

# Cerebral Venous Thrombosis and Sickle Cell (SC) Disease: Case Report and Literature Review

Heve Guouya \*

Department of Neurology Service Clinical, University Hospital Sylvanus OLYMPIO, Togo

\*Corresponding author: Heve Guouya, Department of Neurology Service Clinical, Service of Neurology of the University Hospital Sylvanus OLYMPIO, Togo, Tel: 0022890221672; E-mail: herve\_guinhouya@yahoo.fr

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## Abstract

The authors report a case of cerebral venous thrombosis in a thirty-seven-year-old sickle cell (SC) patient, who presented with a confusional state during hospitalization for a non-displaced fracture of the right leg. Exploration of the patient by cerebral CT and MRI brain angiography revealed superior sagittal sinus thrombosis. Biological examinations did not reveal any other hereditary or acquired hypercoagulable condition. The evolution was favorable under effective anticoagulation by low molecular weight heparin with per os relay by xarelto (rivaroxaban). The interest of this file resides in the scarcity of this observation in Black Africa, where, however, Sickle cell disease occurs.

**Keywords:** Cerebral venous thrombosis; Sickle cell disease; SC; Lome

## Introduction

Sickle cell disease [1] is a widespread genetic disorder resulting from a hemoglobin mutation in nearly 50 million people around the world [2] It is estimated that about 80% of sickle cell cases are concentrated in Africa sub-Saharan Africa [3]. Acute complications can include acute attacks of anemia, splenic sequestration, infections, stroke, acute chest syndromes or vaso-occlusive seizures. We report a case of CSV-induced SCR [1-3].

## Case report

Mrs. XY, 37 years old, with sickle cell SC disease, was hospitalized in the trauma department of CHU-SO for non-displaced fracture of the right leg. Three days after admission, the patient presented a state of obnubilation, which motivated her transfer to neurology after performing a cerebral CT scan. In her antecedents, the patient was third gesture, primiparous with two miscarriages at 3 months and 5 months. There was no known cardiovascular risk factor. On examination, in our ward, the patient was sleepy. The blood pressure was 120/63 mmHg, the heart rate was 77 bpm and the breathing rate was 16 per minute. The temperature was 36.2°C and the oxygen saturation under ambient air at 99%. She had conjunctival sub-jaundice. The neurological examination found a confusional syndrome. Examination of other devices was normal. The brain scan revealed a hypodense delta sign (Figure 1). The brain scan will be completed by a cerebral angioIRM which showed a thrombosis of the superior sagittal sinus (Figure 2). Biological tests including NFS-VS revealed anemia with a hemoglobin level of 10 g/dl and a hematocrit of 22%. The white line and platelets were normal. VS was at 20 mmH1. Electrophoresis of hemoglobin confirmed the following proportions of hemocyte S, 50% hemoglobin S and 50% hemoglobin C. Finally, a coagulation balance comprising the determination of antiphospholipid antibodies, homocysteine, prothrombin and factor V Leiden was negative. The patient was put on effective anticoagulation low

molecular weight heparin for two weeks and then relayed by bone Xarel to 20 mg/d for 165 days. The evolution was simple for both fracture and cerebral venous thrombosis. Radiological examination of the leg showed a consolidated fracture at three months. A phleboscanner test at one month showed a complete disappearance of thrombosis.

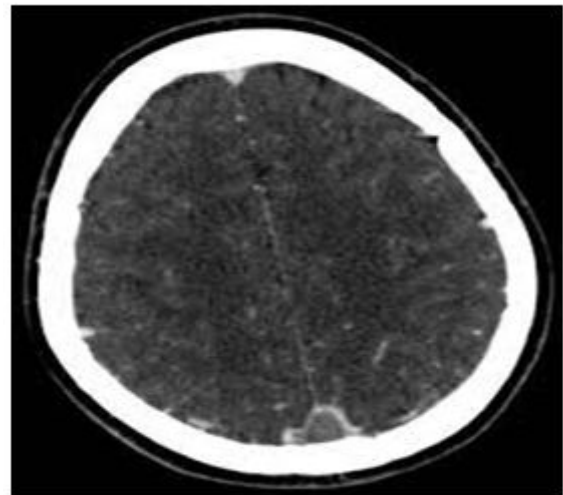
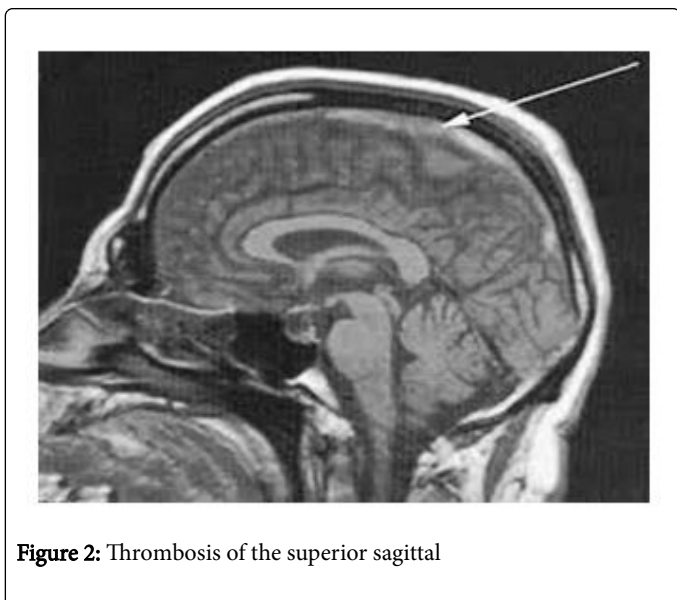


Figure 1: Hypodense delta sign

## Discussion

Sickle cell disease is not considered to be a common cause of cerebral venous thrombosis (CVT) [4]. However, while CTV may affect any superficial or deep cerebral vein, it appears that just as in our study, the sagittal sinus is the most affected [4,5]. Moreover, if the initial symptomatology was a confusional state in our patient, the

symptoms and signs are variable and nonspecific ranging from intracranial hypertension through nausea, vomiting, motor rocking deficiency, epileptic seizures or even coma [5,6].



In any case, the diagnosis is based on the detection of thrombus in the cerebral venous sinus system by CT or contrast MRI [4,7]. If in our patient, no etiology apart from sickle cell SC was found, it should be noted that various mechanisms related to sickle cell disease, according to several authors to explain the activation of coagulation in sickle cell disease [5, 8-12]. In fact, sickle cell patients have lower S, C protein levels, whereas plasma levels of von Will brand factor and Factor VIII are higher, and factors VII and V are low [4,8]. Other authors report in sickle cell patients platelet activation [4] increased adhesion of sickle cells to vessel endothelium [10] and an increase in the number of activated endothelial cells [11]. In addition, during the repeated cycle of sickling of haemoglobins, exposure of phosphatidylserine occurs which promotes increased adhesion of the sickle erythrocytes with activation of coagulation. However, the major pathogenic mechanism remains unknown, but it is thought that sickle-cell haemoglobins play a major role in the thrombophilic state of sickle cell disease [7]. And very often, these mechanisms and complications are manifested in the

homozygous form of sickle cell disease, the SS form. Our patient, however, had a heterozygous SC form. However, several authors are unanimous in recognizing that Sickle cell disease presents a similar clinical and biological picture of the SS form [3,5,8,11]. Finally, contrary to several authors [4] our patient had benefited from a low molecular weight heparin therapy relayed by an oral anticoagulant. It appears heparin potentially reduces mortality and severe disability [5,7] including low molecular weight heparin compared with placebo [10]. Other treatment strategies, in case of deterioration of the clinical condition of the patient despite low molecular weight heparin anticoagulation, may be justified. In particular, endovascular thrombolysis [7] or thrombectomy in hemorrhagic transformation of the lesion in relation to the TVC.

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