CASE REPORT

SICKLE CELL ANEMIA WITH ACUTE SPLENIC SEQUESTRATION

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SUMMARY

A three and half years old girl, a known sickler presented with abdominal pain of four days, vomiting, cough and fever of three days. She had history of chest infections and fever which were treated as pneumonia. On examination she was febrile, had a tinge of jaundice, severe palmor pallor, hepatomegally with span of 11 cm, enlarged spleen 5 cm below costal margin. Respiratory rate was 40/min and crackles were heard on the lower right lung. Pulse rate was 130 beats/min, regular and gallop rhythm was present. Diagnosis of acute splenic sequestration syndrome and non-severe pneumonia was made. Packed cell volume blood transfusion was done twice and antibiotics were given. She was discharged after five days stay in the hospital.

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INTRODUCTION

Sickle cell disease is a clinical syndrome due to a presence of abnormal gene responsible for manufacturing Haemoglobin (Hb). The gene HbS causes replacement of glutamic acid in position 6 of normal β-chain by valine molecule. This results in deoxygenated HbS to be 50 times less soluble than deoxygenated HbA causing Red Blood Cell to sickle at lower oxygen concentration. The gene is autosomal recessive, inherited in Mendelian fashion[1]. The disease was first described by James Herrick in 1910 in a 20 year old West Indies dental student who had recurrent jaundice, fever and chronic ulceration of extremities. Patient was anemic and red blood cells were elongated and crescent shaped resembling the blade of a sickle.[2]

HbS gene occurs widely across tropical Africa, some countries bordering northern shores of the Mediterranean Sea and in parts of Middle East and Southern India. Prevalence may vary from low values to 40%. In black Americans, prevalence is 8%. The distribution of HbS gene corresponds to the area in which Falciparum malaria is endemic or has been endemic[1,3]. In Dar-es-salaam, Tanzania approximately 17% of children entering hospital for any reason were carriers of the sickle cell gene, of these one in every four have homozygous form (HbS). In other areas like Karagwe, 13% of the population are carriers and one in every 30 patient has homozygous sickle cell disease[4].

Sickle cell anemia can be diagnosed at birth but in 90% of population clinical abnormalities do not occur before age of 3-6 months where replacement of HbF by HbS occur. The disease can be diagnosed by sickling test and electrophoresis.[5,6]

PRESENTATION OF CASE

A known sickler, three years and six months old girl from Buguruni (a suburb in Dar) presented on the day of admission with abdominal pain for four days, vomiting, cough and fever for three days. The informant was the biological mother and she came from home.

Abdominal pain was of sudden onset, dull aching, constant and it was all over the abdomen. It was made worse by eating solid food, no relieving factors, no history of diarrhea and it was associated with poor appetite. Fever was of low grade, sudden onset, intermittent in nature, higher in the evening and temporarily relieved by Paracetamol. It was associated with vomiting, but was not however not associated with convulsions, ear discharge or discomfort in micturition. Vomiting occurred 2-3 times a day, immediately after eating especially when given solid foods and was non-projectile. Vomitus contained recently eaten food and mucous, had a colour of bile and was not blood stained. Vomiting was not aggravated by cough and no relieving factors. Cough was of gradual onset, initially dry, later became productive. Sputum was yellowish and small in volume. Cough was not posture related, not associated with difficulty in breathing or wheezing and there is no history of open TB contact. Review of other systems was uneventful.

On past medical history, mother booked at antenatal clinic at sixteenth week of gestation she

was given haematenics, intermittent prophylaxis treatment for malaria. ELISA for HIV and VDRL for syphilis were both negative. She was normotensive and there were no complains during pregnancy. She delivered at term, Spontaneous Vaginal Delivery and the baby weighed 3.7 kg, cried immediately, breastfed after 30 minutes and was discharged on the same day. Baby was taken back to the hospital the following day with septic cord and it was treated. At age of nine months she had fever and diarrhea. It was treated with antimalarials. At age of one year, she had chest tightness and difficulty in breathing which was treated as pneumonia. At age of two and half years she had febrile convulsions, tonic clonic for about four minutes, she lost consciousness for ten minutes and was not associated with urine or fecal incontinence. She was very anaemic with severe palmar pallor and pain in the hands. Sickle cell Anemia was suspected and confirmed by sickling and electrophoresis at Muhimbili National Hospital (MNH). She was treated with antimalarials and anticonvulsants, pain was relieved with paracetamol. She recovered fully and she is now on folic acid 5mg once a day and chloroquine 75 mg once weekly. She also attend sickle cell clinic at MNH.

The baby was exclusive breastfed for three months weaned with formula milk and maize flour porridge containing milk, butter and eggs. At present she eats family food and the food was adequate in quality and content

The index is the only child to the mother who is 30 years old with primary education and she owns a hair salon. Father is a 42 years old with primary education and he is a petty trader. They cohabited but currently are separated and he supports his daughter, the father is married and has other children who are doing fine. Both parents have sickle cell trait

On examination she was alert, febrile (38ºC), had a tinge of jaundice, severe palmor pallor, but no digital clubbing, cyanosis, lymph node enlargement and oedema of lower limbs. Weight was 14.5 Kg, height 104.5 cm and mid-upper circumference 15.8 cm. These anthropometric measurements, were normal with exception of Wt/Ht which indicated mild wasting.

On systemic examination, spleen was enlarged 5 cm from the lower costal margin. Liver was also enlarged, not tender extending 11 cm from the fifth intercostal space.

Respiratory rate was 40 breaths per minute, and on auscultation reduced breath sounds and fine crackles were heard on the right lower lung.

Pulse rate was 130 beats per minute, regular, normal volume, non-collapsing and synchronous with femoral artery. Blood pressure was 95/66. Other systems were essentially normal.

Investigations carried out were blood haemogloin level which was 3g/dl, PVC 9%, blood for malaria parasites was negative, chest X-ray showed patchy opacification on the right lower lobe. Mid-stream urine for culture and sensitivity showed insignificant bacterial growth. Full blood picture revealed slightly raised WBC, RBC was very low ($1.48x\ 10^6/mm^3$), Hb was 3.8g/dl and there was hypochromic microcytic anaemia. Reticulocytes were 0.6% and she was blood group A+. Final diagnosis was sickle cell anemia with acute splenic sequestration and Non-severe pneumonia

She was treated with syrup ampicillin 50mg/kg and cloxacillin 50mg/kg for five days, declofenac, folic acid 5mg once daily for one month and increased fluid intake was encouraged. Blood transfusion were carried twice, packed cell volume was given at rate of 10mls/kg for four hours. Progress of patient was good, fever subsided, splenic size was reduced, palmor pallor was reduced. jaundice was minimal, breath rate 30 breaths/min, pulse rate 125/min, and abdominal pain disappeared. She was discharged after five days in hospital.

DISCUSSION

Acute splenic sequestration crisis (ASSC) is caused by intrasplenic trapping of red cells causing precipitous fall in Hb levels and potential for hypovolemic shock. ASSC has become a leading cause of death in children with sickle cell anemia. It may be defined by a decrease of at least 2g/dl from steady state Hb concentration, elevated reticulocytes counts and acute enlarging spleen. In most cases first attack occurs between three months and five years as in this patient. They are associated with viral or bacterial infections. Clinical manifestations are sudden weakness, severe palmor pallor, tachycardia and abdominal fullness. In typical ASSC patients usually present with hypovolemic shock which is life threatening but our index case did not have shock hence atypical ASSC. Aplastic anemia was ruled out because Full Blood Picture didn't show pancytopenia. Also no haemorrhagic manifestations (petechiae, ecchymoses, bleeding in GIT) resulting from

thrombocytopenia. Furthermore splenomegally is rare in aplastic anemia. Immediate treatment is directed toward correction of hypovolemia with red cell transfusion which was done twice. Once transfusion is complete, red cells sequestered in the spleen are remobilized, splenomegally regress and Hb level increases. Patients who have a life threatening episode of ASSC which require acute transfusion should have splenectomy shortly after the event. This was not done because it was first episode and not life threatening. Pneumococcal vaccine is not readily available in Tanzania, so this child was not given the vaccine. Those below two years should be placed on chronic transfusion program to maintain Hb level. An important factor which may influence decision about splenectomy will be widespread availability of conjugated vaccine to prevent *S. pneumoniae* infections which are common in children with sickle cell anemia and require prompt antibiotic therapy[7].

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