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Case Report

Sickle Cell Disease in Teenager with Heart Failure as First Disease Presentation

Elham Babikir*[1], Dr Hayat Osman Abdalla[2], Dr Sarrh Siddig Elamin Sobeir[3]

1, Telford and Shrewsbury NHS TRUST.

2. Jafar Ibn Auf hospital, Sudan Paediatric consultant.

3. Assistant Consultant neonatologist, king Fahad medical city, Saudi Arabia.

***Correspondence to:** Elham Babikir, Paediatric specialist at Shrewsbury and Telford NHS Trust. e.Babikir@nhs.net

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Abstract

A 13-year-old female from western Sudan presented with fever, joint pain, and jaundice for a few weeks. She had a positive family history of sickle cell disease. On examination, she appeared unwell, small for her age, and clinically jaundiced and pale. A cardiac murmur and hepatomegaly were noted. While rheumatic fever was initially suspected, further investigations revealed a sickle cell crisis with haemolytic anaemia and elevated reticulocyte count. Chest X-ray showed enlarged cardiac shadows and no lung abnormalities. Echocardiography confirmed tricuspid and mitral valves regurgitation. The patient was treated with antibiotics and managed with a multidisciplinary approach involving cardiologists and haematologists. The case highlights the importance of considering sickle cell disease as a cause of heart failure in young children, even without previous recurrent sickle cell disease presentations.

Background

This case report emphasizes the significance of early diagnosis and management of sickle cell disease, as well as the need for a comprehensive evaluation when approaching heart failure in young children in developing countries.

Case Presentation

The 13-year-old female from western Sudan presented with fever, joint pain, and jaundice. Her medical history revealed a positive family history of sickle cell disease. Physical examination showed clinical signs of jaundice, pallor, an apical pansystolic murmur, and hepatomegaly. A working diagnosis of rheumatic fever was considered due to the patient's age and endemic area, but further investigations revealed a sickle cell crisis.

Investigations

Laboratory tests showed haemolytic anaemia with a haemoglobin level of 7 g/dL, leucocytosis of 25,000 cells, and an elevated erythrocyte sedimentation rate. Blood cultures were negative, but C-reactive protein levels were elevated. The patient also had hyponatremia on renal profile check. Reticulocyte count was significantly elevated, and HB electrophoresis confirmed HBSS sickle cell disease.

Treatment

The patient received antibiotics and underwent fluid management to address the sickle cell crisis and heart failure. Collaboration between cardiologists and haematologists was crucial in managing the patient's condition.

Outcome and Follow-Up

The patient showed improvement, gained weight, and remained compliant with medications during follow-up visits. She was in good condition, indicating successful management of her sickle cell disease and heart failure.

Discussion

This case report highlights the late diagnosis of HBSS sickle cell disease without early childhood recurrent sickle cell disease presentations. It also emphasizes the importance of a comprehensive approach when evaluating heart failure in young children, considering multiple factors that may contribute to the condition in developing countries.

Learning Points

- Early diagnosis of sickle cell disease is crucial for effective management and prevention of complications and it worth screening in Sudan.

- Consider sickle cell disease as a possible cause of heart failure in young children, even without previous recurrent sickle cell disease presentations.

- Collaboration between different specialties, such as cardiologists and haematologists and general paediatrician is essential for a comprehensive and successful management approach.