

Multiple hematoma in young patient with sickle cell disease

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Abstract

Stroke remains one of the most serious complications of sickle cell disease. Almost 11% of the patients will have a stroke before the age of 20. The etiology is hemorrhagic in a small part of the cases, which is associated with a high mortality. We present the case of a 19 year-old patient, with sickle cell disease and multiple associated complications. The patient was admitted with acute chest syndrome and developed a hemorrhagic stroke while hospitalized.

Keywords: Sickle cell disease, hemorrhagic stroke, anemia, acute chest syndrome.

Palabras clave: Enfermedad de células falciformes. Ictus hemorrágico. Anemia. Síndrome torácico agudo.

Introduction

Stroke is one of the most severe complications, and a major cause of sickle cell disease (SCD) morbidity. It affects 3% to 17% of all patients with SCD, the prevalence is 11% in patients under 20 years of age, with a high recurrence rate. Most cases are ischemic, however, hemorrhagic strokes account for 13% with higher prevalence between 20 and 30 years of age and a substantial higher mortality comparing to ischemic strokes^{1,2}. Several risk factors are identified^{1,3}, among them: low oxygen blood content, vasculopathy, acute infection, recent acute chest syndrome, classical vascular risk factors (hypertension, diabetes, dyslipidemia), previous stroke or a rapid elevation of hemoglobin.

Although many of the hemorrhagic strokes are subarachnoid hemorrhages after an aneurismal rupture, vasculopathy or even a Moyamoya disease, some cases present with other unexplained etiologies⁴. The standard of care is blood exchange transfusion to achieve hemoglobin S concentration <30% and hemoglobin concentration of 10-12 g/dl, but some studies failed to show benefit in patients with established vasculopathy^{4,5}.

Case Report

A 19 year-old African man with homozygotic (SS) SCD and multiple associated complications - recurrent priapism, retinopathy, transfusional iron overload, cholecystectomy after lithiasic cholecystitis, multiple vaso-occlusive crisis - was admitted to our emergency department with fever, dyspnea and lower left limb pain. He was compliant with his medication - hydroxyurea 1500 mg per day.

At clinical examination he was febrile (temperature 40.1 °C) with increased respiratory effort, tachypnea and low oximetry (89%) while breathing air. Pulmonary auscultation revealed diffuse diminished respiratory sounds. He was tachycardic with 130 bpm (sinusal rhythm) and his blood pressure was 115/68 mmHg. On his lower left limb, we observed a cicatricial chronic ulcer without signs of ongoing infection.

The first laboratory results revealed a hemoglobin concentration of 8.6 g/dl, normal white blood cell count with neutrophilia (81%),

Learning points

- Stroke is one of the major causes of sickle cell morbidity and mortality.
- Hemorrhagic strokes are less common (13% of strokes in SCD patients) but have higher mortality, although risk factors are identified, some cases present with unexplained causes.
- Although some studies failed to show benefit with blood exchange transfusion, it remains the standard of care in stroke prevention.

normal C-reactive-protein 1.3 mg/dl, elevation of hemolytic parameters (total bilirubin 15.18 mg/dl, conjugated bilirubin 2.19 mg/dl, LDH 568 UI/l), normal renal function and a blood gas analysis showed severe hypoxemia (pO₂ 58 mmHg, ambient) with hyperlactacidemia (3.7 mmol/l). Anteroposterior chest ray revealed a bilateral pulmonary opacity (figure 1) and cardiomegaly. A bed-side echocardiography was performed showing normal left and right ventricular function and size, moderate tricuspid regurgitation with a calculated pulmonary artery systolic pressure of 39 mmHg.

He was started with intensive IV fluids, analgesic treatment and empiric antibiotics. No anticoagulation nor platelet antiaggregation therapy was given, previously or during his hospitalization.

The patient failed to improve requiring orotracheal intubation and admission to the Intensive Care Unit. He was diagnosed with vaso-occlusive crisis evolving to acute chest syndrome. The repeated laboratory results sustained this clinical course with elevation of inflammatory parameters (leukocytosis 55000 /l, 91% neutrophilia, c-reactive protein 20 mg/dl), lower hemoglobin 6.7 g/dl with increased hemolysis (total bilirubin 30.6 mg/dl, conjugated bilirubin 3.1 g/dl and LDH 732 UI/l) and acute kidney injury (creatinine 2.1 mg/dl, urea 130 mg/dl). His hemoglobin S percentage was 81%, a blood exchange transfusion was performed to achieve HbS < 30% and hemoglobin of 10 g/dl.

Figure 1. Chest radiography at admission



Figure 2. MRI, 11nd day of hospitalization

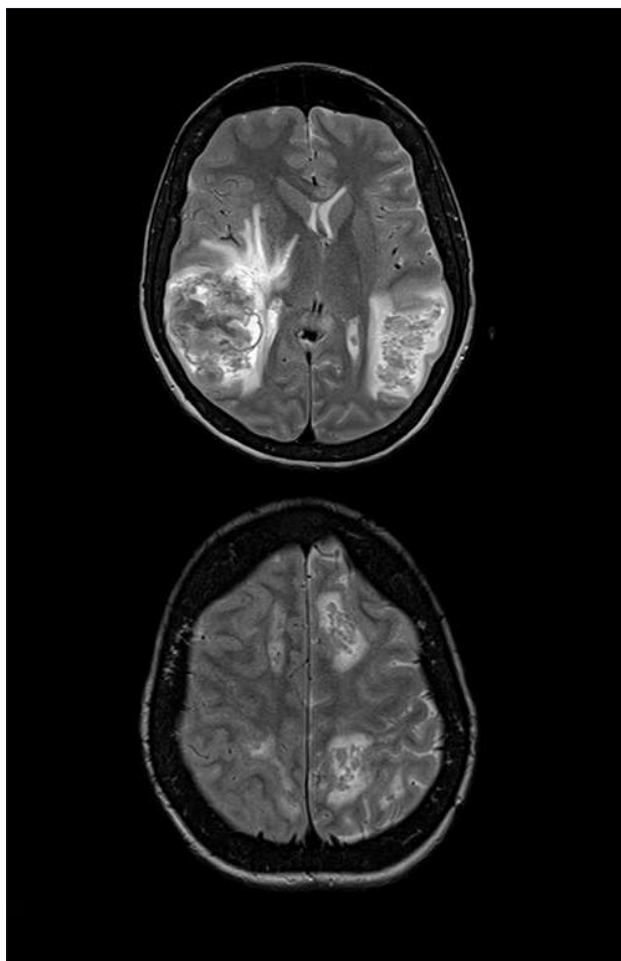
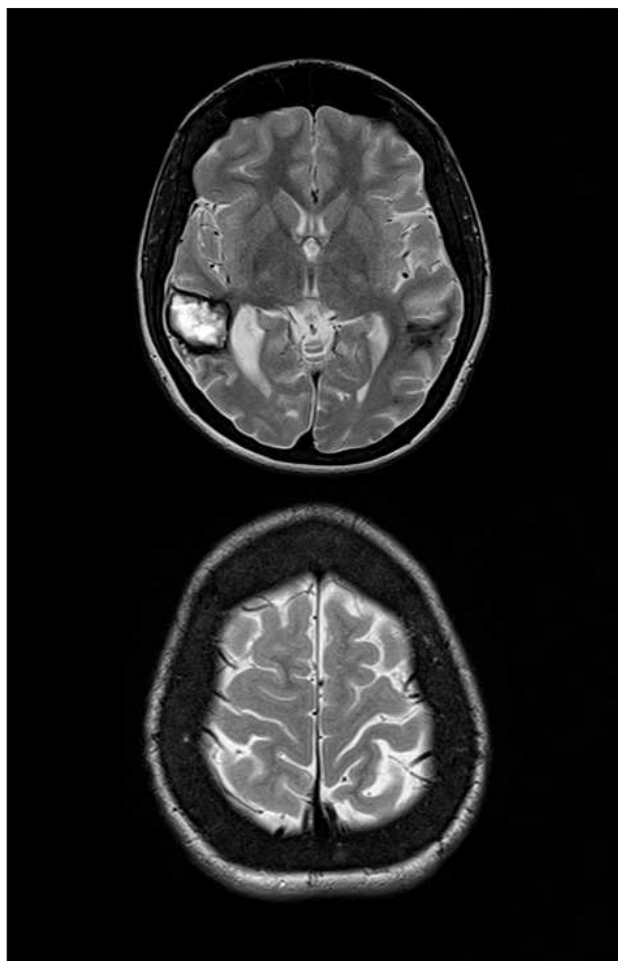


Figure 3. Follow-up MRI



During his hospitalization at the intensive care unit he was mechanically ventilated and given vasopressor agents (norepinephrine) for severe hypotension and septic shock. The patient improved, sedation was stopped daily to assess neurological status. On the 10th day of admission, he was unresponsive and had a hypotonic tetraparesis. His National Institute of Health Stroke score was 33. MRI (figure 2) angiography of the brain showed right (60 mm) and left (45 mm) cortico-subcortical parietotemporal hematoma, multiple frontal and occipitotemporal small lesions. The angiography excluded aneurisms, venous sinus thrombosis and Moyamoya disease pattern. He was evaluated by a Neurosurgery team who excluded vasospasm and decided there was no need for surgical intervention and decompression.

The patient recovered slowly, he was extubated and transferred to the Internal Medicine Department 20 days after admission. On the 28th day of hospitalization he was discharged home walking unassisted, he maintained a left hemiparesis grade 4, and modified Rankin scale of 1 (no significant disability despite symptoms, able to carry out all usual duties and activities).

At follow-up 2 months after discharge, he was symptom free. Repeat MRI (figure 3) showed retraction and reduction of the hematoma.

Conclusion

This case shows one of the most severe complications of sickle cell disease. Hemorrhagic stroke in patients with sickle cell disease has diverse physiopathology. Our patient

was not hypertensive, he did not have vasospasm, Moyamoya disease or venous sinus thrombosis. In the absence of the most frequent causes, the most likely mechanism is endothelial lesion with wall disruption secondary to vaso-occlusive processes. However, he had some risk factors for hemorrhagic stroke: vasopressor use, recent transfusion and acute chest syndrome^{3,4}. He was submitted to blood exchange transfusion according to standard of care which failed to prevent stroke. Despite the severity of the case presented, the patient had a good outcome without any symptoms and was able to carry out all usual activities.

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