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Case Report

A rare case of multiple splenunculiin a patient with β-Thalassemia Major

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Abstract: Fifteen years old male child, a known case of beta-thalassemia major admitted for elective splenectomy because of massive splenomegaly, hypersplenism and increased requirement of blood transfusions. Per operatively, multiple splenunculi were seen over mesentery and along hepato-duodenal duodenal ligament. Both splenectomy as well as removal of all splenunculi was done. On follow-up, the patient is requiring fewer transfusions and is continuing with oral iron chelation therapy.

Keywords: Splenunculi, massive splenomegaly, β-thalassemia major, splenectomy

INTRODUCTION

Ectopic splenic tissue can be categorized into splenunculi and splenosis. Splenunculi is also known as an accessory spleen, supernumerary spleen, or splenules. These are congenital foci of healthy splenic tissues that are found apart from the main body of the spleen due to abnormal deposition of some of the cells from the developing spleen along the path of its formation during embryonic life. [1] Whereas splenosis is caused by auto transplantation of viable splenic tissue in the abdominal cavity. They typically appear as numerous small nodules. It usually occurs after splenectomy and trauma. [2] This report illustrates an unusual case of a 15 years old child with β-thalassemia major who was found to have multiple splenunculi during splenectomy for massive splenomegaly.

CASE REPORT

A 15 years old male child, known case of thalassemia and product of non-consanguineous marriage presented to the Department of Surgery for elective splenectomy due to massive splenomegaly and increasing blood transfusion requirements. The patient had presented with progressive abdominal distension, generalised malaise and fatigue at the age of 1year. Investigations then revealed that patient had beta-thalassemia major. Patient was receiving repeated blood transfusions (BT) since then. The requirement has progressively increased and now he has been receiving it every two weeks. His elder sib also had splenectomy 2 years back at the age of 10 years.

On examination, patient had chipmunk facies, severe pallor and mild icterus. Abdominal examination revealed splenomegaly of 15 cm below left costal margin and reaching up to the umbilicus. Ultrasonography (USG) confirmed the massive splenomegaly. Preoperative vaccination against Streptococcus pneumoniae, Neisseria meningitidis and Hemophilus influenzae B was administered 2 weeks before surgery.

Splenectomy was done after left subcostal incision. Spleen was 22×12×6 cm **(Fig 1),** smooth surface and grey brown in colour with congestion. Multiple splenunculi were seen over mesentery and along hepato-duodenal duodenal ligament **(Fig 2).** Splenunculi over spleen was 5x3×2.5 cm **(Fig. 3)** and reddish brown in colour. Histopathological examination revealed splenunculi having similar architecture as that of the spleen. Post-operative period was uneventful. On follow-up, the patient is requiring fewer transfusions and is continuing with oral iron chelation therapy **(Fig. 4)**.

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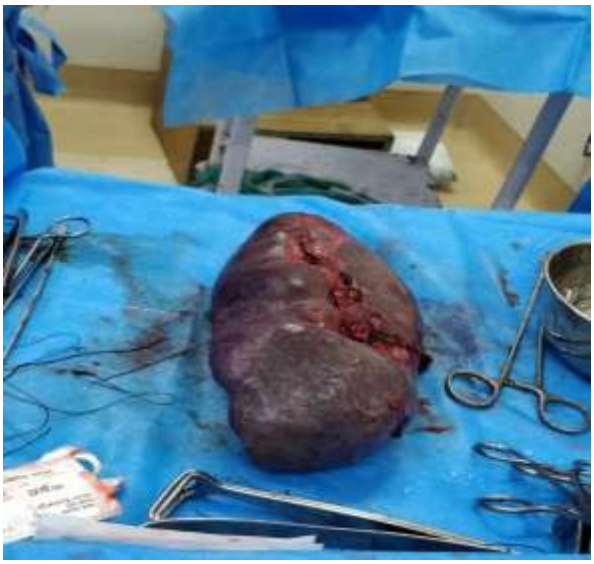

Figure 1: Massive Splenomegaly of size 22×12×6 cm

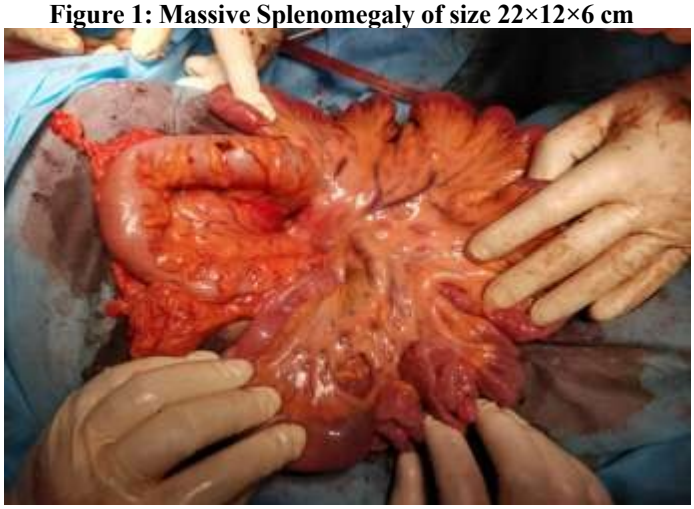

Figure 2: Multiple splenunculi, reddish brown in colour seen over mesentery and along hepato-duodenal ligament

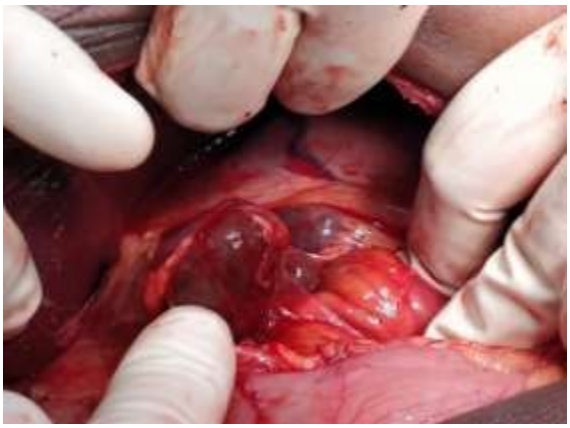

Figure 3: Largest splenunculi 5x3x2.5cm

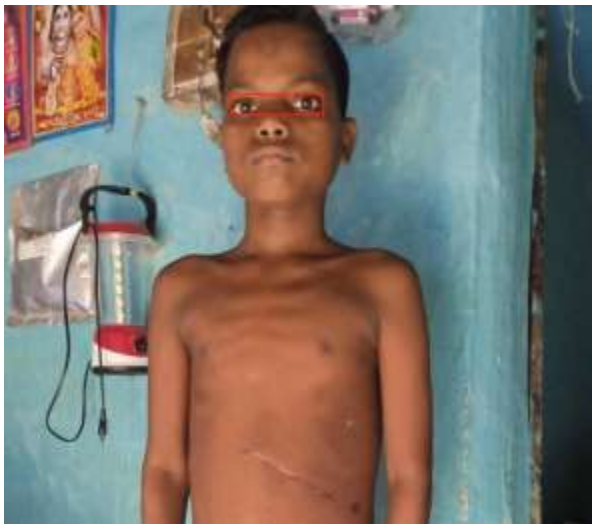

Figure 4: Chipmunk facies of patient (post-splenectomy)

DISCUSSION

Ectopic splenic tissue splenunculi and splenosis are different and hence, cannot be used interchangeably. Splenunculi are morphologically as well as functionally same structural elements as the main spleen. These are of congenital origin and have arterial blood supply from the splenic artery. [6,7] Splenunculi are usually seen near the hilum of the main spleen. However, they may be present anywhere in the peritoneal cavity- in either small and large bowel mesentery, pancreaticosplenic ligament, splenocolic ligament, gastrocolic ligament, greater curvature of stomach, greater omentum,tail of pancreas,retroperitoneal space and in the scrotum very rarely.[5,8,9] The splenunculi have normal splenic histology. However, in splenosis histology is different. It is of varied size and shape, has a capsule which is poorly formed. It does not have any hilum and the architecture is usually distorted. [2]

Splenunculi are usually asymptomatic and discovered incidentally during splenectomy. However, they can clinically present as acute abdomen due to their torsion, cyst formation, spontaneous rupture or hemorrhage and as intraperitoneal inflammatory mass. It may be wrongly implicated as neoplastic mass arising from the tail of the pancreas, gonads, adrenal gland, kidney, gastrointestinal tract or confused as enlarged lymph nodes or abdominal mass during radiological imaging.

In Beta – thalassemia cases, with poor compliance of blood transfusion and iron chelation, the children could develop massive splenomegaly and hypersplenism. Elective splenectomy is advised in such cases if the blood transfusions (packed red blood cell) requirement is more than 220 mL/kg/year or there is evidence of hypersplenism in the form of leucopenia or thrombocytopenia or when there is presence of massive splenomegaly leading to persistent abdominal discomfort. [3] Splenectomy is also advised if reduced RBC survival with splenic sequestration is demonstrated on Cr51 radioisotope studies.

It is very important to identify splenunculi before elective splenectomy because these may enlarge after splenectomy as they become locations of compensatory hematopoeisis and also later result in the recurrence of hemolysis in hemolytic anemia. [10] Ultrasonography (USG) may be used to identify splenunculi and they appear as hypoechoic round mass. However, they may be missed by USGs also. Computed tomography evaluation is a better modality to locate splenunculi. These can be seen in 11.0%–18.8% of cases. Large number of splenunculi are seen in 14.28% of cases. [4]

Hence, thorough search for splenunculi is mandatory during splenectomy for massive splenomegaly in hemolytic anemias. The aim is to remove all functional splenic tissue in such cases so that they don't further worsen the hemolysis in hemolytic anemia. [5,11]

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