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Case Report

Splenic abscess, a rare complication in a case of sickle cell thalassemia

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ABSTRACT

Sickle cell disease (SCD) is an inherited autosomal recessive haemoglobinopathy. Sickle cell beta thalassemia is a variant syndrome of SCD characterised by the compound heterozygosity for sickle and beta thalassemia genes. We present a case of 12 year old male child diagnosed case of sickle cell thalassemia at the age of 2 years with complaints of fever, yellowish discolouration of eyes and drowsiness. USG abdomen was done suggestive of splenomegaly, multiple ill-defined, heterogeneously hypo-echoic, areas scattered throughout the splenic parenchyma with no vascularity within likely representing as splenic micro-abscesses. Child was started on antibiotics covering anaerobic and gram-positive organisms (vancomycin, meropenem, amikacin and metronidazole). Currently child is doing well with huge relieve to his symptoms.

Keyword: Sickle cell anemia, SCD, Splenic micro abscesses

INTRODUCTION

Sickle cell disease (SCD) is an inherited autosomal recessive haemoglobinopathy, resulting from substitution of beta chain amino acid at valine residue, where glutamic acid is replaced by valine at the sixth amino acid position of the β-chain β6 (A3) glutamine valine.[1]Sickle cell beta thalassemia is a variant syndrome of SCD characterised by the compound heterozygosity for sickle and beta thalassemia genes .Based on decrease or complete absence of beta globin chain synthesis it is further classified into sickle cell beta^{+} thalassemia and sickle cell beta^{0} thalassemia. The symptoms in patients with sickle cell beta^{+} thalassemia are less frequent and less severe than those in patients with homozygous SCD or sickle cell beta^{0} thalassemia.[2,3] It is most common among people from Africa, India, the Caribbean, middle east and the mediterranean.

The SCD is most common among people from Africa, India, the Caribbean, Middle East, and Mediterranean.

Splenic abscesses, a complication of SCD are very rare in this disease accounting for 0.14% to a 0.7% of necropsy specimen as these patients tend to have early autosplenectomy.[3] The signs and symptoms of splenic abscess (fever, left quadrant pain and splenomegaly) are generally non-specific leading to delay in diagnosis and treatment with increased mortality.[4] With the recent advances and liberal use of diagnostic techniques such as ultrasonography and CT-scan, increase in early detection of splenic abscess have improved out-come, and emergence of other modalities of treatment for splenic abscess, other than splenectomy. We present a case of splenic abscess in a known case of sickle cell thalassemia.

CASE REPORT

We present a case of 12 year old male child born of third degree consanguineous marriage, a known case of sickle cell thalassemia. He was diagnosed at age of 2 years of age and since then was on regular follow up requiring transfusion once in a month and hydroxyurea. Child had stopped hydroxyurea since 20 days. He presented to us with history of high grade fever since 3 days, yellowish discolouration of eyes since 3 days, one episode of vomiting, loose stools and drowsiness. Examination

showed hepatosplenomegaly. At presentation, child was pale and had signs of congestive cardiac failure with tachycardia (HR-134/min), tachypnea (RR-40/min) and raised jugular venous pressure. He was drowsy with Glasgow coma scale of 13/15. His investigation revealed haemoglobin of 4.7 gm/dl (11.5-13.5), WBC of 22.3/UL (4.5-13.5), platelets of 98/UL (150-350). LFT showed indirect hyperbilirubinemia (total bilirubin of 3.1 mg/dl (<1.5 mg/dl), direct of 0.6 mg/dl (0.2 mg/dl), indirect of 2.5 mg/dl with liver enzymes SGOT of 1007U/L (15-40U/L) and SGPT OF 353U/L (10-55 U/L) which was elevated. Coagulation profile was deranged with (PT-47second (12.4-16.1), INR-1.8 seconds (0.97-1.30) and ApTT of >180 seconds (33.9-46 seconds. Hyponatremia, sodium of 123 mEq/L (125-135 mEq/L) explained the cause of drowsiness. Ammonia was raised with value of 129 mcg/dl (11-32 mcg/dl). Blood culture showed no growth. USG abdomen was done which was suggestive of splenomegaly, multiple ill defined, heterogeneously hypoechoic areas scattered throughout the splenic parenchyma showing no vascularity within representing splenic microabscesses in a known case of SCD. Child was shifted to injection meropenem, vancomycin, amikacin and metronidazole covering gram positive and anerobic organisms for which he responded very well. Child received transfusions for his anemia.

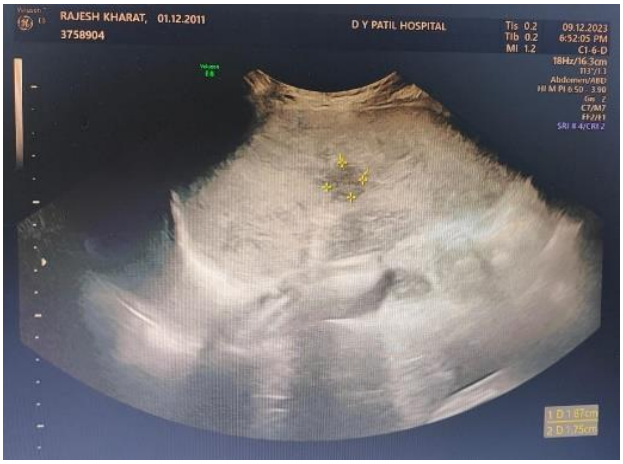

Figure 1: Heterogenous hypoechoeic area in splenic parenchyma.

DISCUSSION

Splenic abscesses are very rare in healthy individuals. It is usually seen in patients with underlying pathologic conditions, such as sepsis, splenic trauma with superimposed infection, spread from adjacent infectious process, or hemoglobinopathies. It is very rare in patients with SCA as these patients tend to have early auto-splenectomy. Persistence of splenomegaly in these patients however, predisposes them to the development of splenic infarction. Bacteremic infection from variety of sites is the most common cause of splenic abscess. Infective endocarditis has been identified as the leading cause to be associated with splenic abscesses.

Splenic abscess is a rare entity, with a reported frequency of 0.05-0.7%.[4] It reported mortality rate is still high, up to 47% and can potentially reach 100% among patients who do not receive the proper treatment. Splenic abscesses is a recognised complication in these children as there is a high incidence of splenic infarction that causes functional asplenia which in turn predisposes them to infection and bacteraemia. Thalassemia patients are at increased risk of infection due to defect in neutrophil chemotaxis and inability of phagocytes to kill microorganism due to iron overload making them vulnerable to infections by unusual organisms. The classical signs and symptoms of splenic abscess may not be specific always so high index of suspicion should be kept in mind for high risk patient groups. Many organisms are know to cause splenic abscess, *Streptococcus* the most commonest followed by *Staphylococcus, Klebsiella pneumoniae*, proteus mirabilis and gram negative bacilli (*Salmonella*).[4] Complications of splenic abscesses are life threatening which include perforation into peritoneum ,rupture into adjacent organs can occur with resulting fistulas into gastrointestinal tract, the plueral space or lung parenchyma.[3,5] Early supportive care and parenteral broad-spectrum antibiotics are importance in this patients.[7] Multiple small splenic abscesses are treated with intravenous antibiotics conservatively but due to high failure rate in this conservative treatment in splenic infarction and infection, it is always followed by splenectomy. Percutaneous drainage is indicated for easily accessible uni-loculated or bi-loculated abscess done under CT or Ultrasound guidance and local anesthesia and is reserved for patients with disease that contraindicated surgery or in young patients to avoid splenectomy.[6] Most of the studies advocate splenectomy and antibiotic coverage for children with SCA and splenic abscess as there is no point in preserving a nonfunctioning spleen which is present in the majority of them.

CONCLUSION

We are presenting this case to highlight the importance of early recognition, raising suspicion of complication and initiation of treatment in high-risk children. As these children are at higher risk for developing the complication, sequestration crisis and splenic infarction superimposed with infection lead to rare entity of gastric splenic fistula. It also highlights the need to give proper antibiotic coverage in these children to improve prognosis and deadly outcomes.

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