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Spontaneous Epidural Hematoma and Cephalohematomas in Sickle Cell Disease

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1. Abstract

Spontaneous epidural hematoma and cephalohematoma are rare complications of sickle cell disease. Although several mechanisms have been proposed, these have most frequently been described in association with bone infarcts involving the calvarium or orbits. We present a case of subacute epidural hematoma and cephalohematomas flanking a frontal calvarial infarct in a 17-year-old patient with sickle cell disease presenting with an acute vaso-occlusive crisis. The patient was managed conservatively with resolution of the hematomas at 2-month followup imaging.

2. Introduction

Sickle Cell Disease (SCD) is the most common inherited hemoglobinopathy with highest incidence in sub-Saharan Africa, the Mediterranean region, and India. A single mutation in the gene encoding the hemoglobin beta globulin chain results in hemoglobin S, which polymerizes in the deoxygenated state causing irregular RBC morphology, hemolysis, and microvascular congestion. Despite improvements in preventive and supportive treatments, life expectancy is currently reduced in patients with SCD by approximately 30 years [1, 2]. Neurologic complications in SCD are most commonly ischemic events. Intracranial hemorrhage has been reported in about one-third of patients with neurologic complications [3], but spontaneous extradural hemorrhage is exceedingly rare [4]. We report a case of a teenage patient with SCD who presented with scalp swelling in the setting of a vaso-occlusive crisis and was found to have a spontaneous epidural hematoma and bilateral frontal cephalohematomas.

3. Case Presentation

A 17-year-old African-American male with known SCD (Hemoglobin SS, HbSS) presented to the emergency department complaining of fever, fatigue, abdominal pain, and forehead swelling. His fatigue and abdominal pain had begun two days prior and gradually worsened. He became febrile the day prior to presentation with a reported temperature of 103°F (39.4°C). Several hours prior to arrival at the emergency department, he developed soft, nontender forehead swelling. The patient had recently been admitted for a vaso-occlusive crisis and had been discharged one week prior. He denied any recent trauma or headache. His home medications included hydroxyurea as well as ibuprofen and acetaminophen as needed for pain.

Initial vital sign measurements were notable for temperature 100.3°F (37.9°C), pulse 91 beats/min, blood pressure 123/79 mm Hg, respiratory rate 18/min, and oxygen saturation 99% on room air. Upon examination, the patient appeared uncomfortable, but was alert and oriented. He had left greater than right frontal scalp swelling without erythema or tenderness to palpation. The only

other notable finding on physical exam was upper abdominal tenderness to palpation without rebound tenderness. Initial laboratory evaluation was notable for hemoglobin level of 7.1 g/dL (hemoglobin at time of discharge one-week prior was 6.5 g/dL) with a reticulocyte count of 8.6%, white blood cell count of 11.4 x 103/μL (61% neutrophils, 26% lymphocytes, 0% band), platelet count of 610 x 103/μL, and creatinine level of 1.76 mg/dL (baseline 0.54 mg/dL).

A head CT was performed after administration of intravenous iodinated contrast given clinical concern for an infectious process, which revealed mixed attenuation collections along the outer table of the frontal calvarium bilaterally as well as a mixed attenuation lentiform epidural collection along the frontal convexity at midline with inferior displacement of the superior sagittal sinus (Figure 1). These intracranial and extra cranial collections were all bounded posteriorly by the coronal suture. Aside from marrow space expansion, the adjacent calvarium had a normal appearance by CT. The epidural collection exerted only mild local mass effect and there was no evidence of dural sinus thrombosis.

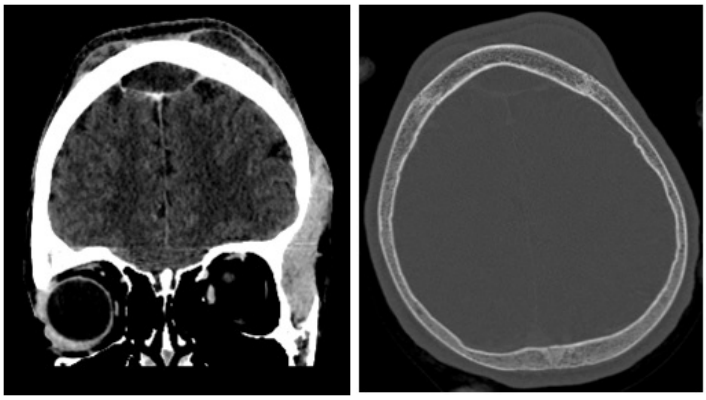

Figure 1. (A) Coronally-reformatted, soft tissue algorithm image from a contrast-enhanced head CT demonstrates bilateral frontal subperiosteal collections and an epidural collection with downward displacement of the superior sagittal sinus. (B) Axial, bone algorithm image shows a mildly expanded diploic space but otherwise normal appearance of the frontal calvarium.

The patient was admitted for pain control and for management of acute kidney injury (AKI). A renal vascular ultrasound excluded renal vein thrombosis and renal artery stenosis, leaving the likely cause of the patient's AKI to be nonsteroidal analgesic use for pain control. A brain MRI was performed without and with gadolinium-based contrast material to further evaluate the collections identified on CT in light of the clinical concern for an infectious process. On MRI, both the intracranial and extra cranial collections showed T1 signal hyper intensity, heterogeneous T2/FLAIR signal intensity, facilitated diffusion, and susceptibility artifact. Additionally, the frontal calvarium demonstrated T1 signal hyper intensity and hypo enhancement adjacent to these collections (Figure 2). The diagnosis of epidural hematoma and cephalohematomas was made given the respect of these collections to the coronal suture. The hematomas were favored subacute given the CT and MRI imaging characteristics.

The patient was evaluated by neurosurgery, who opted for conservative management given small size of the epidural hematoma. Further workup did not identify an infectious process and the patient was discharged two weeks later after improvement in his AKI and weaning from intravenous analgesia.

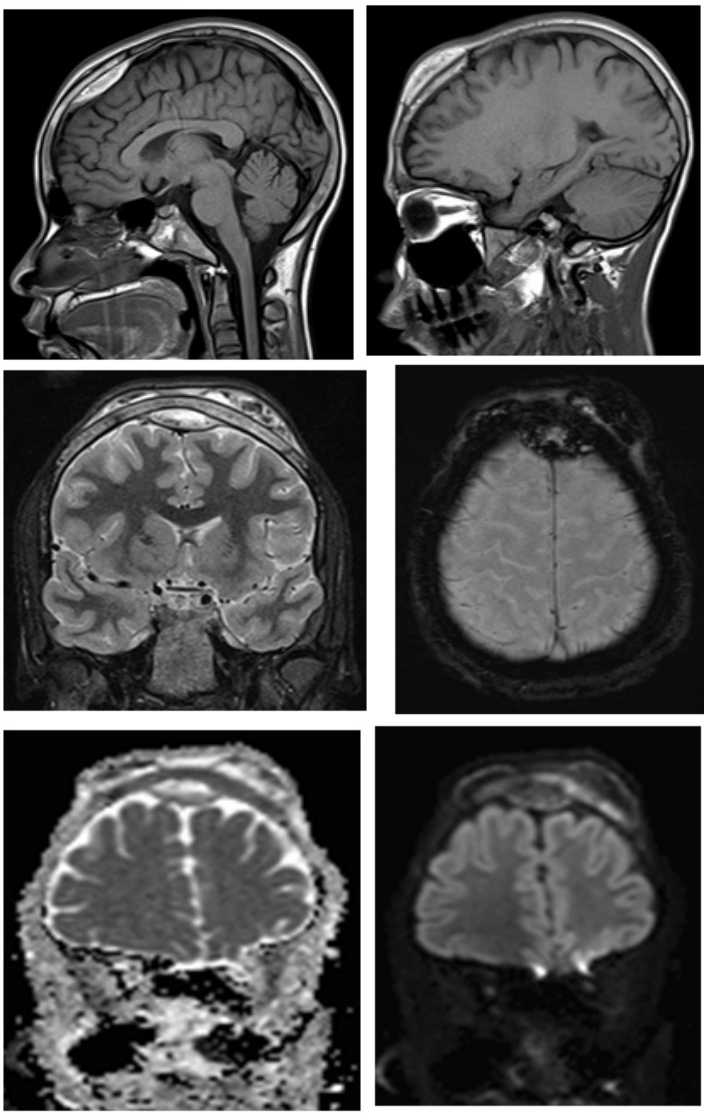

Figure 2. Multiple MRI sequences obtained on the same day as the patient's initial head CT. T1-weighted sagittal images (A and B), T2-weighted coronal STIR (C), susceptibility-weighted imaging (D), ADC map (E), and diffusion-weighted imaging (F). Images demonstrate epidural and subperiosteal collections with associated T1 signal hyperintensity (A and B), heterogeneous T2 signal (C), corresponding susceptibility artifact (D), and no corresponding restricted diffusion (E and F). These findings are consistent with cephalohematomas and epidural hematoma.

A follow-up brain MRI performed nearly two months later showed resolution of the collections as well as development of T1 signal hypo intensity and patchy enhancement in the frontal calvarium,

compatible with an evolving bone infarct (Figure 3). By the time of follow-up imaging, the patient reported complete resolution of his symptoms.

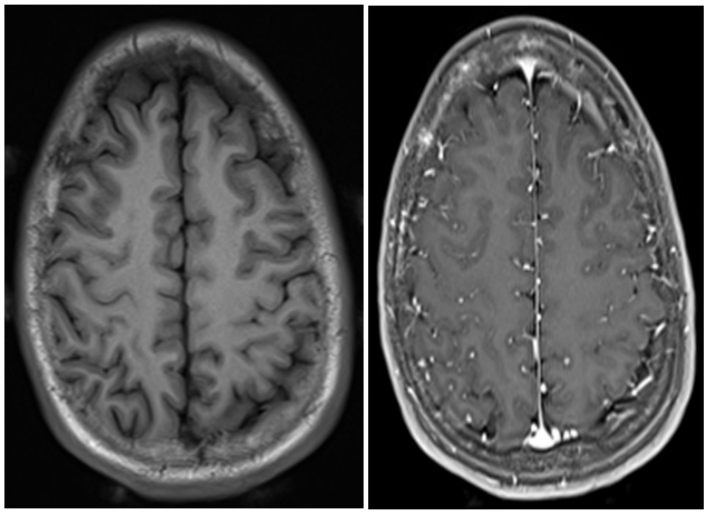

Figure 3. Two-month followup brain MRI. Pre- (A) and Post-contrast (B) axial T1-weighted images demonstrate resolution of the previous epidural hematoma and cephalohematomas with patchy T1 signal hypointensity and enhancement in the frontal calvarium, consistent with an evolving bone infarct.

4. Discussion

Spontaneous epidural hemorrhage (EDH) is rare and potentially fatal complication of SCD [4] and has also been reported in association with craniofacial infections [5], coagulopathies, hemodialysis [6], vascular malformations [7], following cardiac surgery [8], and in association with neoplasms involving the calvarium [9, 10]. We report a case of spontaneous epidural hematoma and cephalohematoma in a patient with SCD, which is an exceedingly rare combination only reported once previously in the literature [11].

Several mechanisms have been postulated with regard to spontaneous EDH in SCD. First, bone infarcts have been proposed as a causative mechanism leading to periosteal uplifting and tearing of adjacent vessels. While bone infarcts have been identified either by imaging or on surgical pathology in some cases [4, 12-15], others have shown no evidence of corresponding bone infarct [16-19]. While bone infarcts are a known complication of SCD, they most commonly involve long bones, vertebrae, or ribs, with the calvarium infrequently involved [14, 20]. Second, a veno-occlusive process leading to impaired venous drainage and resultant hemorrhage has been proposed [20]. Finally, hematopoietic tissue expansion related to acute on chronic anemia has been proposed as a mechanism for spontaneous hemorrhage as marrow space expansion may lead to calvarial microfracturing and corresponding hemorrhage [18, 19]. While the cause for spontaneous hemorrhage may certainly vary from case to case, we favor bone infarct as the precipitating factor for spontaneous hemorrhage in our patient given the progression of MRI findings in keeping with bone infarct in the frontal calvarium. The patient did not present with acute anemia, nor did contrast-enhanced CT or MR imaging show evidence for venous thrombosis.

Management of spontaneous EDH in patients with SCD may be conservative or surgical, with approximately half of cases of spontaneous EDH in SCD reported in the literature having undergone surgical management [4]. In our case, operative intervention was not pursued because of the small hematoma size and the patient's reassuring clinical status. The patient experienced no further intracranial complications and his hematomas resolved spontaneously.

Spontaneous scalp hematomas are less worrisome than intracranial hemorrhage, but given the association between subgaleal hematomas and intracranial hemorrhage in patients with SCD [12, 15], intracranial hemorrhage should be excluded in these patients. Unlike subgaleal hematomas which accumulate deep to the galea aponeurotica and superficial to the periosteum and commonly cross suture lines, cephalohematomas are subperiosteal in location and are typically bounded by sutures [21]. To date, subperiosteal hematomas in SCD have primarily been reported in the orbits, where they may be associated with orbital compression syndrome [11, 22]. In one case, both epidural hematoma and cephalohematomas/subperiosteal hematomas involving both the orbit and frontal calvarium were reported in a teenage patient [11]. Like our case, this patient also had imaging findings consistent with a corresponding frontal bone infarct. A literature review using the search terms "sickle cell disease" and "cephalohematoma" in the title, abstract, and/or keywords of articles indexed in the PubMed (MEDLINE) database returned only three additional case reports of cephalohematomas in infants with SCD.

Interestingly, our patient presented with only a minor complaint of forehead swelling and did not report headache or exhibit neurologic deficits. Because of the potentially falsely reassuring history and physical exam findings, it is important to have a high index of suspicion in patients with SCD presenting with headache, signs of increased intracranial pressure, or even scalp complaints to avoid delayed diagnosis and treatment [16]. It is important for clinicians to be aware of this association to ensure prompt diagnosis and management.

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