were resistant to analgesic treatments and required a fundus examination. Indeed, frequent headaches in sickle cell patients are often interpreted as a clinical manifestation of chronic anemia, they may reveal secondary intracranial hypertension (ICH) and much more rarely idiopathic intracranial hypertension (IIH) [13]. Fundus examination in our patient revealed right unilateral papilledema. Papilledema is the clinical expression of various conditions rarely described as a complication of sickle cell disease. It is a liquid swelling of the optic nerve head, with filling of the excavation towards the inside giving a champagne cork aspect as it was noted in our patient. It is due to a

blockage of axoplasmic flow at the level of the sieve blade of the ethmoid. This papilledema can lead to blindness by optic atrophy in case of delay in diagnosis and management [14]. Note that papilledema was not described by Golberg in his classification of sickle cell retinopathy [10]. Papilledema in a sickle cell patient requires the exclusion of an acute intracerebral vascular lesion as a priority [13]. The CT scan requested to search for a cerebral vascular lesion in our patient allowed us to detect a subdural hematoma (SDH). Subdural hematoma occurs in more than 90% of cases over 50 years of age and in 50% of cases without any traumatic history and mostly in males [15]. Schneider and Hagarty in 1951 were the first to describe subdural hematoma as a complication of sickle cell disease. Its occurrence is exceptional in sickle cell disease and often not reported as a complication [16, 17]. The discovery of SDH is rarely fortuitous, most often manifesting itself by signs of ICH with headache resistant to the usual analgesics, vomiting and visual disorders such as diplopia and papilledema, as in the case of our patient, who was discovered with the help of CT scan, a key diagnostic test with a sensitivity of over 90% [18]. The pathophysiological mechanism of extradural hematoma in homozygous sickle cell patients remains poorly elucidated [19], although some authors have suggested a cranial infarction due to chronic extramedullary hematopoiesis due to anemia, which leads to proliferation of hematopoietic tissues, resulting in disruption of the cortex with extravasation of blood into the subdural space [20]. The prescription of analgesics was indexed by certain authors as being at the origin of the diagnostic delay [21], as was also the case for our patient. Surgery remains the treatment of choice for extradural hematoma by many authors [21], while others opt for medical treatment, especially in fragile patients, but with more careful monitoring because of the risk of secondary aggravation [15]. In his series Agaly *et al.*, 7.5% of patients benefited from medical treatment alone with corticosteroids and clinical improvement. Our patient also benefited from corticosteroid therapy associated with a hypotonic treatment based on acetazolamide with a favorable clinical response and radiological response: improvement of headaches, resorption of papilledema and subdural hematoma.

CONCLUSION

In the presence of headaches that are resistant to the usual analgesics in all sickle cell patients, a fundus examination is essential and the discovery of papilledema should lead to a search for cerebral complications such as subdural hematoma, even in the absence of clinical neurological manifestations. Prompt and adequate medical treatment can ensure clinical and paraclinical improvement avoiding surgery in these often fragile patients.

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