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Spontaneous Extra Dural Hematoma in Sickle Cell Disease: A Case Report

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Abstract **Case Report**

Summary: Extra dural intracranial or intrarachid hematoma is frequently post-traumatic. Spontaneous extra dural hematoma is a rare entity, often associated with coagulopathy or vasculitis. We report a case of acute spontaneous extra dural hematoma causing altered consciousness in a 10-year-old child known to have sickle cell disease. Various pathophysiological mechanisms are involved.

Keywords: Extra dural hematoma, spontaneous, sickle cell disease.

INTRODUCTION

Sickle cell disease - or sickle cell anemia - is an autosomal recessive hemoglobinosis, whose carriers are often of African origin. Its clinical presentation includes a chronic hemolytic anemia associated with acute vaso-occlusive attacks of diffuse bone, pain and headache. Cerebral complications, being both rare and serious, are often ischemic and exceptionally hemorrhagic. [1, 2]

We report the case of an 11-year-old child known to be homozygous sickle cell anemic under treatment, admitted in our training for management of a spontaneous extra dural hematoma associated with a thrombosis of the superior sagittal sinus.

OBSERVATION

The patient M.S, 11 years old, followed-up for 9 years for homozygous sickle cell disease SS under hydroxycobalamin, folic acid and antibiotic prophylaxis, hospitalized several times for vaso-occlusive crises and repeated infections.

The history of the disease dates back to one day before his admission with diffuse bone pain for which he was admitted to the pediatric medical service. The evolution was marked by the appearance of intense headaches complicated secondarily by a loss of consciousness with a Glasgow score of 11 and right anisocoria, without any notion of seizure. He was hemodynamically and respiratory stable.

The patient underwent brain imaging with contrast injection objectifying:

- A 20-mm right fronto-parietal extradural hematoma responsible for the onset of subfalcine herniation.
- Thrombosis of the superior sagittal sinus.
- No visible bone fracture lines.

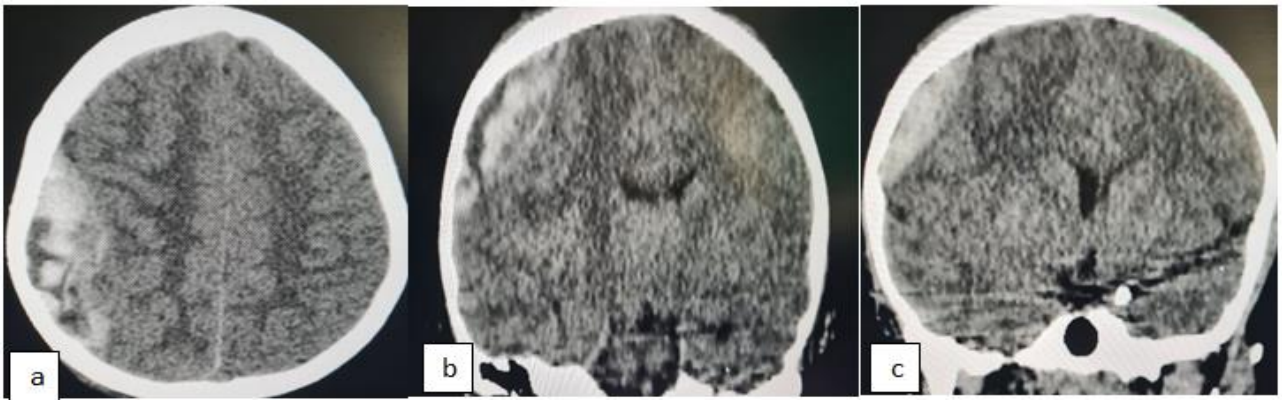

Figure 1: Spontaneous contrast brain CT a: axial section, b, c: coronal section

Extra-dural collection in a biconvex lens form, spontaneously hyperdense, with hypodense areas testifying acute bleeding, responsible for a mass effect on the midline with subfalcine herniation.

On the lab resultats, hemoglobin: 8.3, platelets: 250,000, PT 82%, total bilirubin 43, direct bilirubin 18.

Our management in terms of intensive care and anesthesia was as follows:

- Standard + invasive blood pressure monitoring
- Co-filling co-transfusion with 1 phenotyped O+ red blood cell + 2 fresh frozen plasmas.
- Rapid sequence induction + orotracheal intubation.
- Managing and struggling SBISOs (Secondary Brain Insults of Systemic Origin).
- Administration of 1 g/kg of Mannitol 10%.

The procedure consisted of a craniotomy, evacuation of the hematoma, control of the bleeding, and repositioning of the flap. There were no macroscopically visible lesions of the middle meningeal artery or the cranial vault.

The immediate postoperative radiological control was satisfactory.

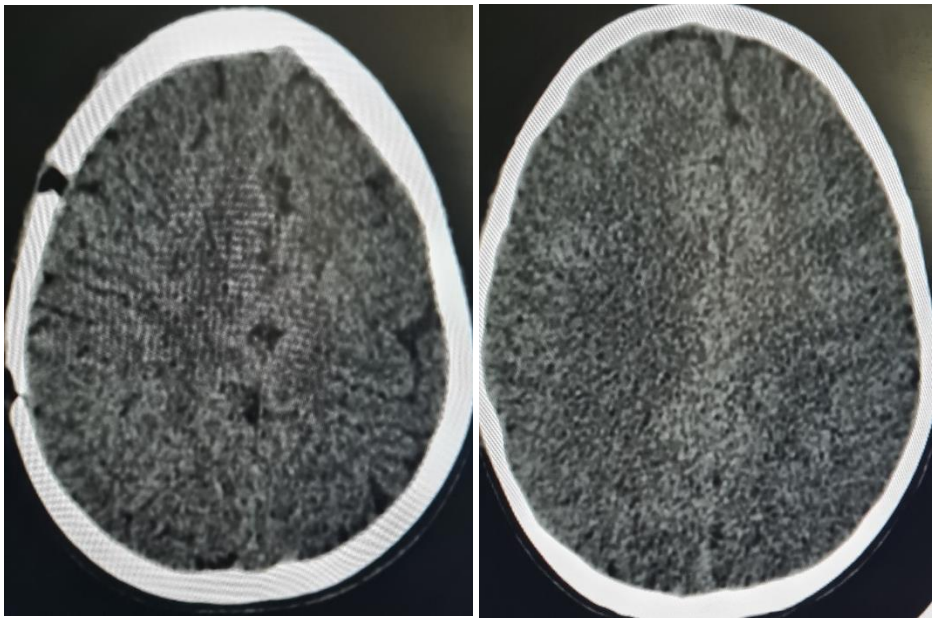

Figure 2: Spontaneous contrast axial slice brain CT: Complete regression of the extradural hematoma with right parietal bone defect in relation to the operative procedure

The postoperative evolution being favorable, the patient was extubated in the ICU after awakening, warming, analgesia and optimal gasometry. He was transferred 2 days later after complete motor and cognitive recovery.

DISCUSSION

During these extra-dural hematomas, the classical picture; associating to the cranioencephalic traumatic context with initial loss of consciousness after a free interval and a secondary aggravation, is replaced by headaches without notion of trauma [3, 4]. These

may be inaugural, or be part of the algesic picture of vaso-occlusive crises, as in the case of our patient.

The prescription of analgesics, a normal attitude in these cases, may be the cause of a diagnostic delay [5]. The appearance of consciousness disorders and/or focal signs should point to neurological complications.

CT and/or MRI will thus make the diagnosis by objectifying the hematoma. They may also show, in MRI in this case, bone remodelling aside the hematoma, such as an increase in the thickness of the skull diploë with hypersignal focus within it. These bone remodelling contribute to the understanding of the pathophysiological mechanism.

Indeed, while post-traumatic extra-dural hematomas are mainly due to traumatic vascular lesions [5], those that are spontaneous are due to other mechanisms. The hypotheses put forward to explain them can be classified into two groups:

- Observed in the long bones [6]. The extradural hematoma would be the result of Extensive hematopoiesis in the diploë and infarction of the skull bone [5].
- Extensive hematopoiesis that occurs in a localized area of the skull is the consequence of rapid deglobulation. This extensive hematopoiesis is responsible for an increase in the thickness of the diploë, and consequently the rupture of the internal and external tables of the skull bone, which will be at the origin of the bleeding [3]. Localized infarction of the cranial vault: this would occur during vaso-occlusive crises, as it is bleeding either by rupture of the internal table, which is associated with the elevation of the periosteum in the infarcted zone [7]; or by rupture of the vessels in the infarcted zone and/or its surroundings. This rupture would concern the veins, which, because of the weak blood flow, become congested.

CONCLUSION

Sickle cell disease being a frequent disease in our context, the hemorrhagic manifestations should not be underestimated considering their rarity, especially in front of a neurological deterioration contrasting with a non traumatic context. The physiopathology, although obscure, should be investigated more deeply in order to better understand these phenomena on the diagnostic, therapeutic and preventive levels.

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