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Cholecystitis – Mode of First Presentation of Sickle Cell Anemia in an Adult Nigerian: A Case Report

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Author's contribution

The sole author designed, analyzed and interpreted and prepared the manuscript.

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Case Study

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ABSTRACT

Background: This article aimed to sensitize the healthcare providers to the possibility of late and atypical presentation of sickle cell anemia (SCA).
Case Presentation: A 24 year old female university undergraduate presented to the Medical out-patient Department of our hospital with complaint of abdominal pain, vomiting, and yellowness of the eyes, all of 2 days prior to presentation. Abdominal pain was located at the right hypochondrial region, colicky in nature, not relieved nor aggravated by any known factor and does not radiate to any part of the body. There was associated vomiting, fever, weakness, and loss of appetite. Patient has been on apparent good health. No previous history of hospital admission(s), surgery nor blood transfusion. Physical examination showed an acutely ill-looking young girl in painful distress, febrile to touch, moderately pale, icteric, no pedal edema and no peripheral lymphadenopathy. Abdominal examination showed right hypochondrial tenderness, hepatomegaly and splenomegaly. A provisional diagnosis of hepatitis was made. However, in the course of investigations, there was no evidence of hepatitis rather cholelithiasis was reported by abdominal ultrasound scan. In the course of investigating for the cause of cholelithiasis, patient was diagnosed to have sickle cell anemia.. She subsequently had cholecystectomy done and is currently being followed up.
Conclusion: Patients with SCA usually present in infancy or early childhood with dactylitis, bone

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pain and/or symptomatic anemia which usually require frequent hospital admissions and blood transfusion especially in the absence of adequate care and follow up. This case is therefore to alert the health care providers to have high index of suspicion for late presentation in an atypical manner.

Keywords: Sickle cell anemia; cholecystitis; splenomegaly; Nigeria.

1. INTRODUCTION

Sickle cell anemia (SCA) is an inherited form of anemia in which a mutated form of haemoglobin distorts the red blood cells into crescent shape at low oxygen levels [1]. It mainly affects people of African, Eastern Mediterranean, Middle eastern, Caribbean and Asian origin [1].

The most common clinical manifestation of SCA is vaso-occlusive crisis which occurs when the microcirculation is obstructed by sickled red blood cells causing ischaemic injury to the organ supplied with resultant pain [2]. SCA may lead to various acute and chronic complications, several of which have high mortality [3]. It usually present in infancy or early childhood with one of the earliest clinical manifestation being dactylitis. Other modes of presentation include generalized bone pain and symptomatic anemia. Splenic sequestration is common in children with SCA, particularly in those less than 5years old. It is not common in adults because repeated vaso-occlusion and infarction over time leads to autosplenectomy, [4] which increases the risk of infection.

The care of people with SCA may include infection prevention with vaccination and antibiotics, high fluid intake, folic acid supplementation, pain medication, malaria prophylaxis, blood transfusion and bone marrow transplantation [5].

Early detection, proper care and follow up helps to prevent or delay development of complications and early death [6]. Without adequate care, patients usually present at early age with poor clinical outcome. I present here an adult patient who presented with cholecystitis and was diagnosed for the first time to have sickle cell anaemia.

2. CASE REPORT

A 24 year old female university undergraduate presented to the Medical out-patient Department of our hospital with complaint of abdominal pain, vomiting, fever and yellowness of the eyes, all of 2 days prior to presentation. Abdominal pain was located at the right hypochondrial region, colicky in nature, not relieved nor aggravated by any known factor and does not radiate to any part of the body. Vomitus was made up of recently ingested food particles, non-bloody and non-bilous. There was associated yellowness of the eyes, fever, weakness and loss of appetite but no associated pruritus, facial, leg nor abdominal swelling. No change in urine volume. However, patient's urine colour became deep yellow. Patient has been on apparent good health. No previous history of hospital admission(s), surgery nor blood transfusion, though she has been having recurrent yellowness of the eyes with intermittent feeling of unwell. Physical examination showed an acutely ill-looking young girl in painful distress, febrile to touch, moderately pale, icteric, acyanosed, not dehydrated, no pedal edema and no peripheral lymphadenopathy. Abdominal examination showed right hypochondrial tenderness, hepatomegaly of about 6cm below the right costal margin and splenomegaly of about 8cm below the left costal margin. No demonstrable ascities. Bowel sound was normoactive. Other systems were normal. A provisional diagnosis of hepatitis with hypersplenism was made.

Investigations done showed haemoglobin level of 7.5 g/dl, white cell count of 4.5 X 10^9/l and platelet count of 446 X 10^9/l; HBsAg and HCV Ab were negative, liver function test showed elevation of both total and conjugated bilirubin with normal level of liver enzymes. Abdominal ultrasound scan reported cholelithiasis with normal thickness of the gall bladder wall, hepatomegaly and splenomegaly. Urinalysis showed urine colour to be deep amber with presence of bilirubin. Diagnosis was thereafter changed to obstructive jaundice querry cause to rule out sickle cell disease. Haemoglobin electrophoresis done showed Hb SS genotype. This was confirmed with High Performance Liquid Chromatography (HPLC) which still reported Hb SS (with fetal haemoglobin level of 2.6 and Hb A_2 of 2.8) following which a diagnosis of cholecystitis secondary to cholelithiasis in sickle cell anaemia was made. Patient

subsequently had cholecystectomy successfully done and is currently being followed up.

3. DISCUSSION

Sickle cell anemia usually manifest in infancy or early childhood [7]. For the first 6months of life, infants are protected largely by elevated levels of haemoglobin F; soon after, the condition becomes evident as haemoglobin F level begins to be replaced with haemoglobin S as age advances. Absence of adequate medical care is usually associated with high morbidity and mortality. The index case manifested for the first time in adulthood which is unusual, despite the fact that she was not being followed up and/or receiving any kind of medical care. Her elevated haemoglobin F level may have contributed. Studies have reported that high haemoglobin F level reduces the chances of red blood cell sickling [8].

Cholelithiasis is common in patients with SCA, as chronic hemolysis with hyperbilirubinaemia is associated with the formation of bile stones [9]. Cholelithiasis may be asymptomatic or result in acute cholecystitis, requiring surgical intervention. Though studies have reported that the most common clinical manifestation of SCA is vaso-occlusive crisis, [10] the index case manifested with cholecystitis secondary to cholelithiasis.

Splenic sequestration is usually seen in children with SCA, particularly in those less than 5years of age [11]. This complication is characterized by the onset of life-threatening anemia with rapid enlargement of the spleen and high reticulocyte count. However, because of repeated vaso-occlusion and infarction over time, the spleen is usually absent in adult because of autosplenectomy. Splenic infarction leads to functional hyposplenism early in life, which in turn increases the risk of infections. Presence of functional spleen in the index case may have also contributed to reducing her chances of having crisis as infections have been reported to precipitate crises [12]. On the other hand, splenomegaly may also predispose her to the risk of splenic sequestration [13].

4. CONCLUSION

Patients with SCA usually present in infancy or early childhood with vaso-occlusive crisis in form of dactylitis, bone pain and/or symptomatic anemia which usually require frequent hospital admissions and blood transfusion especially in the absence of adequate care and follow up. This case is therefore to alert the health care providers to have high index of suspicion for late presentation in an atypical manner.

CONSENT

The author declared that written informed consent was obtained from the patient for publication of this paper.

ETHICAL APPROVAL

The author obtained ethical approval from the hospital Research and Ethics Committee.

COMPETING INTERESTS

Author has declared that no competing interests exist.

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