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Unusual features of haemolytic crisis in sickle-cell anaemia

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Summary

Olanrewaju DM, Ogunfowora CB and Njokanma FO. Unusual features of haemolytic crisis in sickle-cell anaemia. *Nigerian Journal of Paediatrics 1992;* **19:44.** The case of a 27-month old patient with sickle-cell anaemia who developed haemolytic crisis following a febrile illness of one week's duration, is presented. The unusual features of the case included a dangerously low PCV of three percent, absence of any significant jaundice or heart failure, or any manifestations of hypoxic encephalopathy, such as seizure. The patient was fully conscious and not in shock on admission. Urgent blood transfusion was given to the patient within 30 minutes of arrival in the hospital. Recovery was rapid and the post-transfusion PCV reached 15 percent; the patient was discharged some 72 hours after admission. The rapid recovery without any sequalae could be attributed to the urgent treatment that included pre-transfusion diuretic and oxygen therapy and blood transfusion.

Introduction

HOMOZYGOUS sickle-cell anaemia (SCA) is one of the commonest problems of African children and in some parts of the continent as many as two percent of all newborn babies are known to have the haemoglobin-S genotype.[1] The gene frequency ranges between 10 percent in North Africa and 25 percent in West Africa.[2] Haemolytic crisis is one of the commonest complications of the disorder and in the tropics, it usually follows infections. Typically, patients in crisis present with fever, varying degrees of pallor, jaundice and enlargement of the liver and spleen.[3] Patients tolerate relatively low packed cell volumes (PCV) during crisis and anaemic heart failure does not usually occur until the PCV falls to very low values. Besides meningitis and cerebrovascular occlusion, severe anaemia in SCA may also be associated with encephalopathy, manifesting as alteration in level of consciousness, recurrent seizures or even coma,[4] the lower the PCV the greater the chances of these events occurring.

To the best of our knowledge, there has been no previous report in Nigeria of a PCV as low as three percent during haemolytic crisis in a sickler who was not in heart failure; the patient was fully conscious on presentation at

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the hospital and had no residual disability on recovery. The purpose of this short communication is to report such a case with unusual features as described above.

Case Report

KO, a 27-month old male child, was admitted to the children's emergency room (CHER), Ogun State University Teaching Hospital (OSUTH), Sagamu, with a history of weakness, poor appetite and vomiting for one day. One week prior to the admission, he had been given some treatment for a febrile illness associated with mild jaundice and swelling of the hands and feet at a local chemist's drug store. The mother denied any other past history of swelling of the hands and feet, or jaundice; there was no past history of hospitalization. Delivery was by spontaneous vertex at term in a hospital following an uneventful pregnancy. Birthweight was unknown.

Physical examination revealed an ill-looking child, weighing 9.0 kilograms. There was extreme pallor of the conjunctivae, buccal mucosa and the nail beds. There was no jaundice. The temperature was 37.2°C. The respiratory rate was 54 per minute, but the chest was clinically clear. The heart rate was 150 per minute. There were no other abnormal signs in the cardiovascular system. The liver was enlarged to 2cms below the costal margin and the spleen also enlarged to 3cms. In the central nervous system, there was no remarkable sign.

Investigations revealed a PCV of three percent, Hb 1gm/dl, blood group O Rh +ve and HbSS on haemoglobin electrophoresis. The total WBC was 8.5 x 10^9/L (8,500/mm^3) with 60 percent lymphocytes and 40 percent neutrophils. Blood film examination for malaria parasites was negative; urine and blood cultures yielded no growth. Initial management consisted of nursing in a cardiac position, administration of oxygen by nasal catheter and intravenous administration of fursemide (1mg/kg); these measures were taken as precautions against heart failure which could occur with blood transfusion. Transfusion with packed red blood cells was commenced within 30 minutes of arrival in CHER and he received a total of 10ml/kg of blood over a period of two hours. Chloroquine was administered subcutaneously, 5mg/kg stat, followed by an oral medication (10mg/kg/day) for two days. No antibiotics was administered. By the next day, the clinical condition of the patient was stable except for mild mucosal pallor. Folic acid, multivitamins and pyrimethamine (Daraprim) were added to the treatment. Normal feeding was restored. The temperature remained normal and the respiratory rate dropped to 40 per minute and the pulse rate fell to 120 per minute. The child was discharged 72 hours after admission with a post-transfusion PCV of 15 percent and without any neurological disability. The most recent PCV was 18 percent at the outpatient follow-up, some six weeks after discharge.

Comments

Sickle-cell anaemia is a common cause of morbidity and mortality in several countries where the condition exists. In our experience at OSUTH, SCA is second to malaria infection as a cause of childhood anaemia. Severe haemolytic crisis in SCA typically presents with anaemic heart failure and features of hypoxic encephalopathy.[3,4] The clinical features in our patient were, without any doubt, consistent with severe haemolytic crisis, secondary, possibly to malaria infection; therefore, absence of some of the typical features of this complication of haemolytic crisis makes the case unusual. Firstly, the dangerously low PCV of three percent is considered as an unusual case in HbSS disease. It is noteworthy that such a low PCV may occur in acute sequestration crisis complication, but such a sickler usually

presents in shock, due to the rapidity of fall in the haematocrit.[5] Our patient was fully conscious and not in shock on admission and this feature would exclude the complication being a sequestration crisis. The survival of our patient without any disability was also unusual. It is our belief that the complete recovery of the patient was due partly to the absence of any bacterial infection, as evident by the sterile urine and blood cultures and partly to the urgent blood transfusion that was started within 30 minutes of admission. Secondly, the absence of significant jaundice in the patient was probably due to the very low haemoglobin content of the erythrocytes with the consequent production of minimal bilirubin on lysis of the cells. It should be noted that this feature of haemolysis without jaundice is similar to that of Thalassaemia major in which jaundice may not be present, or indeed very mild and due to poor haemoglobinization of the erythrocytes.[6] Thirdly, the absence of heart failure and seizures was also an unusual feature of the case. This may be explained on the basis of the suddeness and the short duration of the haemolytic process.

References

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