RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 7 | July 2015 | ISSN - 2249-555X

Autosplenectomyin Sickle Cell Anaemia-A Case Report

KEYWORDS: Sickle cell anaemia, autospleenectomy

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ABSTRACT *A 17 year old femalepatient of sickle cell anaemia since 3 years on ultrasonography revealed autosplenectomy. Patient presented with complaints of bony pain and pain in abdomen. Initial ultrasonography showed obscured spleen but the complaints were persistent hence ultrasonography was repeated which showed no spleen. NCCT A/P was done showing non visualisation of spleen. Hence confirmingautosplenectomy in a case of sickle cell anaemia a rare finding.*

Introduction:
The global prevalence of sickle cell disease and other data estimated that around 20-25 milllion individuals have homozygous sickle cell disease[1] The clinical manifestation of sickle cell anaemia is mainly due to repeated vaso-occlusion chronic intravascular hemolysis ,microvascular ischemia and organ damage. Spleenic complications are common as a consequence of progressive injury from repeated sickling of sickle haemoglobin in red blood cells. In early life splenomegaly has known to be common. The spleen is dark purple and soft with smooth surface with thin capsule. In later years however autosplenectomy commonly occurs,spleen becomes small wrinkled remnant often buried in adhesions which is a rare finding.

Autosplenectomy denotes spontaneous infarction of the spleen with resulting hyposplenism. Autosplenectomy is most frequently encountered in patients with homozygous sickle cell disease. Complete autosplenectomy is achieved by the age of 8. Radiographically spleen shrinks and calcifies as little as 1 cm in diameter. Ultrasound will either not be able to demonstrate spleen at all or identify a small irregular and shadowing nodule in the splenic bud but CT easily identifies the abnormally small and irregular remnant which is usually calcified[7]

Case report:
A 17 year old female came with complaints of pain in abdomen and bony tenderness since 15 days. She was diagnosed as sickle cell anaemia 3 years back. Her lab reports 3 months back when she was admitted in private hospital were sickling positive,HbA2-1.8%,HBS-90%,HBF-1.2% and ultrasonography abdomen pelvis showed mild hepatomegaly, spleen-partially obscured however revealed small size with heterogenous echotexture. Patient was treated and discharged when was symptomatically better. Patient presented again with complaints of fever and abdominal pain despite being on treatment. She was on tab hydroxyurea 500mg bd and tab folic acid 5 mg od. On examination she was hemodynamically stable, left hypochondriac tenderness was present. Liver was palpable 3cm below the costal margin and spleen was not palpable. There was no dull note in left hypochondriac region and no signs of fluid retention. Her present lab reports were as follows Hb-9.4,Tlc-6700,Platelets-2.4 lakh, renal function and liver function tests were within normal limits. Her present USG was suggestive of liver span 14.9cm,spleen not visualised. Finding was confirmed on NCCT A/P showing non visualisation of spleen.

Discussion:
Sickle cell disease was seen more common among people of African origin. There are three independent occurences of sickle cell gene in Africa. However a fourth independent occurrence of sickle cell mutation occurred in Asia and is shared by people of eastern province of Saudi Arabia and throughout central India and is known as Asian halotype.[1]Sickle cell disease in homozygous state has a significant contribution to the mortality and morbidity of this disorder during crisis.

In our study autosplenectomy was determined by non visualisation of spleen during ultrasonographic examination. This is consistent with the report by Bakhieta Ibrahim Atalla where autosplenectomy was detected in 43(47.8%) patients.[2]Similarly it was also established in reports from Jamaica and US where autosplenectomy was found in most adult patients with sickle cell disease.[3,4]In a prospective study conducted in GMC and HH Bhopal autosplenectomy was observed in 11%.[5] In a study of J Mohanty conducted in Orissa autosplenectomy was found in 8% of the patients thus concluding it a rare finding.[6]

Conclusion:
This study confirms the view of autosplenectomy in sickle cell anaemia. Therefore we advocate observation and avoidance of precipitating factors, nutritional, transfusion support, as well as use of hydroxyurea in management of these patients.

Volume : 5 | Issue : 7 | July 2015 | ISSN - 2249-555X

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