Available online at **www.ijmrhs.com**

ISSN No: 2319-5886

International Journal of Medical Research & Health Sciences, 2017, 6(7): 40-41

Sickle Beta Plus Thalassemia Presenting as Acute Pancreatitis – A Case Report

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ABSTRACT

Sickle beta plus thalassemia (Hb S/β+ Th) is a rare inherited disorder among haemoglobinopathies. This disorder is inherited in an autosomal recessive manner. Sickle beta plus thalassemia patient usually have chronic haemolytic anaemia and rarely can present as vaso-occlusive painful crisis. Abdominal pain is frequently seen in vaso-occlusive painful crisis and may masquerade diseases such as acute appendicitis and cholecystitis. Acute pancreatitis is rarely included as a cause of abdominal pain in these patients. There are no systematic literature available which can give insight about this complication and its management. Here we report a case of sickle beta plus thalassemia presenting as acute pancreatitis.

Keywords: Sickle beta plus thalassemia, Acute pancreatitis, Haemoglobinopathies

INTRODUCTION

Sickle cell beta thalassemia (Hb S/β Th) is an inherited form of sickle cell disease that affects red blood cells, both in the production of abnormal haemoglobin, as well as the decreased synthesis of beta globin chains [1]. Individuals with sickle cell beta thalassemia have one abnormal beta chain, βS, and a defective beta-globin gene, either in decreased synthesis, β+, or complete absence of synthesis, β0. Sickle beta thalassemia is inherited in an autosomal recessive manner. High HbF levels and the β-globin gene cluster haplotypes influence the clinical presentation of sickle cell disease. Most of the cases are from North Africa, India, and the Mediterranean region, especially Greece and Turkey. Sign and symptoms include anaemia (low levels of red blood cells), repeated infections, frequent episodes of pain, pulmonary hypertension, acute chest syndrome, stroke, enlarged spleen and/or liver, heart murmurs, delayed puberty, slowed growth, jaundice. Hydroxyurea, a drug approved by the U.S. Food and Drug Administration for the treatment of sickle cell disease, can decrease the frequency and severity of pain episodes; reduce the need for blood transfusions; and increase life span [2].

CASE REPORT

A 17 years old female patient presented with fever, pain in abdomen, vomiting, high coloured urine, and jaundice since 6 days. She was initially treated in medical college hospital and discharged with a diagnosis of pancreatitis with multi-organ failure.

Examination

Conscious, oriented, febrile, moderately dehydrated, pulse - 106/min, BP - 90/50 mm of Hg, pallor and icterus present. P/A - tenderness in per umbilical region, splenomegaly (4 cm), free fluid and peristaltic sounds present. Respiratory system-bilaterally decreased VBS, others - within normal limits.

Investigation

Hb – 4.2 gm%, WBC count – 7200/cu-mm (P-69, L-24, M-6, E-1), Platelet count – 92000/cu-mm, LDH – 2819 u/l, Indirect bilirubin – 2.8 mg/dl, Direct bilirubin – 0.9 mg/dl, Iron – 40 µg/dl, Ferritin – 432 ng/dl, TIBC – 197.0 µg/

dl, % saturation – 20.30%, PBS – RBC - microcytic hypochromic with moderate anisopoikilocytosis- eliptocytes, ovalocytes, and polychromatic cells; WBC – normal; platelets- reduced; reticulocyte count- 3.39%, haemoglobin HPLC-HbA0 – 6.7%, HbA2 – 5.3%, HbF – 12.8%, HbS – 70.7%; RBC – 1.62 million/cumm, Haematocrit – 11.4%, MCV – 71 cubicμ, MCH – 21.9 pg, MCHC – 31.1%, RDW – 17.0%. G6PD deficiency not detected, direct and indirect coombs test- negative, Amylase – 99 u/l, Lipase – 753 u/l, TSH – 1.67, creatinine, electrolytes and lipid profile – normal, hepatitis A/E/B/C – negative, HIV – negative, INR – 1.28, PT – 14.9s, aPTT -38.6s, SGOT – 110 u/l, SGPT – 87 u/l, urine R/E-normal, Throat swab for c/s – *Klebsiella* species, blood c/s- *pseudomonas* fluorescents, chest X-ray – normal, USG abdomen – GB wall oedema with minimal ascites & per nephric fluid, splenomegaly; CECT abdomen – diffuse GB wall oedema with moderate ascites, splenomegaly (170 mm), bilateral moderate pleural effusions, intra-splenic abscess (14 × 14 mm).

Diagnosis and management

Diagnosis is made on the basis of the clinical picture, laboratory investigation and electrophoresis as sickle beta plus thalassemia. She was managed over 3 units of blood transfusion, antibiotics as per the culture sensitivity, IV fluids, hydroxyurea, and folic acid.

DISCUSSION

Prevalence of the sickle beta plus thalassemia in eastern India [3] was found 0.26%. Sickle beta plus thalassemia patients usually have chronic haemolytic anaemia and rarely can present as vaso-occlusive painful crisis. The vascular occlusion is a complex process and accounts for the majority of the clinical manifestations of the disease. Abdominal pain is often seen in vaso-occlusive painful crisis and may masquerade diseases such as acute appendicitis and cholecystitis. Acute pancreatitis is rarely included as a cause of abdominal pain in these patients. In Sergeant's extensive review [4] of clinical feature of sickle cell anaemia, he states that the frequency of pancreatitis and its role in abdominal painful crisis are entirely unknown. One such case was reported [5] earlier, but there is no literature available about acute pancreatitis in sickle beta plus thalassemia. When it occurs it may result from biliary obstruction, but in other instances it might be a consequence of micro vessel occlusion causing ischemia. In this case we describe acute pancreatitis in patients with sickle beta plus thalassemia apparently due to microvascular occlusion and ischemic injury to the pancreas. Patient responded to conservative management.

CONCLUSION

Acute pancreatitis should be considered in the differential diagnosis of abdominal pain in patients with sickle beta plus thalassemia. Patient responds well with conservative management.

DECLARATIONS

Conflict of interest

None.

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