Case Report

Stroke in sickle cell disease: case report

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ABSTRACT

Sickle cell disease is an inherited blood disorder that affects red blood cells. It is characterized by polymerization of haemoglobin, erythrocyte stiffening, and subsequent vaso-occlusions. These can lead to microcirculation obstructions, tissue ischemia, infarction and acute stroke. Transient ischemic attack, Ischaemic stroke, haemorrhagic stroke, silent cerebral infarction, headache, Moyamoya disease, neuropathic pain, and neurocognitive impairment are neurological complications of sickle cell disease. Here we report a case of ischemic stroke in a patient of sickle cell disease. For early diagnosis and proper management of sickle cell disease neurological complications require specialised haematological and neurological expertise. The newly used medications under ongoing research will be the hope to overcome this devastating disease and its complications.

Keywords: Neurological complications, Sickle cell disease, Stroke

INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive hemoglobin disorder.¹ In SCD there is substitution of the amino acid glutamine by the valine in the sixth position of beta globin chain.² SCD is characterized by haemoglobin polymerization, erythrocyte stiffening, and subsequent vaso-occlusion.³ Increased red blood cells adhesion and shear stress may initiate the injury of endothelial cells of the blood vessels including the cerebral arteries.

A key contributor to vaso-occlusion may be the increased tendency of the sickle red cells to adhere to the vascular endothelium.⁴ As sickled red blood cells adhere to the vascular endothelium, blood flow is impeded and thereby increases the capillary transit time.⁵ The precipitating factors of sickling phenomenon include hypoxia, dehydration and metabolic acidosis.⁶

The sickled red blood cells can obstruct microcirculation and cause tissue infarction because they are less deformable. Beside the neurocognitive impairment, acute stroke and chronic cerebral ischemia are among the most disabling vascular anomalies of SCD.⁷,⁸ Neurologic complications (i.e stroke) are a major cause of morbidity and mortality in sickle cell disease. It affects from 6 to 8% of patients with sickle cell anemia, especially between 2 to 10 yrs of age. However it may also present in adults.

CASE REPORT

A 27 year old right handed male patient presented in the casualty of Mahatma Gandhi hospital, Jaipur with chief complaint of slurring of speech since 8 hrs, deviation of mouth to the right, drooling of saliva from left angle of mouth since 3-4 hrs and weakness of left side of upper and lower limb which was acute onset and non-progressive.

There was no history fever, trauma, seizure, urinary abnormalities and no other complaints suggestive of involvement of cranial nerves other than facial nerve. No significant personal history. He had history of recurrent jaundice since childhood, for which he took treatment
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intermittently from local physician. On examination, patient had a regular heart rate of 84 beats/min, respiratory rate of 16 breaths/min, blood pressure was 134/96 mmHg in right arm supine position and an oxygen saturation of 96% on room air. Pallor and icterus was present.

On neurological examination, GCS-14/15, speech was impaired with intact comprehension, all cranial nerve examination was normal except left sided UMN type of facial palsy with left sided UMN hemiparesis (Table 1).

**Table 1: Motor system examination.**

|        | Right | Left  |
|--------|-------|-------|
| Bulk   | Normal| Normal|
| Tone   | Normal| Increased|
| Power  | Proximal | 5/5   | 2/5  |
|        | Distal  | 5/5   | 1/5  |
| LL     | Proximal | 5/5   | 4/5  |
|        | Distal  | 5/5   | 3/5  |
| DTRs   | B      | ++    | Brisk|
|        | T      | ++    | Brisk|
|        | S      | ++    | Brisk|
|        | K      | ++    | Brisk|
|        | A      | ++    | Brisk|
| Plantar| Flexor | Extensor |

On sensory examination, pain, touch, temperature sensation were absent on left side of body. Signs of meningeal irritation were absent. Gait and coordination could not be assessed. Other systemic examination were within normal limit.

MRI brain was done which was suggestive of acute infarct in right internal capsule region (Figure 1).

On blood investigation there is anaemia with indirect hyperbilirubinemia. So we further investigated the patient as a case of young stroke. We have done various test:

- Anti Cardiolipin Ab: Both Ig G and Ig M negative.
- Lupus anti-coagulant: Negative.
- ANA: was negative.
- RA factor: <8.6, negative(<12),
- CRP: <5,
- 24 hrs urine protein: 240 mg/24hrs(42-225),
- S. homocysteine levels: 11.08(4.7-14.8),
- Factor VIII: 217%(50-150),
- Protein C: 40IU/L (65-135), Protein S: 90(>75).
- Cryoglobulin III: Reactive

In view of anaemia PBF was examined in which some sickle cells were found, then sickling test was done which was positive (Figure 2 and 3).

**Figure 2:** PBF show sickle cells.

**Figure 3:** Sickling test positive (within 30 min).

**Figure 4:** Hb Electrophoresis.
Hb electrophoresis was done which show (Figure 4).

- Hb A: 5.3%
- Hb F: 22.2%
- Hb S: 70.4%
- Hb A2: 2.1 %

**DISCUSSION**

In patients with SCD, stroke may occur as an acute clinical syndrome presenting with hemiplegia, convulsions and visual or hearing loss. Stroke, presenting as cortical blindness, although unusual, has been reported in SCD patients and in strokes due to other conditions. The outcome following cortical blindness with respect to regaining vision is variable, but in some patient, there was an improvement in visual acuity, suggesting that prognosis is good. The event could have occurred during or immediately following surgery, or during the convalescent period. The stress of surgery is a recognised precipitating factor for vasocclusive events in SCD.

Various therapeutic options have been found to be useful in reducing the risk of events occurring during surgery and in a district hospital with limited resources preoperative blood transfusion, adequate postoperative analgesia, inhaled nitric oxide are therapies that would have improved the anaesthetic course of the patient and possibly prevented the stroke. However, a recognised difference between childhood and adult stroke is overlapping risk factors and in some patient other factors that may have contributed to the event were anaesthesia and thromboembolism secondary to the atrial septal defect with possible arrhythmias. In addition, there is a possibility that the cerebral infarction seen in some patient was a consequence of recurrent neurological events. With increased understanding of the natural history of SCD particularly from studies in Jamaica and USA, it is apparent that there are numerous vascular events that occur in SCD patients, which are ‘asymptomatic’. These subclinical events may manifest as neuropsychological deficits, impaired intellectual function, behavioural problems etc. and have been demonstrated in Tanzania and more extensively elsewhere.

Many reports have shown that there is an increase in prevalence of SCD surviving into adolescents and adulthood in Africa suggesting that there will be a rise in the incidence of chronic end-organ damage. Most vascular events occur between the ages of five and ten years. Children with an increased risk of stroke can be identified by measuring cerebral blood flow velocity using transcranial doppler ultrasonography (TCD). Extensive studies showed that increased cerebral blood flow velocity (CBFv) of 200cm per second was associated with a 40% risk of stroke within three years.

Options for both primary and secondary intervention, include blood transfusion, hydroxyurea and comprehensive care have been effective enough to be incorporated into national policies. However, such strategies are costly to a resource-poor health system, and associated with a high risk. It is therefore important to define the high-risk groups based on evidence from studies in Africa before such interventions are proposed and integrated into appropriate management and preventative guidelines and policies at all levels of health care.

**CONCLUSION**

The incidence of ischemic and hemorrhagic stroke is increase in adults with sickle cell disease relative to general population. The prevalence of stroke is 3.75% in patients with sickle cell disease. Control of the cardiovascular risk factors is the usual strategy to prevent stroke. Simple investigations like peripheral blood film should be given proper importance. By proper diagnosing the disease as early as possible we can prevent the further complications of disease.

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