Spontaneous Subgaleal Hematoma: An Unusual Complication of Sickle Cell Disease

Renee-Pier Fortin-Boudreault*, Eden Story, Ewurabena Simpson, Donna Johnston and Christine Chretien
Division of Pediatric Hematology-Oncology, Children's Hospital of Eastern Ontario, Ottawa, Canada

Abstract

We report the case of a 17-year-old male with sickle cell disease who initially presented with headache and chest pain and subsequently developed fever and swelling of the scalp. On imaging, he was diagnosed with a subgaleal hematoma, and an underlying osteomyelitis could not be excluded. The patient was treated medically with analgesia, hydration and antibiotics and fully recovered. Spontaneous subgaleal hematomas are a rare complication of sickle cell disease with only few cases reported. Although it is thought to be associated with an underlying bone infarction, it is important to recognize this entity, as it can present with osteomyelitis or an epidural hematoma, which need prompt medical and in some cases surgical treatment.

Keywords: Sickle cell; Headache; Osteomyelitis; Subgaleal hematoma; Vaso occlusive episode; Hemolysis; Cephalohematoma

Introduction

Sickle cell disease is an autosomal recessive condition in which a mutation in the beta globin results in the formation of sickle-shaped red blood cells. Clinical manifestations are varied and include chronic anemia, acute vaso-occlusive episodes, infection and chronic organ dysfunction [1]. Spontaneous subgaleal hematomas are a rare manifestation of sickle cell disease, with very few cases reported in the literature [2,3]. They have been described more often in association with nontraumatic epidural hematomas [4-8], which can have devastating consequences including death [6-9].

Case Report

A 17-year-old male with known sickle cell disease (HbSS) treated with hydroxyurea was transferred to our emergency department from a peripheral hospital with complaints of chest and rib pain, as well as a bitemporal and parietal headache. He denied any accompanying neurological or infectious symptoms. Upon presentation, his vital signs were normal including temperature. His physical examination, including his neurological exam, was unremarkable. On investigation, his white blood cell count was 15.8 × 10^9/L, hemoglobin 97 g/L, platelets 290 × 10^9/L, reticulocytes 330 × 10^9/L, bilirubin 60 umol/L (conjugated bilirubin 0) and C-reactive protein (CRP) 40 mg/L. A chest x-ray was done and was normal. Because of the intensity of his headache (rated as a 10/10), the patient underwent a CT scan of his head. The CT scan was normal aside from an incidental finding of diffuse widening of the diploic space in the skull and a falx calcification at the anterior interhemispheric fissure. The patient was admitted to the inpatient unit and treated with a morphine infusion, regular ibuprofen and intravenous hydration for his pain. He was also continued on ceftriaxone, which had been started at the referring hospital for an oral temperature of 37.9°C. The day after his admission, the patient developed a fever up to 39°C orally with his headache getting better. Although unlikely, osteomyelitis and an underlying osteomyelitis could not be excluded. The patient was treated medically with analgesia, hydration and antibiotics and fully recovered. Spontaneous subgaleal hematomas are a rare complication of sickle cell disease with only few cases reported. Although it is thought to be associated with an underlying bone infarction, it is important to recognize this entity, as it can present with osteomyelitis or an epidural hematoma, which need prompt medical and in some cases surgical treatment.

Figure 1: MRI showing subgaleal hematomas over the (a) frontal and (b) parietal bones.

Keywords: Sickle cell; Headache; Osteomyelitis; Subgaleal hematoma; Vaso occlusive episode; Hemolysis; Cephalohematoma

*Corresponding author: Renee-Pier Fortin-Boudreault, Division of Pediatric hematology-oncology, Children's Hospital of Eastern Ontario, Ottawa, Canada, Tel: +1 613-249-8942; E-mail: rfortin-boudreault@cheo.on.ca

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and persistent high fevers and elevated inflammatory markers support hemolysis but could have been related to the subgaleal bleeds. Scalp swelling appeared, which was attributed to ceftriaxone induced and MRI. He also had a significant drop in his hemoglobin the day the diffuse widening of the diploic space in the skull, shown on the CT scan showed signs of extramedullary hematopoiesis as demonstrated by the marrow in the subgaleal and epidural spaces [4]. Our case definitely hypothesis is that patients with sickle cell disease have an abnormal is spontaneous rupture of vessels in the area of infarcted bone. A final and epidural spaces. Another related mechanism that has been suggested hypothesis is an underlying bone infarction that disrupts the cortical bone, causing periosteal elevation resulting in bleeding in the subgaleal and epidural spaces. Another related mechanism that has been suggested is spontaneous rupture of vessels in the area of infarcted bone. A final hypothesis is that patients with sickle cell disease have an abnormal anatomy of their skull due to extramedullary hematopoiesis and, in response to acute anemia, the hematopoietic tissue proliferates and expands, which disrupts the cortex and causes extravasation of blood and marrow in the subgaleal and epidural spaces [4]. Our case definitely showed signs of extramedullary hematopoiesis as demonstrated by the diffuse widening of the diploic space in the skull, shown on the CT scan and MRI. He also had a significant drop in his hemoglobin the day the scalp swelling appeared, which was attributed to ceftriaxone induced hemolysis but could have been related to the subgaleal bleeds.

In our patient, images on the MRI were suggestive of a superinfection and persistent high fevers and elevated inflammatory markers support this. There was no positive blood culture and we did not obtain a tissue specimen, so we could not be certain if there was an underlying soft tissue or bone infection. However, because of the clinical picture, and the fact that the hematoma would be an ideal medium for bacterial growth, we treated him as such. Distinguishing vaso-occlusive episodes from osteomyelitis can be very difficult in children with sickle cell disease. Both pathologies can present with pain, fever, and swelling. Moreover, there is no easy laboratory test or imaging that can reliably distinguish between the two entities [10]. Studies have shown that a vaso-occlusive episodes are 50 times more frequent than osteomyelitis in children with sickle cell disease [11]. One study concluded that swelling in a single site combined with fever and/or pain increased the probability of osteomyelitis in children with sickle cell disease [10]. Since no definite diagnosis could be made in our patient, we treated him as an osteomyelitis based on clinical findings and imaging.

In conclusion, we describe a patient with sickle cell disease who presented with headache and developed a non-traumatic subgaleal hematoma during his hospitalization. Only a few similar cases have been reported thus far [2,3]. It is important for physicians to recognize this occurrence, as it can be associated with epidural hematomas or infection, which can have a significant impact on the patient's outcome.

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Figure 2: Follow-up MRI showing interval resolution of previously seen subgaleal collections and cephalohematomas.