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An infant Presenting with Cerebrovascular Accident was Diagnosed as a Sickle Cell Disease Patient: a Case Report

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Abstract

Sickle cell disease (SCD) is a known inherited hemoglobin disorder featured by the presence of sickle shaped erythrocytes in the blood. It can cause cerebrovascular accident (CVA) in adults and children and is responsible for the majority of the strokes in children. Repeated blood transfusion are often required in an attempt to dilute blood thus reducing the risk of vaso-occlusion and stroke in this patients. We report a case of a 13 months old boy, known patient of sickle cell disease with some special signs and symptoms.

Keywords: Sickle cell, Cerebrovascular accident, Children, Stroke

Introduction

Cerebrovascular accident (CVA) is a major complication of patients with sickle cell disease. Sickle cell disease (SCD) is a common inherited hemoglobin disorder characterized by the presence of sickle shaped erythrocytes in the blood (1, 2). This vaso-occlusive phenomena can lead to various adverse effects in different organs, recurrent painful episodes, life-long disabilities and even death (3).

Cerebrovascular accident is an expected event in adults. However, this stroke in children is mainly due to sickle cell anemia. In fact, sickle cell disease is the most prevalent cause of childhood stroke. Based on the previous reports, the reported risk of first CVA in the first 20 years of life is 0.761 per 100 patients with sickle cell anemia (4).

The majority of strokes in patients with sickle cell anemia disease, mainly occur in the age range of 3 to 14 years old and this event is relatively rare in children less than 3 years of age (5). In addition, cerebrovascular accident has wide spectrum of neurologic signs and symptoms. The symptoms of ischemic stroke may include hemiparesis, dysphasia, gait disturbance and a change in the level of consciousness (6).

In this study, we report a case of a 13 months old boy, known patient of sickle cell disease, who presented with recurrent stroke and neurological symptoms.

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A 13-month-old boy patient presented in October 2015 with the complaint of URI (Upper Respiratory Infection), fever, repeated vomits, reduced consciousness level and status epilepticus since a night before admission. History of his illness indicated that the patient was admitted at the department of respiratory medicine in Shahid Mohammadi Hospital, Bandar Abbas in six months old for Pneumonia. Parents were suffering from thalassemia minor and had no family consanguinity. Results of his physical examination on admission were blood pressure 90/60 mmHg with a heart rate (HR) of 120 beats per minute (bpm), respiratory rate (RR) 68, and oral temperature 38.9 C. Bilateral rales were heard by lung auscultation. Child opened his eyes only by painful stimulation and mydriasis was observed in both pupils. DTR (Deep Tendon Reflex) in right eye was decreased and the patient was admitted by the principle diagnosis of encephalitis and broad-spectrum antibiotic treatment was initiated. The convulsion of patient was not controlled by phenytoin and phenobarbital. Therefore, midazolam and management techniques for raised intracranial pressure were used to manage the problem. Thereafter, complete blood count (CBC) test was conducted and the results were as follows: MCV: 62.8, PIT:760, Hb:6, RBC:3360000 and WBC:6800. In addition, the blood samples were sent to check anemia and the patient received packed cell. The results of electrophoresis were as follows:

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HbD:56.9, HbF:35.9 and HbA2:7.4. Based on these findings, the patient was suffering from sickle cell anemia. After stabilizing the patient, Computed Tomography (CT) scan was requested. It revealed sub-arachnoied hemorage (SAH), subdural hematoma and hydrocephalus. Thus, peritoneal shunt method was used to cure the problem and then the patient was transferred to the ward for subsequent observation and management. The patient was discharged a few days later.

Discussion

As mentioned above, CVA is one of the leading cause of death in children with sickle cell anemia. For this reason, it is necessary to identify the candidates for CVA, as soon as possible. In this regard, Trans-Cranial Doppler Ultrasound (TCD) is a useful tool to identify children at high risk for stroke, before they have suffered a clinical stroke (7). According to the previous investigation, routine use of transcranial Doppler screening, coupled with regular blood transfusion therapy, has decreased the prevalence of overt stroke from 11% to 1% in children with sickle cell anemia (8). In this issue, children with Hb SS or HbS beta 0-thalassemia between the ages of 2 and 16 should be considered candidates for TCD screening every 12 months (9). Depending on a previous studies, there are several risk factors of stroke in sickle cell patients including blood current speed in cranial transcranial sonography, low levels of Hemoglobin, the number of white blood cells (WBC), brain silence and acute chest syndrome (ACS) (10). CVA includes any acute neurologic event secondary to arterial occulsion or hemorrhage that results in an ischemic event associated with neurologic signs or symptoms (11). Among first CVAs in patients with SCD, 54% were caused by cerebral infarction, 34% due to intracranial hemorrhage, 11% by TIA and 1% had characteristics of both infarction and hemorrhage (4).

Current management strategies and therapies for secondary prevention of strokes and silent cerebral infarcts also include regular blood transfusion therapy and in selected cases, hematopoietic stem cell transplantation (12). In fact, blood transfusion therapy and bone marrow transplantation can reduce the danger of stroke (8). In the other words, these strategies are mandatory to maintain the level of HbS at <30% to reduce the risk of recurrence (13).

Conclusions

This report revealed that CVAs in patients with SCD is probable in the first months of birth and has a wide spectrum of manifestations which is required more experiments to clarify. Thus, it is suggested that all of the SCD patients be considered as the candidates of Trans-Cranial Doppler Ultrasound (TCD) method from birth.

Conflict of Interest

The authors declare that they have no conflict of interests.

References

1. Alam M, Lodhi M, Khan D. Cerebrovascular accident in sickle cell disease. Journal of the College of Physicians and Surgeons--Pakistan: JCPSP. 2003;13(1):55-6.
2. Serajee FJ, Sarnaik SA, Altinok D, Huq AM. Stroke in sickle cell disease. Journal of Pediatric Neurology. 2010;8(3):299.
3. Okpala I, Daniel Y, Haynes R, Odoemene D, Goldman J. Relationship between the clinical manifestations of sickle cell disease and the expression of adhesion molecules on white blood cells. European journal of haematology. 2002;69(3):135-44.
4. Ohene-Frempong K, Weiner SJ, Sleeper LA, Miller ST, Embury S, Moohr JW, et al. Cerebrovascular accidents in sickle cell disease: rates and risk factors. Blood. 1998;91(1):288-94.
5. Kwiatkowski J, Ohene-Frempong K. Stroke in sickle cell disease. Practical Management of Haemoglobinopathies. 2004:134-44.
6. Edlow JA, Newman-Toker DE, Savitz SI. Diagnosis and initial management of cerebellar infarction. The Lancet Neurology. 2008;7(10):951-64.
7. Charache S. Treatment of sickling disorders. Current opinion in hematology. 1996;3(2):139-44.
8. Kassim AA, Galadanci NA, Pruthi S, DeBaun MR. How I treat and manage strokes in sickle cell disease. Blood. 2015;125(22):3401-10.
9. Ware RE. How I use hydroxyurea to treat young patients with sickle cell anemia. Blood. 2010;115(26):5300-11.
10. Kral MC, Brown RT, Nietert PJ, Abboud MR, Jackson SM, Hynd GW. Transcranial Doppler ultrasonography and neurocognitive functioning in children with sickle cell disease. Pediatrics. 2003;112(2):324-31.
11. Prengler M, Pavlakis SG, Prohovnik I, Adams RJ. Sickle cell disease: the neurological complications. Annals of neurology. 2002;51(5):543-52.
12. Hulbert ML, McKinstry RC, Lacey JL, Moran CJ, Panepinto JA, Thompson AA, et al. Silent cerebral infarcts occur despite regular blood transfusion therapy after first strokes in children with sickle cell disease. Blood. 2011;117(3):772-9.
13. Wanko SO, Telen MJ. Transfusion management in sickle cell disease. Hematology/oncology clinics of North America. 2005;19(5):803-26.