**INTRODUCTION**

Sickle cell disease (SCD) is a genetic hemoglobin disorder characterized by sickling of red blood cells under hypoxic conditions. SCD is the most common hemoglobinopathy, with high prevalence in areas of the Mediterranean region, Africa, South America, and East Asia.\(^1\) Its clinical manifestations are variable and can include chronic hemolytic anemia, vaso-occlusive crises (e.g., pain syndromes, stroke and bone infarction), and organ dysfunction.

Subgaleal hematomas can be classified into traumatic and non-traumatic hematomas. In SCD, spontaneous subgaleal hematoma is a very rare complication of unclear pathophysiology, with few cases reported in the literature.\(^2,3\)

We report a case of a young patient with SCD presented with a headache and painful scalp swelling due to subgaleal hematoma.

**CASE DESCRIPTION**

A 15-year-old male patient known to have SCD (HbSS) presented to the emergency department complaining of a 4-day history of persistent headache and two painful scalp swellings. The headache was constant with fluctuating intensity and was not relieved by simple analgesia. He denied any history of similar attacks nor trauma.

He had been taking hydroxyurea and folic acid for the management of SCD. His past medical history included multiple vaso-occlusive crises and a stroke with no residual neurological deficits.

Upon examination, he appeared pale, in pain, and had an icteric tinge. He was febrile (37.9°C), tachycardic (105 beats/min), and had a consistent blood pressure (105/65 mm Hg). His oxygen saturation was 99% on room air. The two scalp swellings, involving the right parietal and left occipital...
regions, were tender and fluctuant with normal overlying skin. The rest of the examination was unremarkable.

His laboratory results revealed a hemoglobin level of 9.3 g/dL, a hematocrit of 26% with a reticulocyte count of 7.4%, a white blood cell count of 13.6 × 10⁶/µL with a left shift (75.5%, neutrophils), and a platelet count of 415 × 10⁶/µL. Hemoglobin electrophoresis revealed an HbS of 81.1%. His liver function tests showed indirect hyperbilirubinemia (serum total bilirubin, 2.7 mg/dL; conjugated bilirubin, 0.3 mg/dL), while his renal function test results and coagulation profile were unremarkable. He was provisionally diagnosed as scalp hematoma with possible secondary infection and abscess formation. He was admitted for further investigations and management. He was commenced on intravenous hydration, morphine infusion, and regular lornoxicam for pain control. He was also empirically commenced on amoxicillin-clavulanate.

A computed tomography scan (CT) of his head revealed skull bone infarction at the right parietal and left occipital regions, with increased bone thickness. Additionally, two subgaleal fluid collections were observed in the same regions, measuring 8 and 7 mm at their maximum dimensions, respectively, without evidence of intracranial extension (Figure 1). Atrophy of the right hemisphere at the fronto-temporo-parietal lobes with dilatation of the ipsilateral ventricle was observed, which was consistent with the previous ischemic insults. We planned to continue on conservative management.

His headache gradually improved with nonsteroidal anti-inflammatory drugs and opioids. However, 2 days later, he started to develop right periorbital swelling associated with blurred vision. His ophthalmological evaluation showed no restriction of extraocular movements and intact pupillary reflexes. As orbital involvement was possible, he was kept under close observation. Over the next few days, no progression was noted, with gradual resolution of his symptoms. Therefore, the patient was discharged after 5 days. At follow-up 2 weeks after discharge, he was asymptomatic with complete resolution of his scalp swellings.

### DISCUSSION

Sickle cell disease is an autosomal recessive hemoglobin disorder resulting from the substitution of glutamic acid by valine at the sixth position of the beta-globin gene, which leads to sickling of red blood cells in response to hypoxic conditions. SCD is the most common hemoglobinopathy in Saudi Arabia, with variable prevalence across the country reaching up to 2.6% in the Eastern Province.

Pain is the hallmark and prominent cause of morbidity in patients with SCD. Frequent attacks of headache are common in children and adolescents, with an estimated prevalence of 24%-43.9%. Acute attacks represent a diagnostic dilemma, with multiple differential diagnoses including severe anemia, stroke, effect of opioids, vascular diseases, bone infarction, osteomyelitis, and intracranial or extracranial bleeding.

Extracranial bleeding may occur in different layers of the scalp. In neonates, different scalp hematomas may be frequently seen, especially after instrumental deliveries, including caput succedaneum and cephalohematoma. Subgaleal hematoma, defined as collection of blood below the galea aponeurotica layer, can be classified into traumatic and non-traumatic according to its mechanism. Traumatic subgaleal hematoma can be caused by minor head trauma such as hair pulling, whereas non-traumatic cases occur because of coagulopathies or rupture of superficial temporal artery aneurysms or arterio-venous fistulas.

Anatomically, the subgaleal space is located between the skull bones’ periosteum and the galea aponeurotica, extending anteriorly to the orbital ridges, posteriorly to the nuchal ridge, and laterally to the temporal fascia. It contains loose connective tissue and small emissary vessels connecting the extracranial venous system and the intracranial venous sinuses. Subgaleal hematoma results from the disruption of these emissary veins.
TABLE 1  Reported cases of subgaleal hematoma and/or epidural hematoma in patients with sickle cell disease

| No. | Year | Author | Age (y) | Gender | Hb defect | Scalp swelling | Skull infarction | Epidural hematoma | Subgaleal hematoma | Therapy                        |
|-----|------|--------|---------|--------|-----------|---------------|------------------|-------------------|-------------------|--------------------------------|
| 1   | 1987 | Mallouh| 13      | Male   | HbSS      | No            | Yes              | Yes               | No                | Bilateral craniotomies      |
| 2   | 1991 | Karacostas | 19      | Male   | HbSS      | Yes            | Yes              | Yes               | No                | Supportive care and dexamethasone |
| 3   | 1995 | Tony    | 35      | Male   | HbS-Thalasemia | No            | Yes              | Yes               | No                | Supportive care             |
| 4   | 1996 | Resar   | 13      | Male   | HbSS      | Yes            | Yes              | Yes               | Yes               | Supportive care             |
| 5   | 1996 | AlDabbous | 11      | Male   | HbSS      | Yes            | Unknown          | No                | Yes               | Supportive care             |
| 6   | 1996 | Pari    | 25      | Male   | HbSS      | Yes            | Yes              | No                | Yes               | Supportive care             |
| 7   | 1997 | Cabon   | 14      | Female | HbSS      | No             | Yes              | Yes               | No                | Bilateral craniotomies      |
| 8   | 2000 | Naran   | 16      | Male   | HbSS      | Yes            | Yes              | Yes               | No                | Supportive care             |
| 9   | 2001 | Ganeshe | 11      | Male   | HbSS      | Yes            | Yes              | Yes               | Yes               | Supportive care             |
| 10  | 2004 | Okita   | 2       | Male   | HbSS      | No             | Unknown          | Yes               | No                | Craniotomy                   |
| 11  | 2007 | Okita   | 12      | Male   | HbSS      | Yes            | Yes              | Yes               | No                | Supportive care             |
| 12  | 2009 | Dahdaleh| 18      | Male   | HbSS      | Yes            | No               | Yes               | Yes               | Bilateral craniotomies      |
| 13  | 2011 | Arends  | 19      | Male   | HbSC      | No             | Yes              | Yes               | No                | Supportive care             |
| 14  | 2011 | Sangle  | 15      | Male   | HbSS      | Unknown         | No               | Yes               | No                | Bilateral craniotomies      |
| 15  | 2012 | Babatola| 18      | Male   | HbSS      | No             | No               | Yes               | No                | Craniotomy                   |
| 16  | 2012 | Bölké   | 19      | Male   | Unknown   | Unknown         | No               | Yes               | No                | Craniotomy                   |
| 17  | 2012 | Patra   | 13      | Male   | Unknown   | Unknown         | No               | Yes               | No                | Bilateral craniotomies      |
| 18  | 2014 | Akodu   | 12      | Male   | HbSS      | Yes            | Unknown          | Unknown           | Unknown           | Supportive care             |
| 19  | 2014 | Ilhan   | 15      | Male   | HbSS      | Yes            | Yes              | Yes               | No                | Supportive care             |
| 20  | 2014 | Page    | 20      | Male   | HbSS      | Yes            | Yes              | Yes               | Yes               | Supportive care             |
| 21  | 2014 | Page    | 7       | Female | HbSS      | Unknown         | Yes              | Yes               | No                | Craniotomy                   |
| 22  | 2014 | Serarslan| 19      | Female | Unknown   | Unknown         | No               | Yes               | No                | Craniotomy                   |
| 23  | 2015 | Hettige | 7       | Female | HbSS      | Unknown         | Yes              | Yes               | No                | Craniotomy                   |
| 24  | 2015 | Oka     | 19      | Male   | HbSC      | Yes            | Unknown          | Yes               | Yes               | Supportive care             |
| 25  | 2016 | Boudreault| 17     | Male   | HbSS      | Yes            | Unknown          | No                | Yes               | Supportive care             |
| 26  | 2017 | Mishra  | 18      | Male   | Unknown   | Yes            | No               | Yes               | Yes               | Craniotomy                   |
| 27  | 2017 | Saul    | 18      | Male   | HbSS      | No             | Yes              | Yes               | No                | Unknown                     |
| 28  | 2018 | Gupta   | 16      | Male   | Unknown   | Yes            | Unknown          | No                | Yes               | Unknown                     |
| 29  | 2018 | Nelson  | 13      | Male   | HbSC      | Yes            | Yes              | Yes               | Yes               | Craniotomy                   |
| 30  | 2019 | Boukassa| 10      | Male   | HbSS      | No             | Unknown          | Yes               | No                | Craniotomy                   |
| 31  | 2019 | Boukassa| 16      | Male   | HbSS      | No             | No               | Yes               | No                | Craniotomy                   |
Spontaneous subgaleal hematoma is very rare in SCD and is often accompanied by epidural hematoma. Subgaleal hematoma usually resolves spontaneously with conservative management. However, it may necessitate surgical evacuation if it persists.  

The pathophysiology of spontaneous subgaleal hematoma in patients with SCD is not clearly understood, with different presumed explanations in the literature. The common explanation is the development of subgaleal hematoma as a result of cortical bone disruption secondary to periosteal elevation after bone infarction. This theory can also explain its common association with epidural hematoma. Spontaneous rupture of vessels adjacent to the infarcted bone may represent an alternative mechanism. Abnormal anatomy of skull bones secondary to chronic medullary hematopoiesis may cause rapid expansion of the bone marrow, particularly during acute episodes of anemia, precipitating the extravasation of blood into the subgaleal and epidural spaces. Additionally, impaired venous drainage could be the precipitating factor leading to excessive edema and, subsequently, hemorrhage.  

Skull bone infarction has been sporadically reported in SCD without accurate incidence. Bone infarction, usually seen in the long bones in SCD, results from a vaso-occlusive crisis. If the skull bones are involved, bone infarction commonly affects the orbital bones, mandible, and skull base. When it occurs, skull bone infarction is also managed by conservative management, as the case of long bone infarction.

Orbital involvement is scarce in such cases. It is caused by the accumulation of blood at the superior orbital ridges, which may disrupt the attachment of the arcus marginalis muscle, leading to blood accumulation within the orbital cavity, exophthalmos, decreased vision, and ophthalmoplegia.  

A literature review using the search terms “sickle cell disease,” “subgaleal” “hematoma,” “spontaneous,” “skull bone infarction,” and “headache,” in the title, abstract, and/or keywords of articles indexed in the Medline, Scopus, and Google Scholar databases revealed only 12 cases of spontaneous subgaleal hematoma in patients with SCD in the English literature, seven of which were associated with epidural hematoma (Table 1).

The current case had four main entities: headache, multiple subgaleal hematomas, skull bone infarction, and peri-orbital edema. There was no associated intracranial hemorrhage or exophthalmos, and the patient had mild, self-limited visual symptoms. We proposed that a vaso-occlusive crisis caused severe headache and skull bone infarction, which led to the subgaleal hematoma. The peri-orbital edema can be explained by the anterior extension of the subgaleal hematoma. Our decision to manage our patient conservatively was successful, with a gradual resolution of all symptoms.

Spontaneous subgaleal hematoma is a rare clinical entity, with sporadic cases reported as a complication of SCD. Most cases resolve with conservative measures. However, this condition could be associated with life-threatening intracranial hemorrhage. Skull bone infarction should be considered a possible cause of severe acute headache in patients with SCD.

**CONFLICT OF INTEREST**  
The authors declare that they have no competing interests.

**AUTHOR CONTRIBUTIONS**  
MF: writing the manuscript. AH: drafting the manuscript and reviewing the literature. AQ: editing the manuscript and reviewing the literature. MS: editing the manuscript and reviewing the literature.

**CONSENT**  
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the consent is available for review by the Editor-in-chief of this journal on request.

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