Case Report

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A case series of hemorrhagic neurological complications of sickle cell disease: Multiple faces of an underestimated problem!

Sangita D Kamath, Manish Ganesh Pai[1]

Abstract:

Sickle cell disease (SCD) is a group of hemoglobinopathies that vary in severity, the most severe form, homozygous sickle cell anemia, is more commonly associated with neurologic complications. These are attributed to the vaso-occlusion and micro-obstruction in the circulation of the central nervous system. The incidence of various neurologic complications in SCD ranges from 6% to 30% in various series. The commonly reported in literature include silent cerebral infarction (SCI), ischemic stroke, transient ischemic attacks (TIAs), headaches, seizures and neurocognitive impairment. However, hemorrhagic complications like subarachnoid hemorrhage (SAH), hemorrhagic stroke, extradural and subdural hematomas, especially in absence of trauma are rarely thought of. We report three uncommon spontaneous hemorrhagic manifestations of sickle cell anemia – one case of parenchymal (intracerebral) bleed who presented with acute onset of parkinsonism and two cases of extradural hematoma (EDH) of which one patient had recurrent EDH at the same site which is hitherto not reported in the literature.

Keywords:

Anemia, hematoma, neurological, sickling, stroke

Introduction

Sickle cell disease (SCD) is an autosomal recessive hemoglobinopathy characterized by the presence of sickled red blood cells causing obstruction to the microcirculation and tissue hypoxia.[1] Neurological complications in patients with SCD contribute significantly to the morbidity and mortality of the disease. Although the incidence of acute ischemic stroke and chronic cerebral ischemia is higher in SCD,[1] hemorrhagic complications also occur but are rarely thought of. We present a series of three rare cases of SCD, who had hemorrhagic central nervous system (CNS) manifestations.

Departments of Internal Medicine and [1]Neurosurgery, Tata Main Hospital, Tata Steel, Jamshedpur, Jharkhand, India

Address for correspondence:
Dr. Sangita D Kamath, D-25, Vijaya's Heritage, Anil Sur Path, Uliyan, Kadma, Jamshedpur - 831 005, Jharkhand, India.
E-mail: dr.sdkamath@gmail.com

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Case Reports

Case 1

A 34-year-old female unmarried, was admitted to the Tata Main Hospital with a history of severe headache, giddiness, and generalized weakness of 2 days duration. There was no history of vomiting, fever, double vision, or convulsions. She denied the history of head trauma. She was a diagnosed case of SCD and had undergone cholecystectomy at the age of 7 years for cholelithiasis, splenectomy at 10 years of age, and total hip replacement for chronic arthritis of the right hip joint secondary to avascular necrosis of the femur neck in 2013. She had last received blood transfusion 2 years back for anemia. She did not have the history of any other significant medical ailment in the past. She did not

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consume alcohol and tobacco in any form. She was the second-affected sibling in the family.

On admission, she was lean, had mild pallor and icterus, no pedal edema, and cyanosis. She was afebrile, with a pulse rate of 106/min, blood pressure 110/70 mm Hg, and respiratory rate of 18/min, regular and abdominothoracic type. Examination of the CNS revealed awake person, apathetic, Glasgow Coma scale (GCS) E4 (vacant), M6V2–12/15, pupils were normal size and reacting to the light, and preferential left-sided gaze and right hemiparesis with right up-going plantar. Neck was soft. Examination of the other systems such as cardiovascular, respiratory, and gastrointestinal was within the normal limits. Considering the possibility of a cerebrovascular accident (CVA) and an urgent noncontrast computed tomography (NCCT) of the brain was done which showed acute bleed in the right parietal lobe with surrounding edema and mass effect. Basal cisterns were open. There was no midline shift [Figure 1a and b].

Her hemoglobin on admission was 6.9 g/dL, total white blood cell (WBC) count 10,200 cu mm with 35% neutrophils, 53% lymphocytes, 8% monocytes, mean corpuscular volume (MCV) 74.9fL, and platelet count 1.5 lakhs/cu mm. Her liver function tests (LFT) revealed total bilirubin 3.96 mg/dL, direct fraction 0.98 mg/dL, indirect fraction 2.98 mg/L, alanine transaminase (ALT) 17.9 U/L, aspartate transaminase (AST) 52.6 U/L, alkaline phosphatase (ALP) 82.6 U/L, total serum proteins 7.01 g/dL, serum albumin 4.01 g/dL and globulin 3.0 g/dl, activated partial thromboplastin time (APTT) 12.4 s, control 14 s, prothrombin time (PT) 19 s, control 11 s, and PT international normalized ratio (INR) 0.99. Renal function tests showed blood urea 17.2 mg/dL and serum creatinine 0.56 mg/dL. Serum iron was 70.5 mcg/dL, and serum ferritin was 1444.8 ng/mL. Her hemoglobin electrophoresis showed fetal hemoglobin (HbF) 11%, adult hemoglobin$_2$ (HbA$_2$) 3.7%, sickle hemoglobin (HbS) 85.3%, and no HbA. Her chest radiograph was normal.

She was treated conservatively with 20% mannitol 100 ml for 3 days intravenously, three units of compatible packed red blood cells transfusion, pantoprazole 40 mg OD, Ryle's tube feed of 1.8 L/day, and other supportive measures. She developed the features of acute parkinsonism in the form of mask such as face, hypertonia, and cogwheel rigidity on day 4 of admission. Her contrast-enhanced CT of the brain done after 10 days showed resolving hemorrhage with surrounding edema with effacement of the right lateral ventricle and midline shift [Figure 2a and b]. She did not worsen neurologically and was given injection dexamethasone 8 mg three times a day intravenously for 5 days and subsequently discharged with advice to continue the physiotherapy and follow-up in outpatient department.

Case 2

A 18-year-old male, known to have sickle cell anemia (SCA) was admitted to our hospital with a history of chest, lower back pain for 3 days, and frontal headache of 1 day duration. He had a previous history of the several vaso-occlusive crises requiring simple analgesia. He was not on hydroxyurea. He was born at full term after uneventful delivery and was the first-affected sibling. At the time of admission, he was afebrile, had significant pallor, icterus, tachycardia, and normal blood pressure. Examination of the CNS revealed restless, irritable, and confused patient with GCS of 9/15 (E3M4V2). There was mild right hemiparesis with upgoing plantar. Rest of the systemic examination did not reveal any abnormality. Laboratory values on the admission showed Hb 6.7 g/dL, hematocrit 20.1%, total WBC count 12,200 cu mm with 65% neutrophils, 33% lymphocytes, 2% monocytes, MCV 78.9fL, and platelet count 75,000/cu mm. LFT revealed total bilirubin 8.96 mg/dL, direct fraction 0.98 mg/dL, indirect fraction 7.98 mg/L, ALT 28.2 U/L, AST 42.6 U/L, ALP 162.6 U/L, PT 14 s, control 12 s, and PT (INR) 1.18, and normal APTT. His hemoglobin electrophoresis showed HbF 14%, HbA$_2$ 3.7%, HbS 82.8%, and no HbA. His

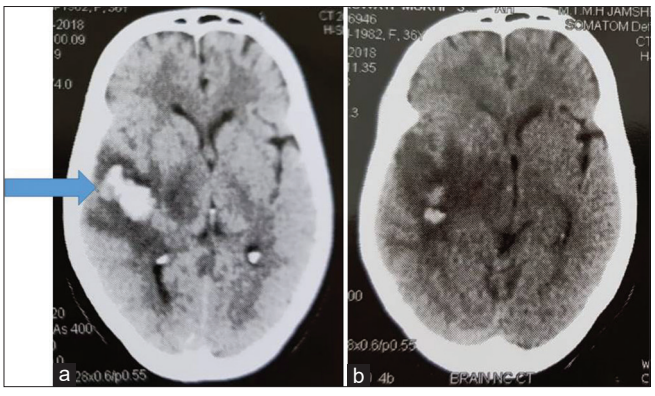

Figure 1: (a and b) Non contrast computed tomography of the brain showing acute bleed in the right parietal lobe with surrounding edema and mass effect (arrow)

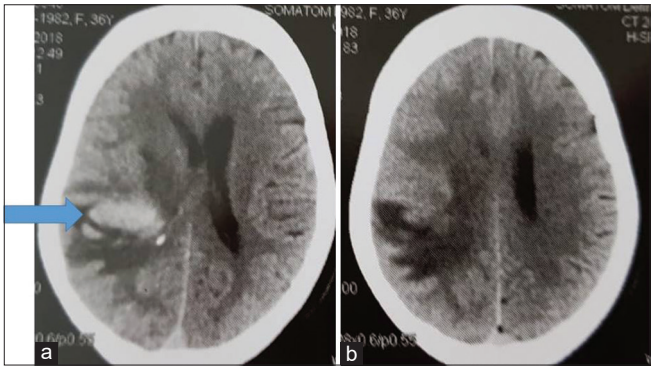

Figure 2: (a and b) Noncontrast computed tomography of the brain showing resolving bleed in the right parietal lobe with surrounding edema extending into basal ganglia and mass effect (arrow)

serum lactate dehydrogenase (LDH) was 1025 U/L. His hemoglobin 3 months before this admission was 7.4 g/dL. His headache intensified the next day requiring escalation of pain analgesia to opiates. In view of the focal neurological deficit and headache, he underwent an urgent NCCT brain which showed an extradural hematoma (EDH) overlying the left frontal-parietal lobe with mass effect, and midline shift to right [Figure 3a and b]. Neurosurgical opinion was sought and he underwent the emergency left-sided craniotomy for draining of the hematoma. Subsequently, he was treated with analgesia, packed red cell transfusions, antibiotics, anti-epileptic (levetiracetam), and other supportive measures. He made a remarkable recovery with rehabilitation and was discharged after 20 days.

This patient was readmitted 1 year later with body aches, severe headache of 2 days duration, and one episode of generalized convulsion. On admission, he had pallor, icterus, and was restless with GCS of 7/15. Pupils were asymmetric; the right pupil was dilated (4 mm), sluggishly reactive to light and the left pupil (2 mm) showed normal reaction. There was right hemiparesis with positive Babinski's sign. Laboratory values showed Hb 6.2 g/dL, hematocrit 18.6%, total WBC count 13,200 cu mm with 75% neutrophils, and 23% lymphocytes. LFT revealed total bilirubin 6.9 mg/dL, direct fraction 0.9 mg/dL, indirect fraction 6 mg/L, and liver enzymes were normal. His NCCT brain showed a large left-frontal EDH with midline shift to right. He underwent an emergency left-sided craniotomy. The extradural hematoma was evacuated, and dural arterial bleeding was controlled. He was managed postoperatively in the intensive care unit with three units of packed cell transfusions, antibiotics, and mechanical ventilation. Two days postsurgery; he had a sudden cardiac arrest and could not be revived. There was no time to repeat CT scan of the brain. The cause of death was probably, reaccumulation of hematoma with herniation of the brain.

Case 3

A 26-year-old male, known case of SCA was admitted with a history of severe headache involving the whole head with pain in the small joints of the body with low-backache of 2 days duration. There was no fever, vomiting, altered sensorium, visual disturbance, or epileptic seizure. No history of head trauma was found. He had one episode of acute chest syndrome 2 years back. Examination of the CNS showed normal orientation to time, place, person, and no focal neurological deficit. Rest of the systemic examination was normal except for mild hepatomegaly. His blood tests revealed Hb 7.7 g/dl, hematocrit 23.1%, total WBC count 13,200 cu mm with 75% neutrophils, 23% lymphocytes, 2% monocytes, MCV 76.9 fL, and platelets 1.25 lakh/cu mm. LFT revealed total bilirubin 8.32 mg/dL, direct fraction 1.12 mg/dl, indirect fraction 6.8 mg/L, ALT 38.2 U/L, AST 48.3 U/L, ALP 192.6 U/L, PT 12.1 s, control 12 s, and PT (INR) 1.01. Other coagulation parameters such as APTT and serum fibrinogen were normal. His hemoglobin electrophoresis showed HbS level of 76.8% and no HbA. His serum LDH was 925 U/L. NCCT of the brain done showed an EDH overlying the left-parietal lobe [Figure 4a and b]. He was managed conservatively (in view of small EDH) with analgesia, blood transfusion, antibiotics, and other supportive measures. A repeat CT scan after 1 month showed near complete resolution of the hematoma.

Discussion

SCD is a qualitative hereditary hemoglobinopathy characterized by the presence of hemoglobin S (sickle hemoglobin).[2] This arises from the substitution of the amino acid glutamine by the valine in the sixth position of 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 South. In the eastern regions, it is common in Odisha, Jharkhand, and Bengal.[3] SCD is characterized by the hemoglobin S polymerization, erythrocyte stiffening, and subsequent vaso-occlusion.[1] These changes lead to obstruction to microcirculation, tissue ischemia, and infarction in the various organ systems including the cerebrovascular system. All the three patients in our series had homozygous form (HbSS), thus representing severe form of the disease.

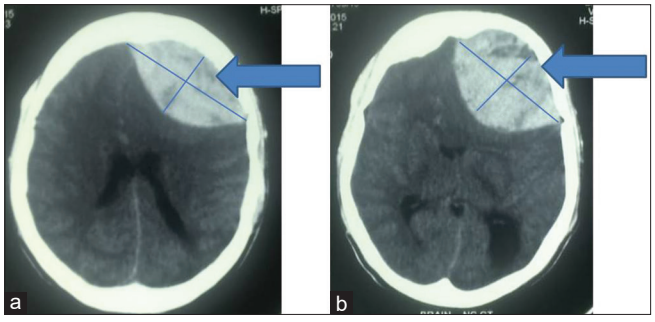

Figure 3: (a and b) Axial views of non-contrast computed tomography of the brain showing left extradural hematoma (arrow)

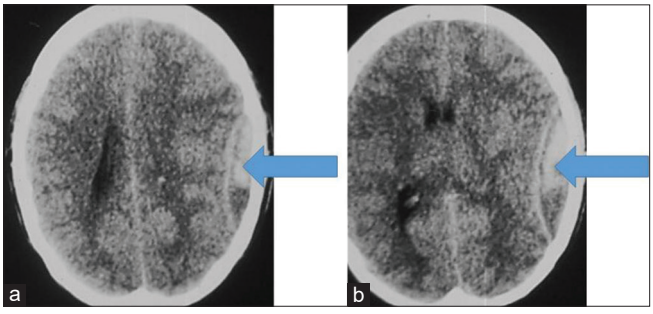

Figure 4: (a and b) Axial views of non-contrast computed tomography of the brain showing left parietal extradural hematoma (arrow)

Patients of SCD have the 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).[1,4-6] Approximately, 70%–80% of all strokes are ischemic, and 20%–30% are hemorrhagic in nature.[1,7]

Primary hemorrhagic stroke has a reported mortality of 24%–65%.[8,9] Infarctive strokes are relatively more common in children than in adults while the reverse is true for hemorrhagic stroke.[8] As per the cooperative study of SCD (CSSCD) report, 5 (9.6%) of 52 first strokes in SCD-SS patients <20 years were hemorrhagic, whereas 14 (52%) of 27 first stroke in those over 20 years of age were hemorrhagic.[8,9] Hemorrhagic complications recognized in SCD include intracerebral hemorrhage, intraventricular bleed, subarachnoid hemorrhage (SAH), and subdural and epidural hematoma. In a study of neurological complications among 325 patients with SCD followed at the University of Illinois between 1975 and 1989, 11 cases of SAH were identified, of which 10 had aneurysms.[6,10]

Our first patient had hemorrhagic acute CVA. CVAs are a catastrophic complication of SCD and a leading cause of death in both children and adults. CVAs occur most commonly in the HbSS genotype with an incidence of 0.61/100 patient-years.[11] The CSSCD determined prevalence and incidence rates of stroke based on data from 4,000 patients followed for up to 10 years from 1978 to 1988. Overall prevalence of stroke in all forms of SCD was 4%; and 5% in those with homozygous SCD (SCD-SS).[12] CSSCD report revealed that the incidence of hemorrhagic stroke (0.44/100-patient years) was the highest in patients aged at 20–29 years.[13] It was associated with a low-steady-state hemoglobin and a high-WBC count. Subarachnoid hemorrhages were less common while extradural bleeds were rare.[14] The other identified risk factors, for the development of hemorrhagic stroke in children, include hypertension, recent blood transfusion,[11] and use of corticosteroids or nonsteroidal anti-inflammatory drugs,[13] whereas in adults include not only hypertension but also diabetes mellitus, hyperlipidemia, atrial fibrillation, and renal disease.[6,11] Hemorrhagic strokes in SCD could also be associated with advanced sickle cell hepatopathy, where the coagulation profile is significantly impaired.[15] However, none of these factors was observed in our patient.

The pathogenesis of neurologic complications in sickle cell anemia is not completely understood. CT and magnetic resonance angiography (MRA) have revealed the prominent large arterial occlusive disease in the terminal intracranial portions of the internal carotid arteries and proximal segments of their main branches but rarely in the vertebrobasilar or extracranial carotid systems.[9,16,17] These changes are referred to as large-vessel cerebral vasculopathy and lead to the formation of a mass of small, friable new blood vessels in response to the severe stenosis or occlusion of the major intracranial vessels. This is called moyamoya phenomenon.[18] Rupture of moyamoya's vessels leads to cerebral hemorrhage. It is observed in 5.5%–17% of patients with SCD.[19]

Cerebral hemorrhage in adults is also related to aneurysm formation. In a study by Nabavizadeh *et al*. involving 709 imaged patients, the prevalence of aneurysm was 10.8% in adults with SCD.[20] Those which rupture were typically relatively small (2–9 mm), and located at the bifurcations of major vessels both in the anterior and posterior circulation.[20,21] Histopathology studies have shown degeneration and fragmentation of the internal elastic lamina of the wall of the aneurysms. This is described as "elastorrhexis" of the vascular wall.[8,22] The hemorrhage is typically subarachnoid but may be intraventricular or parenchymal. Hence, bleeding in any form, except for traumatic subdural hematoma warrants further evaluation for a surgically correctable aneurysm by MRA even if the bleeding appears to be primarily intracerebral. Our first patient presented with an acute parkinsonism due to intracerebral bleed. She could not be subjected to MRA due to the metallic implant in the hip joint which was not MR compatible. The subarachnoid and intracerebral hemorrhage may also be caused by ischemic changes in the capillaries and arteriolar walls which lead to diapedesis of red blood cells into the subarachnoid space and into the surrounding cerebral tissue.[7]

Since 1978, 15 cases have described the occurrence of spontaneous extradural hematomas as a rare complication of SCD.[23-27] Most of these reports identify the skull bone infarction in the same area as the extradural hematoma. Majority of the extradural hematomas are a complication of underlying bone infarction disrupting the cortical bone, causing periosteal elevation and subsequent bleeding into the extradural space.[26,27] Another explanation suggested was spontaneous rupture of the epidural vessels in the vicinity of the infarcted bone.[27] Bone infarct can be visualized on magnetic resonance imaging (MRI) of the skull. We could not demonstrate the bone infarcts as both the patients of EDH refused MRI due to financial constraints.

Recently, Dahdaleh offered an alternative mechanism suggesting that patients with SCD have abnormal skull

anatomy (hypervascularity) due to chronic medullary hematopoiesis. In response to acute hemolysis, there are rapid hemopoietic tissue proliferation and expansion, which could disrupt the skull cortex and precipitate the extravasation of blood and marrow into the epidural spaces.[28] The consequences of acute bone infarction due to vaso-occlusion are insignificant in the vertebrae and long bones, including femur, humerus, radius, and ulna, which are surrounded by soft tissues.[26] However, as the skull is very thin, hematomas are likely to be much more obvious, particularly if they occur in relation to the inner surface of the skull and compress the brain.

The EDH in our patients could be secondary to acute expansion of hemopoietic tissue or have developed as a periosteal reaction to the infarcted bone. However, as both the patients had a history of vaso-occlusive pain crisis, it is likely that EDH was secondary to skull bone infarct. The fall in hemoglobin and rise in LDH could be due to the development of EDH. Our second patient had recurrent EDH in the same location suggesting some anatomical bone defect. After making on intense literature search, we did not come across a single published report of recurrent EDH in SCD. To the best of our knowledge, this is the first-case report of the recurrent EDH in SCD.

Depending on the location and volume of the hematoma, there could be an alteration of the higher mental functions for the frontal EDH, homonymous hemianopsia in the occipital ones, signs of intracranial hypertension in the frontal and occipital forms, or hemiplegia in the parietal forms.[29] A parietal swelling may develop on the side of the hematoma. EDH is a rare cause of neurological symptoms in SCA, and the clinicians should suspect this complication even in the absence of focal neurological signs or history of trauma. Small EDH without evidence of cerebral compression or raised intracranial pressure undergoes spontaneous resolution and hence, can be managed conservatively while larger ones require evacuation. Hemorrhagic stroke is among the emerging challenges in SCD care, as screening measures and preventive transfusions are reducing the incidence of ischemic stroke.

Conclusion

These cases highlight the rare neurological hemorrhagic complications of SCD. EDH is an uncommon complication of SCD, whereas recurrent EDH is extremely rare. Small EDH may not manifest with neurological deficits while the acute onset of stroke in patients of SCD may not be always related to ischemia. One must have high index of suspicion, as the early diagnosis and treatment reduces the morbidity and mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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