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Case report: Splenic hamartoma: A rare vascular benign tumor of spleen

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Abstract
Splenic hamartoma (or splenoma) is a rare, benign, vascular tumor, often incidentally found at imaging, surgery or autopsy. It is usually asymptomatic. We report a case of 53 yrs old female from Tumkur city which was incidentally found during evaluation for GERD who had a large (9*10*10) splenic mass with small cystic /necrotic areas arising from lower pole of spleen, moderate heterogeneous enhancement on post contrast study, histologically congested sinusoids along with white pulp comprised of lymphoid follicles and immunohistochemically positive for antibodies to T- and B-cells and immunoglobulin light chains, CD 34 and Factor 8.

Keywords: Spleen, hamartoma, sinusoids, factor 8

Introduction
Splenic hamartomas are accidentally detected benign tumors of spleen which was first described by Rokitansky [1] but recent advances in imaging techniques have allowed the preoperative detection of splenic tumors [2]. Splenic hamartoma are also known as splenoma, splenoadenoma, and nodular hyperplasia of the spleen. The incidence of splenic hamartomas in autopsy series ranges from 0.024% to 0.13 % [3]. Lesions which are large present clinically as splenomegaly, pain, rupture, and rarely with thrombocytopenia and anemia [4]. Splenic hamartomas may be associated with other conditions like tuberous sclerosis, Wiskott-Aldrich like syndrome, and other neoplastic conditions [5, 6]. Splenic hamartoma is benign but it is very important to differentiate it from malignant tumors including metastatic tumors [7, 8].
With this background we report a rare case of splenic hamartoma found incidentally while evaluating GERD in a 53-year-old woman.

Case Presentation
A 53 yrs old female from Tumkur city presented to surgery OPD of our hospital with chief complaint of mass in the left upper abdomen which was incidentally found during evaluation for GERD. She also gave history of loss of weight of 10kgs in 1year. Per abdomen examination revealed intraabdominal intraperitoneal mass of 9*10*10 cm in the left upper quadrant which was non tender. All the borders well made out except superior border. Splenic dullness continued with mass dullness. There were no associated co morbidities. By history and clinical examination we conclude that the mass arising from the spleen. Routine investigations like hemogram, renal function test, and liver function test and coagulation profile were essentially normal.
Ultrasound abdomen showed a well defined oval heterogeneously hyper echoic lesion of 80*51 mm arising from inferior pole of spleen, feeding vessels were seen arising from splenic artery and its intraparenchymal branches. Significant vascularity was present. Features were suggestive of low grade malignant tumor or long standing benign soft tissue lesion.

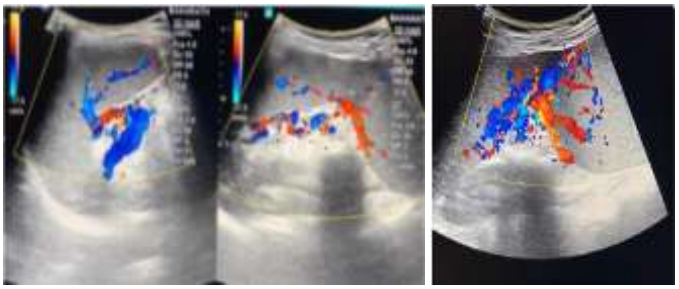

Fig 1: The abdominal ultrasound image (a) showed a round mixed echogenicity mass in the spleen, in which inhomogeneous echogenicity with patchy hyperechoic and iso-echoic foci were noted. Internal color Doppler flow was also observed.

Contrast enhanced computerized tomogram (CECT) abdomen showed exophytic heterogeneous solid mass with small cystic /necrotic areas measuring 9*8 cm arising from lower pole of spleen, moderate heterogeneous enhancement on post contrast study. Features were suggestive of neoplastic mass of spleen mostly splenic sarcoma.

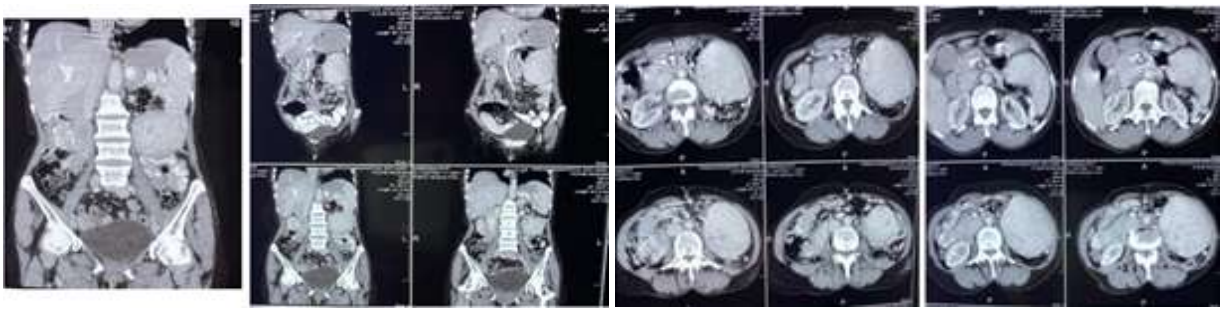

Fig 2: Abdominal computed tomography scan revealing a homogeneous round splenic mass with rim enhancement.

Fine needle aspiration cytology (FNAC) was not done due to hyper vascularity of the mass.

Splenectomy was planned under general anesthesia. Intra operative findings showed smooth globular mass arising from inferior pole of spleen. Excision of the mass was planned but due to massive hemorrhage splenectomy was done. Resected spleen was 10.5*7*3cm with external surface showing intact capsule. Hilum was unremarkable and the cut surface showed congested areas.

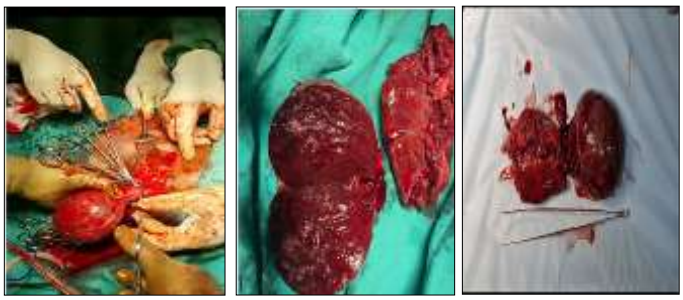

Fig 3: a) Intraoperative findings shows smooth globular mass arising from inferior pole of spleen b) A well-defined homogeneous red-tan mass can be seen on the cut surface compressing the adjacent normal splenic parenchyma c) A well-defined homogeneous red-tan mass can be seen on the cut surface compressing the adjacent normal splenic parenchyma.

Histopthological examination showed

Spleen - Microscopy-congested sinusoids along with white pulp comprised of lymphoid follicles.

Mass – Microscopy –globular mass show partial effacement of splenic structure comprised of varying sized vascular channels lined by plump endothelial cells almost replacing white pulp, without atypia.

The final pathological diagnosis was benign vascular lesion splenic hamartoma.

Immunohistochemical studies, with antibodies to T- and B-cells and immunoglobulin light chains, showed **CD** 34 and Factor 8 positive.

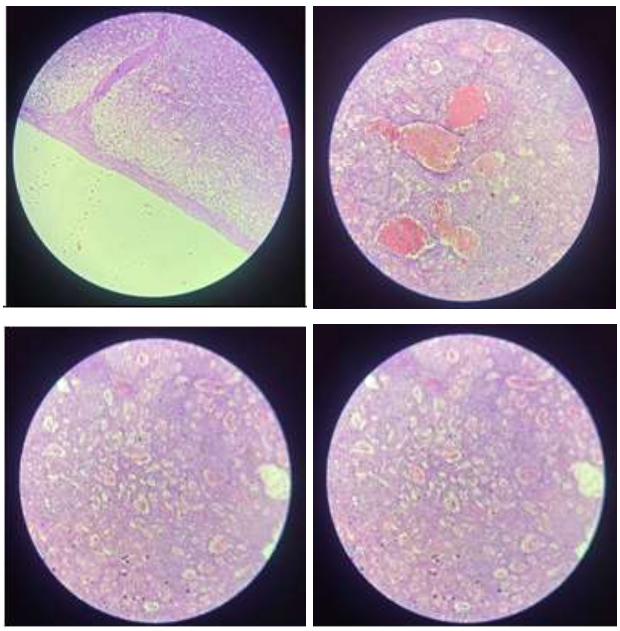

Fig 4: a) Low-power view showing the lesion (right field) composed of unorganized sinusoid-like channels without malpighian corpuscles, less well demarcated from the adjacent normal splenic tissue (left field) (H&E, × 100). c In some areas, the cavernous vascular channels were filled with erythrocytes and foci of fat vacuoles can be noted (H&E, × 100). d-f Many bizarre large cells are scattered in the stroma throughout the lesion, with oval, reniform, multilobulated, or convoluted nuclei. The chromatin is pale, granular or vesicular. Sometimes nuclear grooves (e, arrow) are present. Note large cells (d, arrows) with double nuclei and apparent eosinophilic nucleoli mimicking Reed-Sternberg cells in classical Hodgkin's lymphoma. (H&E, × 400)

Discussion

Splenic hamartoma is a rare benign lesion of spleen often seen in adult patients of an average age of 40–50 years but some of the case reports shown that they can occur even in children. Although there is no data on the incidence of splenic hamartomas but in autopsy studies, an incidence of 0.024-0.13% has been reported and an incidence of 0.015%-2.7% was noted among the patients who had undergone a splenectomy[9]. The lesions vary in size ranging from a few millimeters to 20 cm maximum and in our case it was around 10 cm.

Clinical presentation depend on the size of the hamartoma and common presenting clinical features are splenomegaly, anemia, palpable mass, thrombocytopenia and digestive symptoms, including loss of appetite and abdominal pain [7, 10]. In our case the presenting complaint was GERD and palpable mass. Hematological disorders are more common among children [9].

The common differential diagnoses of splenic hamartoma are atypical vascular tumors like hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma and angiosarcoma [8].

Preoperative diagnosis of splenic hamartoma is very difficult as malignant counterpart appears similar, and the diagnosis has to be confirmed by histopathology. However recent progress in imaging procedures has changed the management approach of splenic hamartomas. Abdominal Ultrasound is usually the first radiological method of investigation. Doppler US, CT, and magnetic resonance (MR) are advanced methods for diagnosis. Doppler US helps to distinguish splenic hamartomas from other differential diagnosis [11]. Splenic hamartoma is suspected when findings of increased blood flow on color Doppler images are seen in association with a homogeneous solid echogenic mass[9]. On ultrasound, hamartomas are well-defined homogenous hyperechoic lesions that shows increased blood flow demonstrated with Doppler. Secondary changes like hemorrhage, degeneration, and calcification makes these lesions heterogenous on ultrasound [12]. In our case ultrasound abdomen showed a well defined oval heterogeneously hyperechoic lesion of 8*5 cm arising from inferior pole of spleen, feeding vessels were seen arising from splenic artery and its intraparenchymal branches. Significant vascularity was present.

Plain CT abdomen shows hamartomas as isodense and in contrast they may appear as heterogenous [12]. In our case Contrast Enhanced Computerized Tomogram (CECT) abdomen showed exophytic heterogeneous solid mass with small cystic /necrotic areas measuring 9*8 cm arising from lower pole of spleen, moderate heterogeneous enhancement on post contrast study.

Surgical procedures for splenic hamartoma include splenectomy, partial splenectomy, laparoscopic splenectomy, and hand-assisted laparoscopic splenectomy [12]. In our case splenectomy was done.

Though imaging studies helps in predicting the diagnosis, but exact diagnosis is often made by histopathological examination of the mass. Histologically, splenic hamartoma are characterized by unorganized sinusoid-like channels without interspersed white pulp. The key immunohistochemical feature for splenic hamartoma is the lining cells of sinusoid-like channels are CD8-positive [13]. In our case histopathology showed congested sinusoids along with white pulp comprised of lymphoid follicles and vascular channels lined by plump endothelial cells almost replacing white pulp, without atypia. Immunohistochemical studies, showed CD 34 and Factor 8 positive.

Conclusion: Splenic hamartomas are rare benign vascular lesion with the characteristic CD8 positive immunophenotype of the lining endothelial cells. Though rare, it must be included in the differential diagnosis of splenic mass-forming lesions. An elective splenectomy should be performed to make a definitive diagnosis by histopathological examination.

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