ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXIX | No. 1 March 2025

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Ahmed Mahmud,
Adeleke Nurudeen Abiola,
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DOI: 10.33962/roneuro-2025-010

Romanian Neurosurgery (2025) XXXIX (1): pp. 61-65
DOI: 10.33962/roneuro-2025-010
www.journals.lapub.co.uk/index.php/roneurosurgery

Spontaneous extradural hematoma in sickle cell anaemia-a stroke mimic. A report of two cases

Adegboye Olakunle Michael, Ahmed Mahmud, Adeleke Nurudeen Abiola, Enaworu Oghenevwoke, Nnara Stanley Onyeka, Wahab Kolawole Wasiu, Odebode Timothy Olugbenga

Division of Neurosurgery, Department of Surgery, University of Ilorin Teaching Hospital, Ilorin, Kwara State, NIGERIA

ABSTRACT

In the absence of trauma, extradural hematoma is rarely considered a cause of hemiplegia in a sickle cell disease patient, rather a cerebrovascular accident, as this occurs in about a quarter of sickle cell disease patients. We report two sickle cell anaemia patients who were initially diagnosed as cases of stroke having presented with hemiplegia/hemiparesis without a prior history of trauma to the head. Cranial computed tomographic scans however revealed extradural hematomas and both of them underwent surgical evacuation of the hematomas with subsequent neurologic recovery.

Keywords
CT scan, hemiplegia, sickle cell anaemia, spontaneous extradural hematoma, stroke

Corresponding author:
Adegboye Olakunle Michael

University of Ilorin Teaching Hospital, Ilorin, Kwara State, Nigeria

omadegboye@gmail.com

INTRODUCTION

Sickle cell disease (SCD) is a hematological condition with multi-systemic manifestations, including central nervous system. Neurological complications are among the most devastating manifestations of the disease and these include stroke, vascular malformations and cranial neuropathies.[1,2] Jeffrey reported that as many as 11% of patients with SCD will develop stroke by age 20 years while the incidence increases to 24% by age 45 years.[3]

Spontaneous extradural hematoma (EDH) is one of the underrecognized complications of sickle cell disease and rarely considered a cause of limb weakness.[4] Delay in making diagnosis of extradural hematoma could be dangerous as there could be expansion of the hematoma with subsequent coning and death. While ischemic and most times, haemorhagic stroke may not be amenable to surgical procedures but rather a long time of physical therapy which may leave the patients with significant residual deficit, surgical evacuation of extradural hematoma may have dramatic post-operative recovery once the offending compressive lesion is removed.

ISSN online 2344-4959
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First published
March 2025 by
London Academic Publishing
www.lapub.co.uk

Case Presentation

Case 1

An 18-year-old right-handed known sickle cell anemia (SCA) male patient diagnosed in childhood who presented to medical emergency room with a 3-day history of generalized headache, weakness of left upper and lower limbs and altered sensorium of two days. He had no history of seizure, vomiting and no preceding history of trauma to the head. He was being managed by the medical team as a case of right hemispheric stroke.

A cranial computerized tomography (CT) scan done however showed biconvex hyperdense right parietal mass lesion (measuring 78 mls) with a significant midline shift (12mm) and effacement of ipsilateral lateral ventricle, suggestive of an extradural hematoma with mass effect and overlying subgaleal hematoma (figure 1A-B). There was no overlying skull fracture. These radiological findings necessitated a consult to our unit (Neurosurgical team).

Upon our review, his Glasgow coma score was 9 and he had left spastic hemiplegia and a right sided parietal minimal diffuse scalp swelling. He was afebrile, anicteric but pale. His packed cell volume (PCV) was 24% (his stable PCV was 23-25%), platelet count and clotting profile were within normal limit.

He underwent emergency right sided parietal craniotomy and evacuation of the hematoma under general anaesthesia. Intraoperative findings were minimal subgaleal clotted blood and massive parietal extradural hematoma, compressing the underlying dura and brain (figure 2A-C). The bone flap appeared grossly normal with punctuate bleeding from the craniotomy edges.

Postoperatively, he made significant improvement, with full regain of consciousness within 2 days and gradual improvement of power in the left upper and lower limbs. On 12th day post - operative day, power had improved to 4 (from pre-operative power of 0) in the left upper and lower limbs. He was then discharged home for outpatient physiotherapy and follow up at the neurosurgery clinic. A clinic follow-up at three weeks post-operative revealed full power in all the limbs and he had no complaints. A repeat cranial CT scan at five weeks post-operative revealed no residual or recurrent extradural hematoma. (Figure 1C)

Case 2

A 10-year-old right-handed known SCA male patient (diagnosed at the age of three years), who presented to the emergency paediatric unit with history of abdominal pain, passage of dark coloured urine and weakness of right upper and lower limbs. He was pale with PCV of 18% (his stable PCV was not known). There was no antecedent history of trauma to the head or abdomen. He was diagnosed with hyperhaemolytic and vaso-occlusive crises and left hemispheric stroke. He could not do cranial CT scan due to financial constraints. He had exchange blood transfusion by the paediatric team and was discharged home after nine days of admission

He however represented after two weeks with history of headache and persistence of the weakness of the right upper and lower limbs. At this time, the mother was persuaded and he was able to do a cranial CT scan which showed a left parietal extradural hematoma and an overlying subgaleal hematoma without overlying skull fracture (figure 3A-C). Full blood count revealed adequate platelet count (206,000 cells/uL) and the clotting profile was within normal limit. A review by our team on being consulted revealed a fully conscious boy with normal pupils and right hemiparesis (power was grossly 3 in the right upper and lower limbs). He also had minimal left parietal scalp swelling.

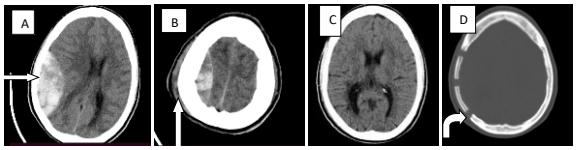

Figure 1. Pre-operative non-contrast cranial axial CT scan showing right parietal extradural hematoma with mass effect (A) and overlying subgaleal hematoma (B), fifth week post-operative cranial axial CT scan showing no residual or recurrent hematoma (C), bone window showing craniotomy burr hole sites (D).

He had surgical intervention under general anaesthesia. At surgery, we found minimal subgaleal hematoma, brittle dusky left parietal bone **(**figure 4A**)** and extradural clotted blood with lysed liquid portion (figure 4B). Due to the unhealthy nature of the bone, the parietal skull bone was excised until healthy bleeding edges (figure 4C) were reached and the hematomas were evacuated.

He made good recovery post-operatively with complete resolution of headache and gradual

resolution of the right hemiparesis. He was discharged home 10 days post-operatively with full power in all muscle groups of the limbs and residual left parietal skull defect of 8cm by 6cm size and counselled for later cranioplasty.

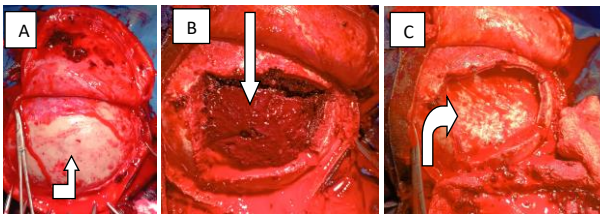

Figure 2. Intra-operative images showing healthy looking skull bone (A), massive extradural hematoma(B) and compressed dura/brain post evacuation of the hematoma (C).

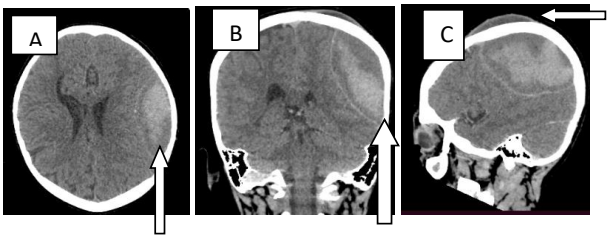

Figure 3. Non-contrast cranial axial (A) and coronal (B) CT scan showing right parietal extradural hematoma, the sagittal view (C) showed overlying subgaleal hematoma).

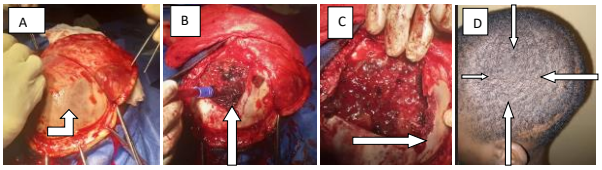

Figure 4. Intra-operative images showing dusky left parietal skull bone, suggestive of skull infarction (A), extradural hematoma(B) and bleeding bone edges, post craniectomy (C), three months post-operative clinical photograph showing left parietal skull defect with overlying scalp depression (D).

Discussion

Stroke, a significant cause of morbidity and mortality, is relatively common in sickle cell disease patients, affecting up to a quarter of these patients.[3,5] This could be in form of silent infarct, overt ischemic or hemorrhagic stroke[5–7] Overt stroke could manifest clinically with limb weakness, focal seizure, cranial nerve deficit among others.[3]

Intracranial EDH usually occurs following trauma and cases of spontaneous EDH are rare. The causes of spontaneous EDH could include coagulopathies, vascular malformations of the dura, dural/skull metastasis and infectious processes of the skull.[8] In the absence of the above aetiological factors and without history of trauma diagnosis of EDH becomes rarer. Spontaneous EDH is a rare complication in SCD patients, suspicion of its diagnosis may become submerged if the patients present with history suggestive of stroke.

Though the pathogenesis of spontaneous EDH in SCD patients has not been clearly defined, certain mechanisms have been proposed. These mechanisms include infarction of the skull bone, disruption of the diploic vein, periosteal elevation and cortical margin disruption and subsequent bleeding into either the extradural or subgaleal space or both.[9] Other suggested mechanism is that the skull could act as extramedullary hematopoietic site in SCD patients and during the period of hemolytic crisis, hyperproliferation and sudden expansion of the bone marrow in response to anemia could lead to expansion of the diploe, disruption of the diploic veins, disruption of inner and outer cortical skull margins and subsequent extravasation of blood.[10] Both of these mechanisms could explain co-existence of subgaleal and extradural hematomas in the same patient as found in the two patients presented. Although the pathogenesis of the spontaneous EDH could not be explained by any of these two mechanisms in the first patient presented, it could be argued that the finding of overlying necrosed bone flap at surgery could be explained by the infarction theory.

As a rare complication of SCD, the possibility of spontaneous EDH may be overlooked if the patients present with history suggestive of stroke. The clinical manifestation of spontaneous EDH varies. In a recent literature review in 2023 of 25 cases of spontaneous EDH by Lintz and Blum, the commonest presenting complaint was headache which occurred in 10 (40%), followed by VOC (32%), 12% of the patients presented with coma.[11] None of the 25 patients in the review presented with hemiparesis/hemiplegia, although Iversen *et al* reported a case of massive spontaneous EDH in a SCD patient who presented with headache, coma, dilated pupils and hemipareisis.[5] The index report is one of the few reports in which patients presented with limb weakness mimicking stroke.

Some pathomechanisms of spontaneous EDH in SCD patients suggest that there could be concomitant subgaleal hematoma in the same

patient. Page *et al* reported that concomitant subgaleal hematoma is found in 50% of SCD patients with spontaneous EDH.[12] The two patients in this case report had scalp hematomas. The presence of more sinister physical signs such as limb weakness may divert the attention of the attending physician from noticing a scalp swelling especially when it is subtle. We suggest that presence of scalp swelling should be specifically sought for in a SCD patient presenting with limb weakness.

The management of extradural hematoma depends on several factors, such as volume of the hematoma, presence or otherwise of mass effect, level of consciousness, presence or absence of neurologic deficit.[11] While some surgeons surgically manage these patients, some toe the pathway of conservative care. Chaurasiya *et al* opined that patients with large extradural hematoma with mass effect and significant neurologic deficit warrant surgical evacuation.[13] Both patients we presented in this report underwent surgical evacuation due to the large volume of hematoma with mass effect, presence of neurologic deficit and altered consciousness in the first patient and neurologic deficit in the second patient. Lint and Blum reported that 13 of 25 patients they reviewed were managed surgically while the remaining 12 patients had conservative care.[11]

The outcomes of patients with extradural hematoma varies, mortality ranges between 20% and 33% in the literature.[11,12,14] Prognosis largely depends on early diagnosis and promptness of management institution. Therefore, it is important to have a high index of suspicion and low threshold for requesting cranial CT scan in SCD patients who present with features suggestive of spontaneous EDH in order to avoid preventable mortality and/morbidity. Page *et al* reported a large EDH in a SCD which was only diagnosed at autopsy with features of brain herniation and infarction of the skull overlying the hematoma.[12]

Conclusion

The presence of limb weakness in sickle cell disease patient may not always be due to stroke. Unfortunately, little attention may be given to the possibility of surgically treatable extradural hematoma, especially in the absence of history of head trauma. Failure of early diagnosis of extradural hematoma and prompt surgical evacuation may lead to mortality. A search for a possible spontaneous extradural hematoma through cranial computed tomography scan should be commenced in earnest as surgical evacuation of such could be life-saving and produce good neurologic recovery.

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