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Case Report

A Rare Case of Splenic Infarcts in Sickle Cell Anemia

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Abstract

Sickle cell disease (SCD) is a genetic disorder which causes debilitating systemic syndrome which is characterized by chronic anemia, acute painful episodes, organ infarction and chronic organ damage. Early screening, systemic clinical follow up, prevention of sepsis and organ damage have led to increased life expectancy among people with SCD.

Keywords: splenic infarct, sickle cell disease, hemolytic anemia

Introduction

Sickle cell anemia (SCA) is a rare genetic disorder, fewer than 1 million cases per year. Sickle cell hemoglobin is a structural variant of normal adult(HbA) caused due to mutation in Hbb gene that leads to substitution of valine for glutamic acid at position 6 of beta globin subunit (β^s) of hemoglobin molecule[1]. Sickle cell disease refers to any condition in which production of HbS leads to pathophysiological consequences. The most common form (>70% of SCD worldwide) [2] results from the homozygous inheritance of β^s mutation and is usually referred to as either SCD SS or as Sickle cell Anemia [3,4]. This rare hemolytic anemia must be considered as differential diagnosis in patients presenting with splenomegaly.

Case Report

A 20 year old male presented with complaints of fever and left hypochondriac pain for 15 days. On examination pallor was present with few prominent palpable cervical and axillary lymph nodes. Examination of the abdomen showed moderate splenomegaly with mild hepatomegaly. Investigations revealed anemia, thrombocytopenia and peripheral smear showed normocytic normochromic anemia with sickle cells and target cells and Howel Jolly bodies. Sickling test using 2% sodium metabisulphite showed sickle shaped RBC. Liver function test, coagulation profile, renal function test and electrolytes were within normal limits. HPLC analysis showed S window 77% suggestive of SCA. Contrast CT of the abdomen showed splenomegaly with ill defined non enhancing hypodense lesions suggestive of splenic infarcts. Under general Anaesthesia laparotomy with splenectomy and prophylactic cholecystectomy were performed for hypersplenism. Histopathological examination showed features of splenic infarct with changes of SCA. The patient received polyvalent vaccine at follow up.

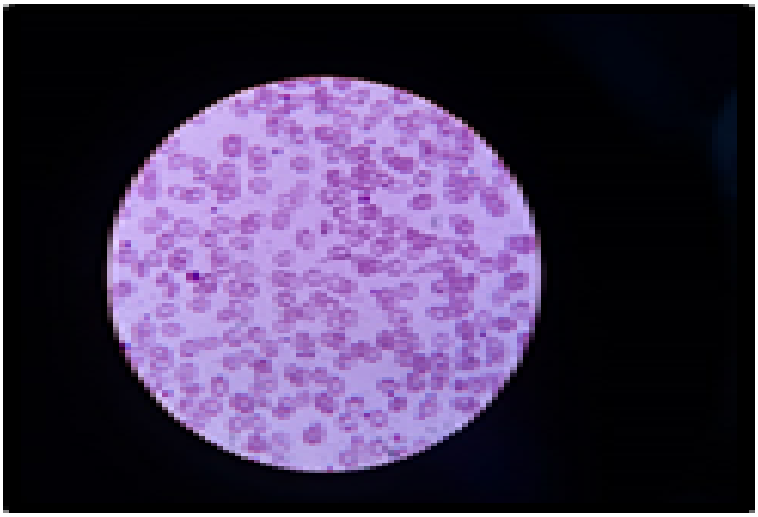

Figure 1a-*Peripheral Smear Showing Sickle Shaped Cells*

Figure 1b-*Sickling Test Showing Sickle Shaped Rbcs*

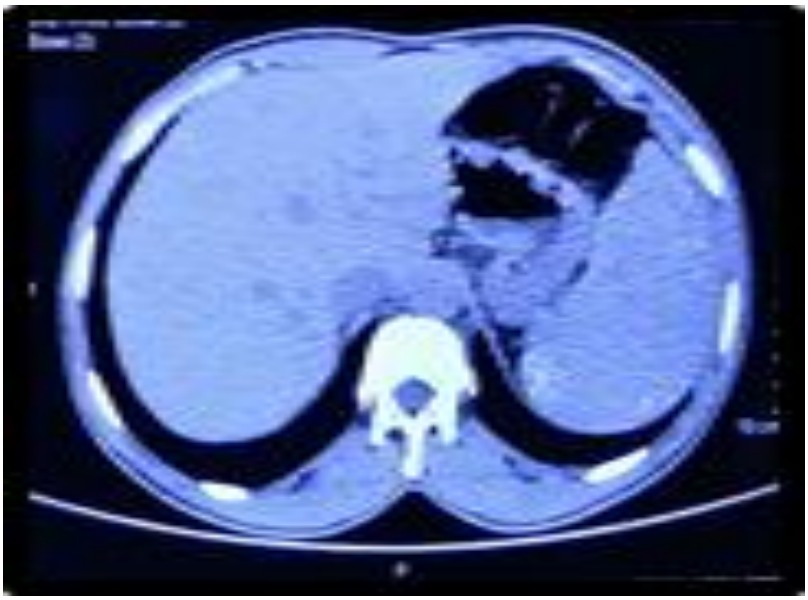

Figure 1c-*Cect Showingsplenomegaly with Ill Defined Non Enhancing Hypodense Lesion*

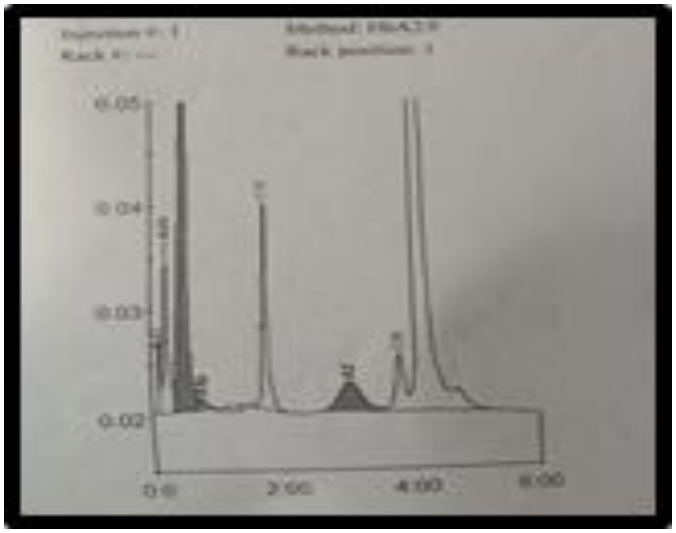

Figure 1d- *Hplc Showing S Shaped Window*

Figure 1: *Showing the Sickle Cell Disease*

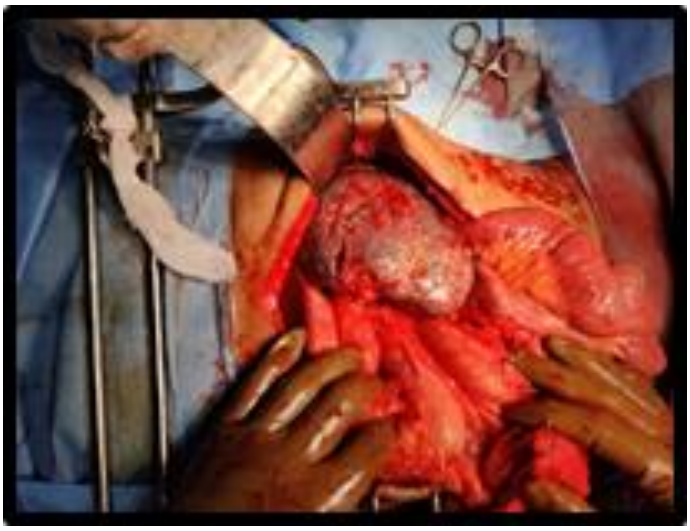

Figure 2a-*Operative Showing Splenic Infarct*

Figure 2b-*Excised Spleen*

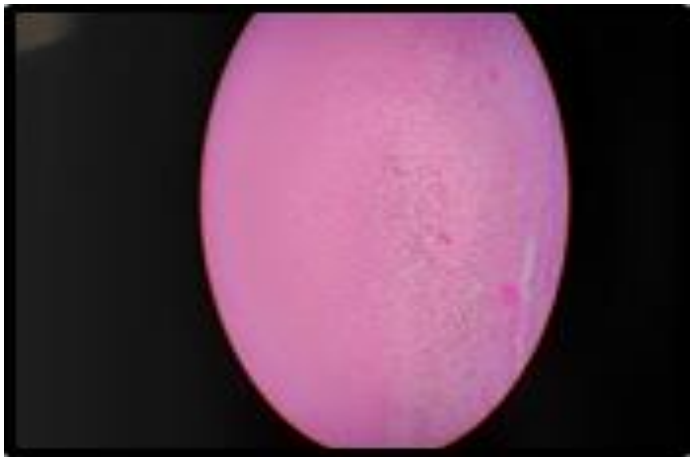

Figure 2c-*Histopathology of the Spleen Showing Congestion*

Figure 2: *Show Perop and Postop Images of Spleen*

Discussion

SCA occurs in 1-4% of population. Associated with aplastic crisis, acute sequestration crisis, hyperhemolytic crisis. Splenic infarct serve as nidus for abscess formation. Splenectomy helps in control of hemolytic anemia.SCA is common in Central India and migrant labours from Jharkand. Prevalence in Central India is about 22-44%.

Abdominal pain remains the leading chief complaint in patients diagnosed with splenic infarct. One of the most common cause of splenic infarct is sickle hemoglobinopathies. In patients with sickle cell disease hypoxic episodes, acidity and cellular dehydration causes polymerization of HbS within erythrocyte leading to their deformation into characteristic sickle shape. In dynamic interaction with the vascular endothelium, this sickling leads to episodic microvascular occlusion, ischemia and reperfusion, vascular and inflammatory stress and increased expression of vascular oxidases, inflammatory cytokines and adhesion molecules. Chronic hemolysis results in anemia, hypoxia, cholelithiasis, fatigue, exercise intolerability, hypercoagulability and vasculopathy which leads to endothelial nitric oxide depletion which causes pulmonary hypertension and ischemic strokes. SCA can affect any part of body but spleen is the most common and earliest organ to be affected. Spleen most commonly enlarges in first decade of life but then undergoes progressive atrophy as a result of repeated attacks of vaso-occlusion and infarction leading to autosplenectomy.

Crisis

Splenic sequestration crisis results from rapid sequestration of red blood cells in the spleen; characterized by sudden onset of anemia and splenomegaly. Treatment usually involves splenectomy. In sickle cell disease, infection with parvovirus causes decreased production of red blood cells leading to aplastic crisis causing sudden and severe anemia, fatigubility and shortness of breath. Blood transfusion is the main stay of treatment.

In Hyperhemolytic crisis, intravascular and extravascular hemolysis due to hypoxia leads to acute worsening anemia causing organ failure and death.Blood transfusion is the mainstay of treatment.Painful vasoocclusive crisis due to bony infarction presents in infants as dactylitis or bony infarction of digits, irritability and swelling of fingers or toes. Infarction can affect any bone or joint mimicking osteomyelitis [5].

Acute chest syndrome is characterized by intrapulmonary ischemia and infarction, systemic hypoxia and pulmonary infiltrates on chest radiography [6].

Standards of treatment include universal or targeted newborn screening programs, implementation of simple treatments such as vaccination and antibiotic prophylaxis, regular followup have led to reduction in early mortality from SCD.

Hydroxycarbamide or hydroxyurea is the only agent that has been proven to reduce painful episodic crisis, ACS and hospitalisations.

Transfusion therapy is mainly indicated for stroke risk reduction, renal failure or recurrent painful crisis that are less responsive to treatment with hydroxycarbamide.

Despite its benefits, chronic transfusion therapy can result in iron overload leading to organ damage, Alloimmunsation, transfusion acquired infections, venous access related issues such as thrombosis and sepsis. Allogenic hematopoietic stem cell transplant is the only curative treatment for SCD and is successful in 85-90% of patients [7]. Transplantation offers disease free survival and stabilization of neurological lesions. Gene therapy aims to abrogate SCD related symptoms by manipulation of hematopoietic stem cells, either by viral vector mediated insertion of functional β globin gene or by various gene editing techniques that reduce intracellular sickling by enhanced production of HbF [8].

Splenectomy

The indications for splenectomy were acute splenic sequestration crisis or Hypersplenism. Preoperative immunization with pneumococcal, meningococcal and H.influenza vaccines should be administered 10-14 days prior to surgery.

Conclusion

Splenic infarct in non infectious setting may be treated with analgesics, hydration and other means of supportive care. In sickle cell hemoglobinopathies treatment mainly involves correction of hypoxia and acidosis may be required. Splenic infarct causes dangerous complications including pseudocyst, abscess, hemorrhage, splenic rupture and aneurysm which warrant emergent surgical intervention.

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