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Original Research Paper

Internal Medicine

RECURRENT CEREBROVASCULAR ACCIDENT IN A YOUNG FEMALE WITH SICKLE-BETA THALASSEMIA: A CASE REPORT.

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ABSTRACT Cerebrovascular accident (CVA) without any predisposed features is typically considered an old age disorder. However, in younger patients, the rigid shape of red blood cells (RBCs) in sickled microcirculation can cause sludging and occlusion of blood vessels, leading to cerebral ischemia. This case report describes a 17-year-old female with sickle-beta thalassemia who presented with recurrent CVA. The report highlights the diagnostic challenges, clinical course, and management of the patient, emphasizing the importance of early screening and awareness of hemoglobinopathies.1,2

KEYWORDS : Young cerebrovascular accident, Sickle-Beta Thalassemia

INTRODUCTION:

Cerebrovascular accidents in younger patients can be linked to sickle cell disease (SCD), where sickled red blood cells cause vascular occlusion and ischemia.[3] SCD patients are also at risk for hypercoagulable states and endothelial injury, leading to stroke.[4] This case report aims to increase awareness and prevent complications associated with hemoglobinopathies in younger individuals.[5]

Case Report:

- **Presentation:**

A 17-year-old female presented with fever, right-sided chest pain, cough, shortness of breath, and joint pain for 5 days. She also developed right-sided weakness. The patient had similar episodes of right-sided weakness two times and left-sided weakness once in the last four years, but she had not received proper medical treatment or investigation. There had been no residual weakness following the previous episodes.

Consequently, her sickle-beta thalassemia remained undiagnosed until this admission. At this presentation, she was diagnosed with acute coronary chest syndrome due to pneumonia, leading to a sickling crisis and subsequent right-sided weakness due to ischemic infarct.

Table 1: Vitals trend during admission

Vital Signs	Day 1	Day 3	Day 5
Temperature (°F)	101.2	99.8	98.6
Pulse (/minute)	106	92	82
Blood Pressure (mmHg)	90/60	100/72	110/86
Respiratory Rate (/minute)	24/minutes, laboured	20/minutes	16/minutes
Neurological Examination	• Conscious oriented to time, place and person • Right sided Upper limb power+2, Right lower limb +3, right sided facial weakness, right sided Babinski sign+ • Cranial nerves: 7th Cranial nerve upper motor type weakness. • Rest all cranial nerves: no abnormalities • Left-sided upper and lower limb examination: normal • No cerebellar signs		
Systemic examination	• Spleen palpated just below the left costal margins		

Investigation:

CECT: contrast enhanced CT, USG: ultrasonography, NRBCs-Nucleated RBCs

Table 2: Investigation during admission

Investigation	Day 1	Day 2	Day 3
White blood cell (cells/cumm)	33000	26400	14000
Red blood cell (millions/cumm)	2.5	3.4	4
Haemoglobin (g/dL)	7.5	9.1	9.6
Mean corpuscular volume (fL)	63.2	66	73
Platelet (cells/cumm)	128000	90000	124000
International normalised ratio/Prothrombin time	1.2	0.9	0.9
Procalcitonin (ng/mL)	5.7	3.5	1.9
C- Reactive protein (mg/L)	172	130	60

Arterial blood gas analysis	PH: 7.12 Lactate: 5.7 mmol/L PaO2: 56 mmhg Bicarbonate: 14 meq/L Paco2: 30 mmhg		PH: 7.32 Lactate: 1.7 mmol/L PaO2: 86 mmhg Bicarbonate: 20 meq/L Paco2: 44 mmhg
Blood Culture			Streptococcus pneumoniae within 48 hours, from 2 blood cultures of different site
Sputum Culture			Streptococcus pneumoniae (sensitive to Ceftriaxone and co-amoxiclav)
Peripheral blood smear examination	• Marked anisopoikilocytosis • Target cells++, sickle cells+ • Few polychromatic RBCs, • 68/100 NRBCs		
Sickling test	Positive		
Hemoglobin Electrophoresis			Double heterozygous for Hbs and B thalassemia • HbS -62.1% • Hb A- 3.4% • Hb F -29.7% • HbA2- 4.8%
CECT Brain	• Gliotic areas in subcortical and periventricular white matter of the left frontal and parietal lobes, left centrum semiovale mid ex vacuo dilation of the left ventricle • parenchymal volume loss in left frontal and parietal lobes with prominent sulci.		
MRI brain without contrast	• Same findings as in CECT brain, features suggestive of acute ischemic infarct in left frontal and parietal lobes		
Chest X-ray (Image 1)	Right-sided middle and lower lobe community-acquired pneumonia.		
USG Chest		Right-sided	moderate pleral effusion (4.5 cm maximum band) with underlying consolidation and collapse.

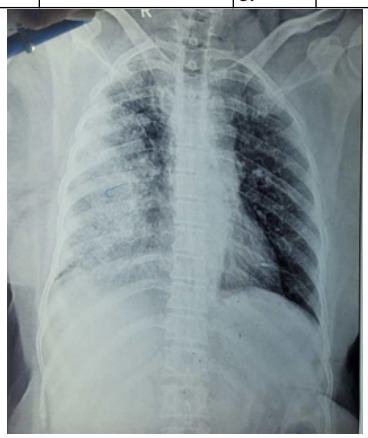

Image 1: Chest X Ray on Day1- suggestive of right-sided middle and lower lobe community acquired pneumonia

- **Treatment And Follow Up:**

Patient was started on intravenous injection Ceftriaxone for 5 days, followed by tablet Amoxicillin-Clavulanic Acid 625 mg three times a day for 5 days upon discharge. For pain management, injection Tramadol 50 mg twice daily and as needed was given. Oxygen Support was Provided at 4 liters per minute for 3 days, which was gradually tapered, and the patient was off oxygen after 5 days. One unit of Packed- Red blood cells were transfused on day 1 and day 2.

Gradually after 7 days of treatment, the patient was discharged with Tablet Aspirin 75 mg once daily, tab Hydroxyurea 500 mg twice daily, tab folic acid 5mg once daily.

- **Follow-Up and Monitoring:**

Routine follow-Up Visits and haemoglobin measurement were scheduled every 2 months with emergency examination if symptoms such as fever or shortness of breath develops. Family members were advised to undergo screening for sickle cell disease and thalassemia.

DISCUSSION:

Patients with sickle cell disease (SCD) are at a higher risk of developing cerebrovascular accidents (CVAs) compared to the general population. The prevalence of stroke in SCD patients is approximately 3.55%.[7] Adults with SCD are particularly vulnerable to recurrent strokes within two years of their initial event.[8] Ischemic strokes in SCD patients are associated with several factors including previous transient ischemic attacks, sepsis, dehydration, elevated systolic blood pressure, acute chest syndrome, and nocturnal hypoxemia.[9]

In this case, the patient's recurrent CVA was likely exacerbated by her pneumonia, which leads to microvascular occlusion due to sickled red blood cells. Hydroxyurea was prescribed as part of the management strategy to reduce the frequency of sickling crises and overall hemolysis by increasing fetal hemoglobin (HbF) levels, which helps reduce

the sickling of red blood cells.[10] Aspirin was included in the treatment regimen to address potential hypercoagulability and reduce the risk of thrombotic events. Both therapies are well-established in managing sickle cell disease complications and preventing stroke.

• CONCLUSION:

Cerebrovascular accidents are rare but severe complications in young patients with sickle-beta thalassemia. Early diagnosis and management are crucial to improve patient outcomes and preventing recurrence. Implementation of routine hemoglobin electrophoresis and sickling tests in newborns and at-risk populations to identify individuals with sickle-beta thalassemia early. Evaluation of family history for hemoglobinopathies to identify individuals at higher risk and initiate early intervention.[11]

This case underscores the need for increased awareness and screening for hemoglobinopathies in young patients presenting with recurrent CVAs. Appropriate treatment and follow-up can lead to significant improvement and reduce the risk of severe outcomes.

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