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AN UNUSUAL PRESENTATION OF SICKLE CELL ANAEMIA: PARALYTIC ILEUS

Medical Science

KEYWORDS :

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INTRODUCTION:

Vaso-occlussive phenomena and hemolysis are the clinical hallmarks of sickle cell disease(SCD), an inherited disorder due to homozygosity for the abnormal hemoglobin, hemoglobin S(HbS). Vaso-occlussion results in recurrent painful episodes and a variety of serious organ system complications that can lead to life-long disabilities and/or early death. Abdominal pain is a frequent symptom among patients undergoing sickle cell crisis. Gastro-intestinal disorders that may accompany sickle cell disease include sickle cell hepatopathy, splenic sequestration, paralytic ileus, cholecystitis and acute abdominal pain crisis. Although association has been stated between sickle cell anaemia and paralytic ileus, it is unusual, easily missed, easily misdiagnosed and not routinely taken into consideration. We report a case of 23 years old male patient came to our hospital with chief complains of breathlessness, fever, abdominal pain and minimal distension of abdomen.

CASE REPORT:

A 23 years old male patient came to our hospital with chief complains of breathlessness on exertion since 15 days. He also had complain of fever since 3 days which was low grade and intermittent associated with abdominal pain and minimal distension of abdomen, which was not relieved on taking local medicines. There was no complain of chest pain, palpitation, cough, back pain, joint pains, leg ulcers, bowel-bladder disturbances, vision loss, priapism, focal neurological defiect or other neuro-cognitive impairment. . On eliciting past history, from younger age he used to get repeated episodes of fever every 2-3 months and also become anaemic, for which he needed multiple blood transfusions. This continued till 18 years of age but since than though he has multiple episodes of fever, blood transfusion was never needed. He was never evaluated for this condition. There was no history of bleeding diathesis or anemia in family of the patient. Vitally patient was stable with systematic examination showing slight tenderness in epigastric region with hepatosplenomegaly, rest was normal. On investigating, he was found to be severly anemic with mild leucocytosis and periphereal smear showed RBC morphology predominantly microcytic hypochromic, marked hypochromia, marked anisopoikilocytosis with sickle cells present, most likely sickle cell anaemia. Renal function tests including electrolytes were found to be normal with mild elevation of indirect bilirubin. Retic count was found to be elevated(6%) with increased S.LDH(1409). All of this was suggestive of hemolytic anemia and keeping in mind the history given by the patient his sickling test was done which was positive and Hb electrophoresis showed compound heterozygous for HbS and Beta thalassemia. Ultrasonography was done which showed hepatosplenomegaly consistent with hemolytic anemia. His CECT abdomen was done which showed mildly dilated distal ileal loops with submucosal edema in terminal ileum likely due to paralytic ileus. He was treated symptomatically and comprehensively with intravenous fluids, blood transfusion, appropriate antibiotics, pain relief by analgesics and initially nil by mouth with naso-gastric decompression followed by enteral nutrition on clinical improvement . Patient improved and was discharged on hydroxyurea, folic acid supplementation and other supprotive medicines and advised for proper oral hydration and regular follow up. 2 years have passed and there is no single episode of sickle cell crisis and drugs are monitored.

DISCUSSION:

Sickle cell disease(SCD; this designation includes homozygosity for hemoglobin S, compound heterozygosity for hemoglobins S and C, compound heterozygosity for hemoglobin S and beta-thalassemia, plus other less common genotypes) is a typical Mendelian disorder. It is associated with hyoxia induced polymerization of the abnormal hemoglobin S molecule, followed by red blood cell sickling, with resultant macrovascular occlusion(vaso-occlussion) clinically manifested most often as an acute painful episode or "crisis", but with many other acute and chronic complication. Clinical manifestations vary markedly among the major genotypes. Acute pain is a first symptom in disease in more than 25% of patients and is the most frequent symptom after the age of 2 years(1). Acute pain is also the complication for which patients with sickle cell disease commonly seek medical attention(2). The frequency of the pain peaks between the ages of 19-39; more frequent pain

is associated with higher mortality rate in patients over age 19(3).

Pain will be precipitated by event such as weather conditions, dehydration, infection, stress, menses, alcohol consumption, nocturnal hypoxemia and rarely obstructive sleep apnea(4). However the majority of painful episodes have no identifiable cause. The episodes can affect any area of body, with the back, chest, extremities and abdomen being most commonly affected. Abdominal pain is mostly seen during vaso-occlusssive of infarctive crisis and per se may be difficult to distinguish from an acute surgical abdomen(eg, appendicitis, cholecystitis) and may be due to such conditions such as cholelithiasis, splenic infarction, pancreatitis, ischemic colitis, and non surgical genitourinary disorders and rarely paralytic ileus(5). There is no test to diagnose a vaso-occlussive event although presence of sickled form of RBCs on peripheral smear are seen. The first phase of the crisis may be charecterised by preferential trapping of deformable cells in the micro-circulation(6), these deformable cells may be more likely to adhere to vascular endothelium, leading to ischemia of the mesentry and thus leading to dilatation of the bowel loops and paralytic ileus. Although ileus has numerous causes(sepsis, drugs, metabolic, endocrine, trauma, neurosurgical procedures and many others), the post operative state is the most common setting, but association with sickle cell though theoretically stated is rarely documented. The optimal management of vaso-occlussive crisis is based upon high index of suspicion and assessment of volume status(oral hydration, or intravenous hydration if hypovolemic) and comprehensive treatment including infection control, nutrition maintainance and pain management strategies(intravenous opiate as initial therapy, rather than a non-opiod analgesic(7).

While defining causality due to vaso-occlusion remains an elusive goal, determining the contribution of specific event will establish the pathogenic importance and potential as targets of rational treatment of patients with sickle cell disease. Thus early suspicion and presumptive treatment may help in reducing the number of episodes of sickle cell crisis and ultimately improving the quality of life.

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