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Case Report

Multiple Neurological Complications in an Adult Sickle Cell Patient: A Case Report from the Libreville University Teaching University Hospital

Complications Neurologiques Multiples chez un Drépanocytaire Adulte : À Propos d'un Cas au Centre Hospitalier Universitaire de Libreville

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ABSTRACT

Neurological complications are common in sickle cell patients, and most of them are either stroke, epilepsy or encephalopathy. Proper diagnosis requires repetitive brain imaging, once signs related to central nerve system impairment appear. The management of these patients often requires a multidisciplinary team because of the number and complexity of the clinical situation. We report the case of an adult female sickle cell patient whose diagnosis was a combination of subacute subdural hematoma, structural epilepsy and encephalitis. The patient recovered with treatment.

RÉSUMÉ

Les complications neurologiques sont fréquentes chez les drépanocytaires, et la plupart d'entre elles sont des accidents vasculaires cérébraux, des épilepsies ou des encéphalopathies. Un diagnostic correct nécessite une imagerie cérébrale répétée, dès l'apparition de signes liés à l'atteinte du système nerveux central. La prise en charge de ces patients nécessite souvent une équipe multidisciplinaire en raison du nombre et de la complexité de la situation clinique. Nous rapportons le cas d'une femme adulte drépanocytaire dont le diagnostic était une combinaison d'hématome subdural subaigu, d'épilepsie structurelle et d'encéphalite. La patiente s'est rétablie grâce au traitement.

INTRODUCTION

Sickle cell disease is a ubiquitous autosomal recessive monogenic hemopathy mainly found in the black population, (1) and the prevalence of which in Gabon is estimated at 21.1% (2). It is a mutation in the ß-globin gene with C and S phenotypes, which will give rise to SS, SC, and CC sickle cell disease (3,4). The SS phenotype is the most frequent in our country (2). Sickle cell disease is characterized by the sickling of red blood cells, responsible for the common clinical manifestations of anemia and vaso-occlusive crises (5,6). However, sickle cell patients are subject to many other complications, which are essential to know because they can be life-threatening, including neurological complications such as strokes, seizures, and extra and subdural hematomas (6–9). We present to you the case of a 42-year-old sickle cell SS patient with a history of ischemic stroke ten years ago, followed by hematology at the Libreville University Hospital Center, who presented with multiple neurological complications.

CASE REPORT

Mrs. MS, with sickle cell disease, aged 42, of Gabonese nationality, was admitted to a hematology hospital for vaso-occlusive crisis (VOC) of osteoarticular expression. Her history includes an ischemic stroke in 2013, following which she suffered motor aftereffects such as right spastic hemiparesis. His baseline hemoglobin is 9g/dL. Clinical and paraclinical investigations in search of the etiology of his CVO revealed a right basal lung disease treated with broad-spectrum intravenous antibiotic therapy. On day 7 of her antibiotic treatment, the patient presented storms of generalized tonic-clonic convulsive seizures with tongue biting and loss of urine. At this point, we strongly suggested the diagnosis of vascular epilepsy in this patient with a history of stroke. However, in the context of CVO, an acute symptomatic attack was not excluded. One requested a brain CT scan performed the same day (figure 1). with a non-specific biological assessment. The CT scan made it possible to retain the diagnosis of semi-recent left subdural hematoma and frontal falx of the brain.

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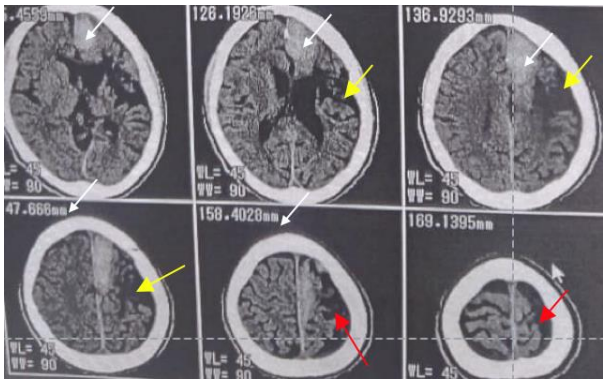

Figure 1 Brain CT in an axial section without injection of contrast product: Spontaneous hyperdensity in favor of HSD of the brain scythe (white arrows). Sequelae of left superficial sylvian ischemic stroke (yellow arrows) Cortical and subcortical atrophy more marked on the left (red arrows).

The patient was commenced on phenobarbital 100 mg/d orally, and a brain MRI was performed on day 15 to refine our diagnosis (figure 2).

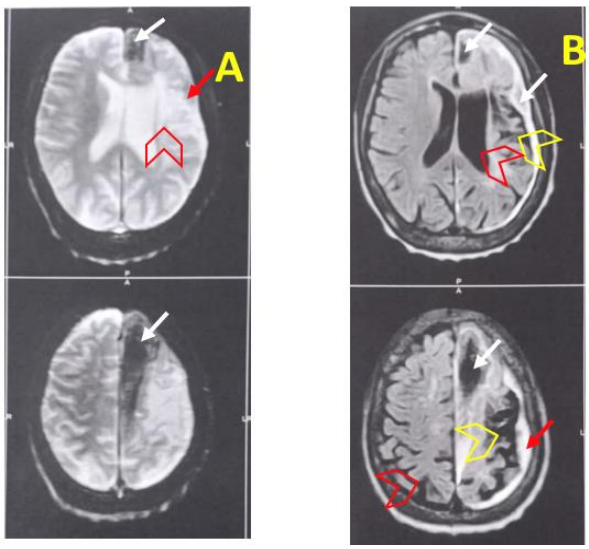

Figure 2 : Brain MRI in axial section T2 sequence (A) highlights hyposignal corresponding to HSD of the brain scythe (white arrows) and frontal (red arrow) and cortical and subcortical atrophy (red arrowhead). The T2 FLAIR sequence (B) confirms the subacute SDH of the brain scythe and frontalis (white arrow) associated with a hypersignal, which could be in favor of pachymeningitis (red arrow). Also, observe the cortical and subcortical atrophy (red arrowheads) and the aftereffect of stroke (yellow arrowhead)

On day 12, the patient presented with febrile confusional syndrome associated with head headaches and visual blurring. We then made the diagnosis of superinfected subdural hematoma after multidisciplinary consultation including the neurologist, the hematologist, and the neurosurgeon, and the patient was placed on dual antibiotic therapy consisting of gentamicin 160 mg/day and Ceftriaxone at a dose of 4g/day. Intravenously. After 21 days of Ceftriaxone and good clinical and biological progress, the patient was referred to the outpatient neurosurgery consultation for continued management of her SDH, to the outpatient neurology consultation for the management of her convulsive seizures and to the outpatient hematology consultation for continued management of his sickle cell anemia. Psychotherapy was also indicated for her. The patient is well controlled with phenobarbital, the neurosurgeon said there is no need for a procedure. After a follow-up of ten months, the patient presented at the outpatient consultation of neurology. She was seizure-free and no longer complaining of headaches. The CT scan showed no more hyperdensity in the subarachnoid spaces (figure 3).

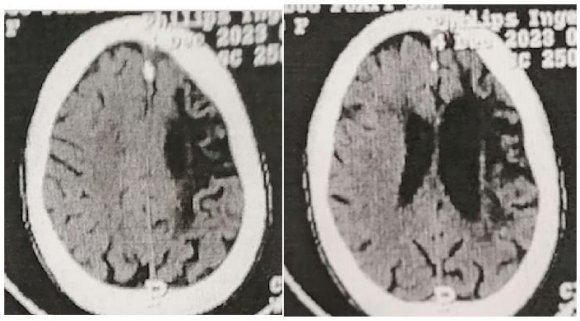

Figure 3 : Control CT Scan at ten months showing signs of ancient ischemic stroke with no signs of subdural hemorrhage.

DISCUSSION

Neurological complications are common in sickle cell patients, and of them, the most frequent are stroke, epilepsy, and encephalopathy (9). The phenotype most often affected by its complications is the SS phenotype (6,9). It is, therefore, appropriate to carry out brain imaging if signs related to brain damage appear. Subsequently, this imagery must be repeated iteratively if necessary. Thus, in our patient, it was the performance of a CT scan and then a brain MRI that made it possible to retain the diagnosis of subacute SDH. Although seizures are frequently associated with subacute SDH (10,11). We also considered the history of stroke in the superficial territory of the middle cerebral artery as a probable etiology of epilepsy. Although our patient's hematoma volume was rather significant, We only made the diagnosis quite late. This silent evolution could be explained by the cortical atrophy shown in imaging. Indeed, cortical atrophy might be responsible for good cerebral compliance in case of the creation of a third sector. (12). After a follow-up of ten months, the patient was seizure-free and headache-free and was showing only right hemiparesis. There was no more hyperdensity of subdural hemorrhage on the brain's CT scan, which tells us that in the case of SDH, we should avoid any neurosurgical procedures if the size of the hematoma does not threaten the patient's life (13).

CONCLUSION

Sickle cell disease is the most common hemopathy in Gabon, with a prevalence of 21.1% in particular series. It exposes us to significant morbidity and mortality, mainly due to its neurological complications, which are very numerous. In certain rare cases, the same patient may

present several neurological complications simultaneously or successively, such as stroke, subdural hematomas, epilepsy, encephalitis, or even meningoencephalitis. It is, therefore, essential to perform brain imaging and repeat it at the slightest suggestive sign. MRI is, consequently, the imaging of choice. The care of a sickle cell patient is, therefore, multidisciplinary.

DECLARATIONS

Authors' contributions

Dr. Michel-Arnaud SAPHOU DAMON worked with Pr. Marielle IGALA on defining the research project and it design. Pr. Marielle IGALA received the patient in the in-patient department of hematology while Dr. SAPHOU DAMON and Dr. ALENE performed the neurological examination. Both authors equally worked on the bibliography. All the authors reviewed the manuscript. Pr. KOUNA NDOUONGO Philomene has approved and reviewed the study at all its stages. All the authors have read and agreed the final version of the manuscript and its content.

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Informed consent form: The authors have obtained a written informed consent form from the patient. This document is in the possession of the corresponding author.

Ethical responsibilities: The study has been achieved by Libreville's Teaching Hospital Center rules regarding data privacy and the Helsinki Accords. The patient has received sufficient information before his participation.

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