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IMAGING FEATURES OF AUTO – SPLENECTOMY IN A PATIENT WITH SICKLE CELL ANEMIA

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Abstract

Sickle cell disease is prevalent in many parts of the world, with around 20–25 million people being affected by it. The first case in India was found in the South in 1952, in the Nilgiri Hills. The clinical manifestations of sickle cell anemia are due to the vascular-occlusive crisis. Repeated infarction of the spleen can lead to splenic atrophy followed by grdaully progressive autosplenectomy. In this case report we describe the imaging features of autosplenectomy in a case of sickle cell anemia, which has been infrequently described in the literature.

Keywords: Vaso-Occlusive Crisis, Autosplenectomy, Sickle Cell Disease, Splenic Infarction

INTRODUCTION

Sickle cell disease is prevalent in many parts of the world, with around 20–25 million people being affected by the homozygous variant of this disease (1). It is also observed that at least half of this population is in the sub-Saharan African region. It is found to be the most common genetic abnormality among Nigerians (1,2).

The first case in India was found in the South in 1952, in the Nilgiri Hills (3,4). Since then, many cases have been identified in central India, as well as in north Kerala and Tamil Nadu. This gene is found to be prevalent in tribal communities, ranging from 1-40% (3,4).

The clinical manifestations of sickle cell anemia are due to the Vaso-occlusive crisis. Repeated attacks of such sickle cell crises lead to hemolysis, ischemia, and infarction of organs and even the bone (5,6). Repeated infarction of the spleen can lead to splenic atrophy , followed by autosplenectomy. The spleen is not absent in the literal sense in this case; it is merely wrinkled and non functional , the remnant of which is found deep within fibrous tissue and adhesions (6,7).

Patients with sickle cell anemia often complain of left upper quadrant pain, which is due to the splenic infarction. Functional asplenia has been reported and is defined as impaired splenic reticuloendothelial function .(8). This is associated with an increased risk of infection but is reversible with timely blood transfusions. Splenic calcification is yet another finding that is only found incidentally on imaging (9).

In this case report, we present the findings of a typical case of sickle cell crises who was incidentally found to have imaging features of autopsplenectomy.

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Case report

This is a case of a 19-year-old male patient who presented to the emergency department of our hospital with pain in the left leg and waist for the past 3 days. Patient was a known case of sickle cell anemia on treatment. The patient had multiple such previous episodes in the past, which were managed symptomatically.

On examination, the patient had a pulse of 90 beats per minute, a blood pressure of 92/60 mm Hg, respiratory rate of 12 cycles per minute, and room air saturation of 96%. Patient was afebrile at the time of examination. The patient was pale in appearance and poorly built and nourished. Systemic examination was unremarkable, except for mild tenderness in the left upper quadrant of the abdomen. There was no hepatosplenomegaly on examination. Examination of the skeletal system was grossly normal.

A working diagnosis of vaso-occlusive crisis was made, and patient was treated symptomatically for the same with IV fluids and antibiotics. Once stabilized, patient was evaluated further.

USG abdomen and pelvis was done, which revealed irregular shrunken spleen with echogenic curvilinear densities suggestive of calcification in the left hypochondrium . (figure 1)

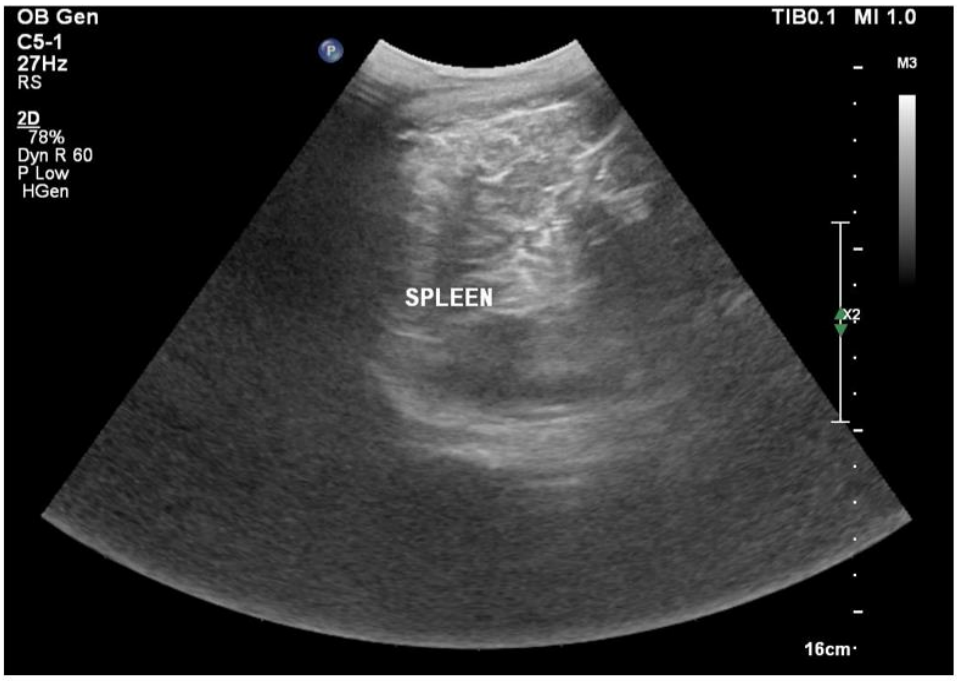

Figure 1: USG abdomen showing irregular shrunken spleen with echogenic curvilinear densities suggestive of calcification

Computed tomography of the abdomen and pelvis was performed, which confirmed the ultrasound findings- Plain CT images showed an irregular atrophied calcified spleen . No significant post contrast splenic parenchymal enhancement was seen on the post contrast images . (Figure 2 a, b) The splenic artery showed uniformly reduced calibre from its origin to the splenic hilum which suggests a long standing hypoperfusion of the spleen(Fig. 2c).

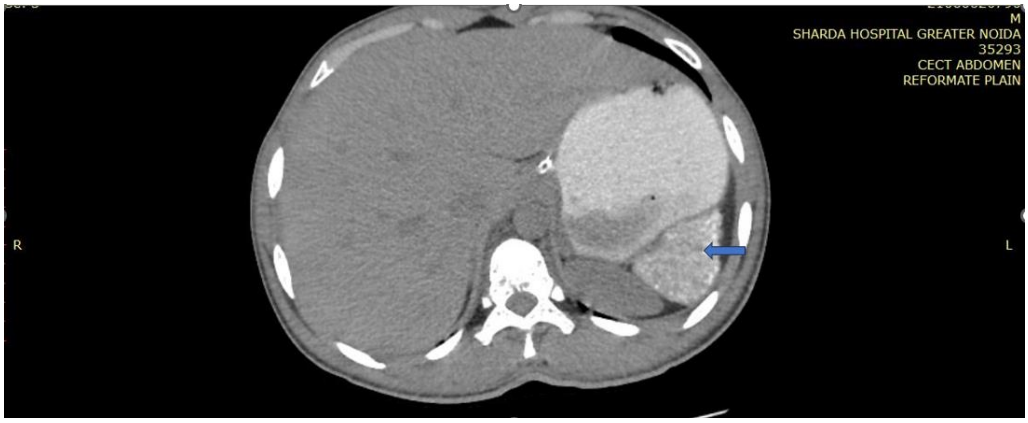

Figure 2a: NCCT Scan Abdomen (Axial Section) Shows irregular atrophied and calcified spleen in the left hypochondrium (blue arrow)

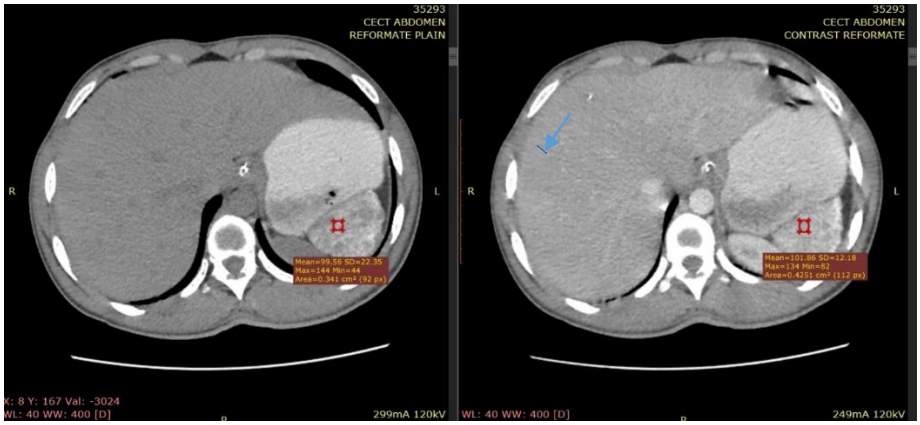

Figure 2b: CT abdomen (Axial section) pre contrast and post contrast images showing shrunken spleen with no significant post contrast enhancement

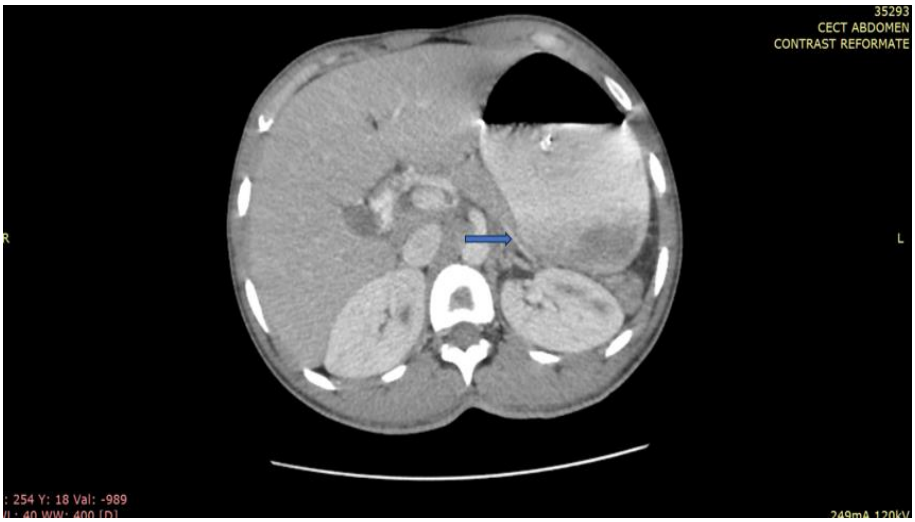

Figure 2c: CECT Abdomen (Axial Section) showing small sized splenic artery (blue arrow)

Patient was then treated symptomatically and received vaccination against capsulated organisms. Once the patient was clinically better, he was discharged. She is now on regular follow-up and receiving timely blood transfusions.

DISCUSSION

Autosplenectomy is an incidental, finding in patients with sickle cell anemia. It is seen more commonly in the homozygous variant of the disease rather than the sickle cell trait due to the increased frequency of sickling crisis in the homozygous variant. In a study done by A A Babadoko et al [(10),] 41 patients were found to have asplenia during ultrasound evaluation, while 23 patients had a shrunken spleen. In a study done by Bakieta et al [(11)], 47.8% of their study participants had autosplenectomy. In a study done by Watson et al and Rossi et al from Jamaica and the United States of America respectively, autosplenectomy was observed routinely in adult patients with sickle cell anemia. The imaging features of autosplenectomy have been infrequently described in the literature.

On Ultrasound imaging the spleen was found to be small sized and showed irregular margins . Multiple curvilinear echogenic foci were seen in the splenic parenchyma which represent splenic calcification . Plain CT images revealed a small sized and irregularly calcified splenic parenchyma . No significant parenchymal enhancement was seen on post contrast images which reflects the almost non existent splenic perfusion . Additionally , the splenic artery was of uniformly less diameter throughout its course (from its origin from the coeliac trunk to the splenic hilum) which suggests a long standing hypoperfusion of the spleen. Thus functional asplenia (autosplenectomy) can be reliably diagnosed on imaging studies as has been demonstrated in our case.

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