Spontaneous Subgaleal Hematoma: A Rare Complication of Sickle Cell Disease

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Abstract

Spontaneous subgaleal hematoma is a rare complication associated with sickle cell disease. Although its cause is not clearly defined, it is frequently associated with vaso-occlusive crises. It may be a neurosurgical emergency when associated with epidural haematoma. The authors describe a case report of an adolescent with sickle cell disease that presented this complication, posing diagnostic and therapeutic challenges. This report highlights the importance of considering this diagnosis in patients with sickle cell disease that present with headache and haematoma of the head.

Keywords: Adolescent; Anaemia, Sickle Cell/complications; Haematoma, Epidural, Cranial/diagnosis; Haematoma, Epidural, Cranial/therapy

Introduction

Sickle cell disease is a serious chronic condition resulting from the presence of haemoglobin (Hb) S in the absence of HbA.[1] It includes homozygosity for the mutation (HbSS, sickle cell anaemia, the most common and severe form) and compound heterozygosities, in which an allele of β globin contains the S mutation and the other allele contains another mutation: HbC, β-thalassemia, HbD and HbO^{Arab}.[2]

Changes in red cell shape and elasticity are conditioned by the intracellular polymerisation of the HbS molecule in a deoxygenated state.[3] These altered erythrocytes cause haemolysis, microvascular vaso-occlusion and endothelial changes, leading to the acute and chronic manifestations and complications of the disease.

Spontaneous subgaleal haematomas are rare complications of sickle cell disease, with few described cases in the literature, especially in the paediatric age.[4,5] Due to their frequent association with non-traumatic epidural haematomas, [6-10] timely diagnosis and treatment are essential. The underlying pathophysiology is controversial and not fully understood.[5,6]

Case Report

A 15-year-old male patient with sickle cell disease under hydroxycarbamide and a history of multiple hospitalisations due to vaso-occlusive crises.

The patient was admitted to the emergency department for intense bilateral temporal and occipital headaches, with its onset five hours prior to admission and intensity of 10 (on a 1-10 numerical rating scale), not associated with trauma and with no improvement with analgesia. On admission, he had no fever, was normotensive, physical examination was normal, including neurological and fundoscopic examination. Analytically, he had haemoglobin 8.1 g / dL, leukocytes 12.3 x 10^9 cells/L (55% neutrophils), platelets 167 x 10^9 cells/L, C-reactive protein 4.05 mg/dL, international normalised ratio 1.3, activated partial thromboplastin time 25 s (21-30 s), D-dimer 10 296 µg/L (reference value <500.0 µg/L), 80% HbS. Head computed tomography without contrast showed no pathological findings.

There was an onset of fever and chest pain worsening on inspiration on the second day of hospitalisation. Pulmonary and cardiac auscultation were normal and chest X-ray showed no changes. An increase in inflammatory parameters (C-reactive protein 12.7 mg/dL) was observed and empirical antibiotic therapy with ceftriaxone was initiated. The patient was still experiencing left parieto-occipital headache, and an elastic and non-movable swelling with about 2 cm of larger diameter appeared (Fig. 1). Ultrasound excluded cellulitis or soft tissue abscess. Magnetic resonance angiography (MRA) of the head excluded acute cerebrovascular disease and head magnetic resonance (MRI) showed an hyperintense intradiploic lesion with left parietal subgaleal component on T2 and T1-weighted scans (Fig 2).

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Figure 1. Left parietal swelling with no associated wound.

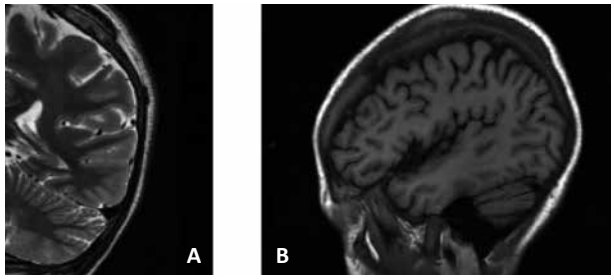

Figure 2. Magnetic resonance imaging. A. Coronal T2-weighted MRI: hyperintense left parietal intradiploic focal structural change, associated with a hyperintense small subgaleal collection on a T2-weighted scan. B. Sagittal T1-weighted MRI: Small subgaleal collection hyperintense on T1-weighted images.

The diagnostic hypotheses of subgaleal and left parietal intradiploic hematoma and extramedullary haematopoiesis were thus equated, as the *diploe* was involved.

Cytohistological analysis of the ultrasound-guided biopsy of the tumour showed a sediment with high cellularity, with predominant polymorphonuclear cells, some lymphocytes and histiocytes and very rare megakaryocytes, in a background of erythrocytes; haematopoietic cells were rare (0.2%). These findings were compatible with the diagnosis of haematoma.

A favourable clinical course was observed, with regression of the swelling and improvement of analytical parameters, with negative cultures. Discharge after erythrocytapheresis (red blood cell exchange with haemodilution of 750 ml; final HbS 46%).

The head MRI after six months revealed almost complete resolution of the previously described findings (Fig. 3). At the two-year follow-up, the patient showed no symptoms or relapse.

Discussion

We present the case of an adolescent with sickle cell disease, who is admitted to the emergency department with severe headache and develops a parieto-occipital swelling approximately 48 hours later with no associated trauma. In this context, it is urgent to exclude acute cerebrovascular disease (stroke), non-negligible complication in paediatric sickle cell patients. Ischaemic stroke

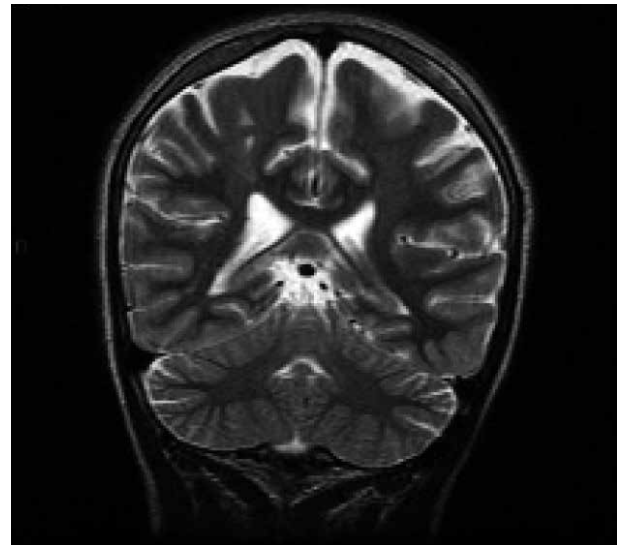

Figure 3. Magnetic resonance imaging. T2-weighted MRI: Significant resolution of the findings described on T2-weighted images six months after discharge.

occurs in 1% of the patients with screening and prophylaxis programmes, but may reach 11% without screening; haemorrhagic stroke occurs in 3% and silent infarcts reach 30%-40% of the patients.[11]

After acute intracranial pathology was excluded, the aetiology of the subgaleal swelling was investigated by head computed tomography and MRA. Since vaso-occlusive crises are frequently associated with infection, the diagnosis of osteomyelitis or its exclusion in these patients is of paramount importance, albeit often difficult. Both conditions are characterised by pain, fever and swelling, and no laboratory or imaging exam distinguishes these two entities perfectly.[12] We opted for broad-spectrum antibiotic therapy and clinical follow-up. Ultrasonography and head MRI reinforced the diagnostic hypothesis of subgaleal haematoma but did not allow for the exclusion of extramedullary haematopoiesis. For definitive diagnosis and symptomatic therapy, an ultrasound-guided biopsy of the tumour was performed, and its cytohistology was compatible with spontaneous acute subgaleal haematoma.

The most frequent cause of non-traumatic subgaleal hematomas in sickle cell disease is the occurrence of a bone infarct, which disrupts the cortical bone, leading to elevation and rupture of the periosteum with subgaleal haemorrhage (which may also reach the epidural space). Another postulated mechanism is the spontaneous rupture of vessels in the region of the bone infarct. A third hypothesis is that, in response to severe chronic anaemia, haematopoietic tissue proliferates and expands, causing the disruption of the cortical bone and extravasation of blood and bone marrow to the subgaleal and epidural spaces.[6] All these hypotheses are equally valid.

Partial red cell exchange transfusion (PET) is a procedure that involves the removal of HbS erythrocytes and their replacement by normal exogenous erythrocytes, improving oxygen transport capacity while preventing the occurrence of new haemolysis and vaso-occlusion events. It is currently considered the first-line therapy in children with sickle cell disease and ischemic stroke, improving its prognosis.[13] Its efficacy has also been demonstrated in primary and secondary prevention of stroke as well as in the treatment of acute thoracic syndrome.[14] On the other hand, haemorrhagic stroke treatment in sickle cell disease is controversial and has no formal evidence.[15] In these cases, PET has the same benefits stated for ischaemic stroke.[15] Given its potential severity, the very frequent association of this entity with the concomitant development of epidural haematomas,[8-10] and although its use is controversial, we chose to perform PET to prevent early relapse. The child did not start chronic transfusion therapy after this event.

In conclusion, this is a rare complication of sickle cell disease, whose nonspecific symptoms and signs require a timely differential diagnosis that includes several conditions. The head MRI is important to guide the diagnosis, although cytohistological analysis is mandatory for its confirmation. The treatment, which is conservative in most cases, may include more differentiated therapies such as exchange transfusion.

WHAT THIS CASE REPORT ADDS

- Although rare, non-traumatic subgaleal haematoma is a complication to be taken into account in sickle cell disease.
- The clinical findings are non-specific and the differential diagnosis includes very diverse conditions.
- The possibility of simultaneous presence/development of intracranial haematomas, which can be life-threatening, should be considered.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Hematoma Epicraniano Espontâneo: Uma Complicação Rara da Doença Falciforme

Resumo:
O hematoma epicraniano agudo espontâneo é uma complicação rara da doença falciforme, de causa não claramente definida, que surge mais frequentemente em contexto de crise vaso-oclusiva. Pode constituir uma emergência neurocirúrgica se associado a hematoma epidural. Reporta-se o caso clínico de um adolescente com anemia falciforme que apresentou esta complicação e que colocou desafios diagnósticos e terapêuticos. Sublinha-se a necessidade de considerar esta entidade no diagnóstico diferencial de doentes falciformes que se apresentam com cefaleia e tumefação craniana aguda.

Palavras-Chave: Adolescente; Anemia Falciforme/complicações; Hematoma Epidural Craniano/diagnóstico; Hematoma Epidural Craniano/tratamento