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Original Article

Neurosurgical crisis in sickle cell disease: Case reports of haemorrhagic stroke in paediatric patients.

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Abstract
A collection of hemoglobinopathies known as sickle cell disease (SCD) ranges in severity; the most severe type, homozygous sickle cell anemia, is more frequently linked to neurological issues. These are ascribed to micro-obstruction and vaso-occlusion in the circulation of the central. nervous system. Across different series, the incidence of neurologic problems in SCD ranges from 6% to 30%. Silent cerebral infarction (SCI), ischemic stroke, transient ischemic episodes (TIAs), headaches, seizures, and neurocognitive impairment are among the conditions that are frequently documented in the literature.
One of the most debilitating symptoms of sickle cell disease (SCD) in children is cerebral vascular problems. Haemorrhagic stroke is a neurosurgical emergency with a high morbidity and fatality rate, whereas ischemic stroke is recorded more often. Subarachnoid hemorrhage (SAH), hemorrhagic stroke, and extradural and subdural hematomas are examples of hemorrhagic consequences that are rarely considered, particularly when there is no trauma.
This article delineates three uncommon spontaneous hemorrhagic manifestations of sickle cell anaemia: a case of subdural hematoma (SDH), a case of extradural hematoma (EDH) concomitant with SDH and subarachnoid haemorrhage, and a case of intracranial haemorrhage. These cases highlight the importance of early neuroimaging, aggressive supportive care, and multidisciplinary management in preventing adverse outcomes.

Keywords: *Anemia, hematoma, neurological, sickling, stroke.*
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Introduction
The hereditary disorder known as sickle cell disease (SCD) is defined by the presence of sickled red blood cells that clog the microcirculation and result in tissue hypoxia. Chronic hemolysis, vaso-occlusion, and progressive vasculopathy are the hallmarks of sickle cell disease, an inherited hemoglobinopathy. (1). 5 to 10 % of children experience neurological issues, with stroke being one of the most severe aftereffects. Neurological problems cause a substantial portion of SCD patients' morbidity and mortality. (2).
While ischemic stroke is more common in pediatric SCD, hemorrhagic stroke, which includes subdural hemorrhage (SDH) and subarachnoid hemorrhage (SAH), is becoming more and more common, especially in teenagers. (3). Endothelial dysfunction, chronic anemia, hypercoagulability, and the creation of frail collateral vessels that are prone to rupture are all part of the pathogenesis. Survival depends on early detection and neurosurgical intervention. (4). This article describes three uncommon SCD patients with hemorrhagic central nervous system (CNS) symptoms.
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Case 1
A 15-year-old male patient, who was a known case of sickle cell disease, had a frontal headache that came on suddenly, followed by strange body motions and a day-long loss of consciousness. He had icterus and pallor when he was admitted. With a heart rate of 130 beats per minute, a respiratory rate of 20 beats per minute, a blood pressure of 138/78 mmHg, and an oxygen saturation of 93% on room air, were his initial vitals. A Glasgow Coma Scale (GCS) score of E1V1M3, which indicates severe impairment of consciousness, was found during a central nervous system evaluation. Anisocoria was observed by pupillary examination, and bilaterally brisk deep tendon reflexes and bilaterally upgoing plantar reflexes indicated involvement of higher motor neurons. The results of the gastrointestinal, respiratory, and cardiovascular exams were ordinary.
A cerebrovascular accident was suspected due to the severe neurological deterioration and the history of sickle cell illness. An acute subdural hemorrhage with signs of active bleeding was discovered during an urgent non-contrast CT (NCCT) scan of the brain. Laboratory tests revealed a normal platelet count (369 lakhs/cu mm), leukocytosis (WBC 18,200/cu mm) with neutrophilic

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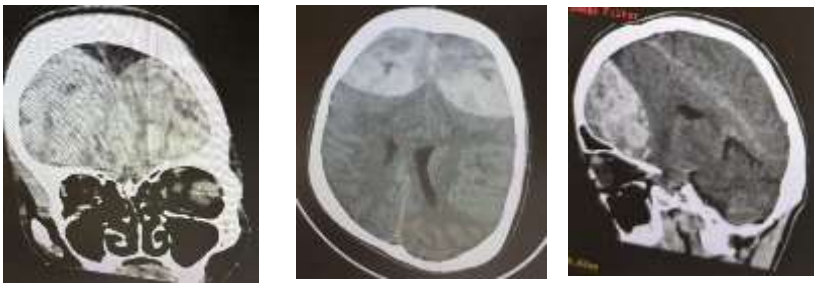
predominance, and severe anemia with hemoglobin of 4.0 g/dL. Liver function tests revealed high transaminases (ALT 122 U/L, AST 55 U/L), elevated alkaline phosphatase (276 U/L), and primarily indirect hyperbilirubinemia (total bilirubin 3.7 mg/dL, indirect 2.8 mg/dL), all of which were indicative of hemolysis and hepatic stress. Renal function was maintained, and coagulation measures were within normal ranges (INR 0.99). With severe anemia and continuous hemolysis, the clinical picture pointed to a serious cerebral hemorrhagic episode in sickle cell disease.

The patient had a known history of sickle cell disease and was on intermittent supportive treatment, including blood transfusions, with poor compliance to regular follow-up. Following diagnosis, the patient was managed in the intensive care unit with aggressive supportive care, including packed red blood cell transfusion, osmotherapy, anticonvulsants, and neurosurgical consultation. Despite intensive management, the patient showed minimal neurological improvement due to the severity of the hemorrhage. No procedure-related adverse events were noted; however, the overall prognosis remained guarded due to severe neurological insult.

Case 2

A 15-year-old male patient, who was a known case of sickle cell disease, had one episode of generalized tonic-clonic seizures and four hours of impaired sensorium. He was admitted in a semiconscious state with noticeable icterus and pallor. Patient was tachycardic (pulse 140/min), tachypneic (respiratory rate 30/min), with raised blood pressure (150/88 mmHg), and low oxygen saturation (90 percent on room air). A Glasgow Coma Scale (GCS) score of E2V3M5, which indicates substantial impairment of consciousness, was obtained after a neurological examination. Anisocoria was discovered by pupillary examination. Bilaterally brisk deep tendon reflexes and bilaterally upgoing plantar reflexes were indicative of elevated intracranial pressure and involvement of higher motor neurons. The results of other systematic evaluations were not noteworthy.

An immediate non-contrast CT (NCCT) scan of the brain was conducted due to the severe neurological impairment in the background of sickle cell disease. With a maximal thickness of 10.7 mm, imaging showed an acute-on-chronic subdural hemorrhage along the right frontoparietal convexity, which resulted in compression of the right lateral ventricle and effacement of the underlying sulci. There was a noticeable midline displacement to the left of around 9 mm. Along with diffuse cerebral edema marked by widespread effacement of sulci and gyri, there was also a thin subdural hemorrhage (2 mm) along the falx cerebri. These results showed elevated intracranial pressure and a potentially fatal intracranial event with mass effect.



Laboratory tests showed a normal platelet count, leukocytosis (WBC 12,300/cu mm), and severe anemia (hemoglobin 5.9 g/dL). Indicative of continuous hemolysis, liver function tests revealed increased AST (107 U/L) and significant indirect hyperbilirubinemia (total bilirubin 5.6 mg/dL, indirect 4.3 mg/dL). Renal function was maintained, and coagulation measures were within normal ranges (INR 0.99). Low ionized calcium and mild hyponatremia were also seen. In a patient with sickle cell disease and severe hemolytic anemia, the entire clinical picture indicated acute neurological decompensation brought on by acute-on-chronic subdural hemorrhage.

The patient had a prior diagnosis of sickle cell disease and had received occasional transfusion therapy. After diagnosis, the patient was managed with supportive intensive care, including blood transfusion, antiedema measures, seizure control, and close neurological monitoring. Neurosurgical intervention was considered due to significant mass effect. The patient showed partial clinical stabilization; however, neurological deficits

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persisted. No major adverse events related to treatment were observed during the hospital stay.

Case 3

A 10-year-old female patient, who was a known case of sickle cell disease, had several partial seizure episodes on the second postoperative day after splenectomy. She was afebrile and alert at admission. With a blood pressure of 100/78 mmHg, oxygen saturation of 98% on room air, a pulse rate of 90 beats per minute, and a respiratory rate of 24 beats per minute, her vital signs were steady. A Glasgow Coma Scale (GCS) score of E4V5M6, which indicates full consciousness, was obtained during neurological testing. Patient's pupils reacted to light and were bilaterally equal. Plantar reflexes were upgoing on both sides, and deep tendon reflexes were brisk bilaterally, indicating involvement of higher motor neurons. There were no noteworthy findings from the examination of the respiratory, gastrointestinal, or cardiovascular systems.

A cerebrovascular incident was suspected in a postoperative sickle cell patient due to the sudden development of convulsions. Subarachnoid hemorrhage along the bilateral Sylvian fissures and cortical infarctions involving the parafalcine regions of the bilateral basofrontal and high frontal lobes were seen on the contrast-enhanced MRI brain. These results pointed to a thrombo-occlusive condition, which is a known consequence of sickle cell disease. There was also moderate hydrocephalus. In accordance with the clinical seizure activity, electroencephalography (EEG) showed an epileptogenic focus in the right hemisphere

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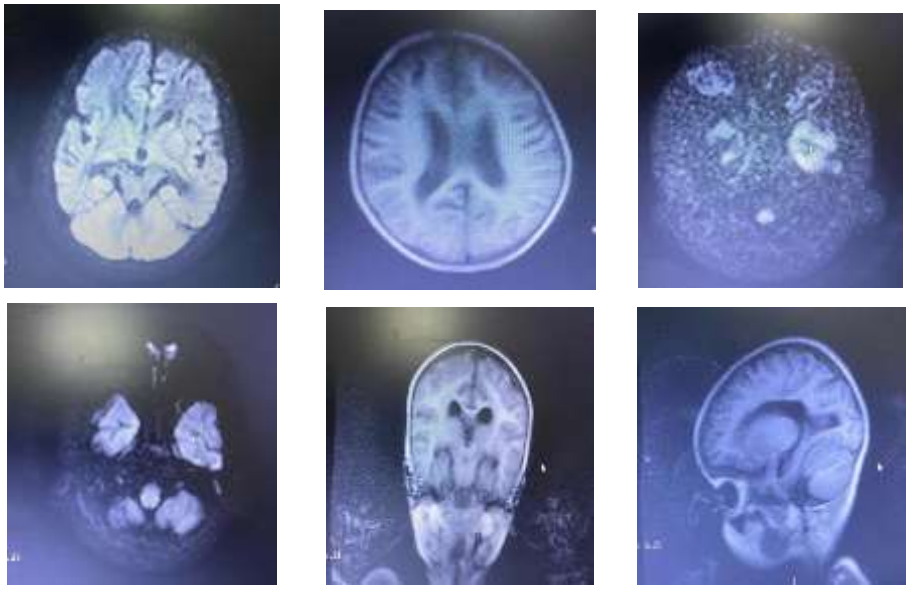

Laboratory tests revealed a normal platelet count, leukocytosis (WBC 12,300/cu mm), and hemoglobin of 8.19 g/dL. According to liver function tests, there was continuous hemolysis, as evidenced by high AST levels and indirect hyperbilirubinemia (total bilirubin 5.6 mg/dL). Renal function was maintained, and coagulation markers were within normal ranges. Low ionized calcium and mild hyponatremia were also noted. Overall, the clinical picture showed focal seizure activity and a postoperative cerebrovascular complication with subarachnoid hemorrhage and cortical infarction due to sickle cell-related thrombo-occlusive pathology.

The patient was a known case of sickle cell disease and had recently undergone splenectomy. Post-diagnosis, management included anticonvulsant therapy, supportive care, and correction of metabolic abnormalities. The patient responded well to treatment, with control of seizures and stable neurological status on follow-up. No

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significant adverse events were reported. The postoperative state likely contributed to increased thrombotic risk, leading to the cerebrovascular event.

Discussion

SCD is a qualitative hereditary hemoglobinopathy characterized by the presence of hemoglobin S (sickle hemoglobin) (5). This arises from the substitution of the amino acid glutamine by valine in the sixth position of the beta-globin chain. Its incidence is high among the people of African, Arabian, and Indian origin. SCD in India is prevalent in the Western, Central, and Eastern regions and in the pockets of the South. In the eastern regions, it is common in Odisha, Jharkhand, and Bengal (6). SCD is characterized by the hemoglobin S polymerization, erythrocyte stiffening, and subsequent vaso-occlusion (7). These changes lead to obstruction to microcirculation, tissue ischemia, and infarction in the various organ systems, including the cerebrovascular system. All three patients in our series had the homozygous form (HbSS), thus representing a severe form of the disease. (8).

Patients with SCD have a higher incidence of cerebrovascular events. The incidence of serious neurologic complications in SCD ranges from 6% to 30% in various series. By decreasing the order of frequency, neurologic manifestations of SCD include silent cerebral infarcts (39% by the age of 18), acute and chronic headache (36% in children), neurocognitive impairment (25%), seizures (7%–10%), ischemic stroke (1% in children with effective screening and prophylaxis, but nearly 11% in children without screening), and hemorrhagic stroke (3% in children and 10% in adults) (9), (10). Approximately 70%–80% of all strokes are ischemic, and 20%–30% are hemorrhagic in nature (11).

There is still much to learn about the pathophysiology of sickle cell anemia's neurological consequences. Large arterial occlusive disease has been identified by CT and magnetic resonance angiography (MRA) in the proximal segments of the internal carotid arteries' main branches and terminal intracranial parts of the internal carotid arteries, but infrequently in the vertebrobasilar or extracranial carotid systems. (12). The development of a mass of tiny, friable new blood vessels in reaction to the severe stenosis or occlusion of the major intracranial vessels is known as large-vessel cerebral vasculopathy. (13).

In addition to highlighting the increased risk of both hemorrhagic and ischemic cerebrovascular events in this population, the three cases discussed also demonstrate the severe and varied neurological consequences that can arise in pediatric sickle cell disease (SCD) patients. Acute neurological deterioration was evident in all three cases, highlighting the unpredictable and potentially fatal nature of central nervous system involvement in sickle cell disease.

In the first two cases, teenage boys with known sickle cell disease (SCD) showed up with seizures and a changed sensorium. Neuroimaging revealed subdural hemorrhages. Significant anemia (hemoglobin 4.0 g/dL) and continuous hemolysis, as evidenced by indirect hyperbilirubinemia, were linked to acute subdural hemorrhage with active bleeding in the first instance. Imaging in the second patient showed diffuse cerebral edema with acute-on-chronic subdural bleeding with a notable mass impact and midline shift. Bilateral extensor plantar reflexes and anisocoria were seen in both patients, which suggested elevated intracranial pressure and serious neurological impairment. Notably, both individuals' coagulation values fell within normal ranges, indicating that endothelial dysfunction, sickle cell-associated vasculopathy, and the development of weak collateral vessels were more likely to be the cause of the hemorrhagic events than primary coagulopathy. It's possible that severe anemia added to the vascular instability and brain hypoxia.

The third instance demonstrates a distinct yet no less severe form of cerebrovascular disease associated with SCD. On the second postoperative day after the splenectomy, this 10-year-old girl experienced several partial seizures. A thrombo-occlusive process was suggested by the MRI data, which showed mild hydrocephalus, bilateral cortical infarctions in the frontal areas, and subarachnoid hemorrhage. The complicated pathophysiology of SCD, which includes the simultaneous occurrence of vaso-occlusion, endothelial damage, hypercoagulability, and reperfusion injury, is reflected in the coexistence of bleeding and infarction. Thrombotic risk may be considerably elevated in the postoperative state. Clinical seizure activity was correlated with an epileptogenic focus confirmed by EEG. When taken as a whole, these instances highlight how, even in the absence of coagulation problems, children and adolescents with SCD are nonetheless at high risk for both ischemic and hemorrhagic stroke. Improving results requires continuous neurological monitoring, interdisciplinary management, aggressive anemia repair, and early neuroimaging. These findings support the necessity of careful screening and prophylactic measures for high-risk SCD patients. (8).

Key Takeaways:

These cases highlight the diverse and severe neurological complications of sickle cell disease in pediatric patients. Early neuroimaging, prompt recognition of neurological

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deterioration, and aggressive multidisciplinary management are critical in improving outcomes. Even in the absence of trauma or coagulation abnormalities, clinicians should maintain a high index of suspicion for hemorrhagic stroke in SCD patients.

Conclusion

In children with sickle cell disease, hemorrhagic stroke is a potentially fatal neurosurgical emergency. Improving outcomes requires aggressive hematological correction, timely imaging, coordinated neurosurgical care, and early detection of neurological deterioration. These incidents demonstrate the uncommon neurological hemorrhagic side effects of sickle cell disease. While EDH is a rare SCD consequence, recurrent EDH is quite infrequent. Small

Neurological impairments may not be a symptom of EDH, and ischemia may not necessarily be linked to the early onset of stroke in SCD patients. A strong index of suspicion is necessary since early diagnosis and treatment lower morbidity and mortality.

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Data Availability:

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Ethical Considerations:

Ethical approval was obtained from the Institutional Ethics Committee. Informed consent was obtained from the parents/guardians of all patients included in this report. Patient confidentiality has been maintained.

References:

1. Parikh T, Goti A, Yashi K, Prasad N, Ravikumar G, Dankhara N, et al. Pediatric Sickle Cell Disease and Stroke : A Literature Review. Cureus. 2023;15(1):1-7. https://doi.org/10.7759/cureus.34003
2. Keyrouz SG, Diringer MN. Clinical review : Prevention and therapy of vasospasm in subarachnoid hemorrhage. Crit Care Vol. 2007;11(4):1-10. https://doi.org/10.1186/cc5958
3. Linguet SL, Verlhac S, Missud F, Holvoet-vermaut L, Brousse V, Ithier G, et al. Stroke without cerebral arteriopathy in sickle cell disease children : causes and treatment. 2024;(October):3346-56. https://doi.org/10.3324/haematol.2023.283773
4. Debaun MR, Kirkham FJ. Central nervous system complications and management in sickle cell disease. Blood [Internet]. 2016;127(7):829-38. https://doi.org/10.1182/blood-2015-09-618579
5. Dominique N'dri Oka, André Tokpa, Alpha Bah LD. Spontaneous Intracranial Extradural Hematoma in Sickle Cell Disease. J Neurol Surg Reports. 2015;76(R1):97-9. https://doi.org/10.1055/s-0035-1544953
6. Sangita Kamath. NJ and BSR. ACUTE CHEST SYNDROME IN SICKLE CELL DISEASE - THE PAIN MALADY (1 YEAR TMH) EXPERIENCE. Int J Recent Sci Res. 2016;7(2):8940-6.
7. Al-jafar HA, Alroughani R, Abdullah TA, Al-qallaf F. Neurological Complications in Sickle Cell Disease. Int J Clin Exp Neurol. 2016;4(1):9-18.
8. Azhar MJ. Extradural Hemorrhage: A Rare Complication and Manifestation of Stroke in Sickle Cell Disease. Oman Med Journa. 2010;25(4):1-4. https://doi.org/10.5001/omj.2010.97
9. Noubiap JJ, Mengnjo MK, Nicastro N, Kamtchum- J. Neurologic complications of sickle cell disease in Africa: A systematic review and meta-analysis. Neurology. 2017;89(October):1516-24. https://doi.org/10.1212/WNL.0000000000004537
10. Alberts AH, Lucke-wold B, Alberts AH, Lucke-wold B. Stroke in sickle cell patients, epidemiology, pathophysiology, systemic and surgical treatment options and prevention strategies Stroke in sickle cell patients, epidemiology, pathophysiology, systemic and surgical treatment options and prevention st. Clin Transl Disc. 2024;4(April):1-12. https://doi.org/10.1002/ctd2.303
11. WOOD DH. Current Concepts of Cerebrovascular Disease - Stroke Cerebrovascular Complications of Sickle Cell Anemia. Stroke. 1978;9(1):73-5. https://doi.org/10.1161/01.STR.9.1.73
12. Mustafa M, Amawi M, Altoonisi MM, Soliman W, Kamal M. Beyond Ischemia : The Rare Occurrence of Hemorrhagic Strokes in Pediatric Sickle Cell Anemia Case Presentation. Cureus.

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Original Article

2024;16(10):1-14. https://doi.org/10.7759/cureus.70871
13. Sangita D Kamath MGP. A case series of hemorrhagic neurological complications of sickle cell disease : Multiple faces of an underestimated problem ! Asian J Transfus Sci. 2021;15:241-6. https://doi.org/10.4103/ajts.AJTS_101_18

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