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Original Research Paper

General Surgery

A RARE CASE OF ADULT SPLENIC LYMPHANGIOMA MASQUERADING AS A HYDATID CYST

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ABSTRACT Adult lymphangiomas of spleen are exceedingly rare neoplasms of spleen. They commonly present in childhood under the age of 2 years of age with no sexual preponderance. This case report highlights the management of splenic lymphangioma in a 30 years old female presenting with vague upper abdominal pain for the past 1 month. Imaging by computed tomography revealed multiple lobulated multiseptated peripherally enhancing lesion (HU-12) with few specks of calcification noted in the spleen largest of size 6.2 x 8.3cm. Possibility of splenic hydatid cyst in background of splenomegaly. Laparoscopic splenectomy was performed after proper preoperative preparation of the patient. Histopathological examination was reported as splenic lymphangioma. This case report highlights the need to differentiate among different cystic lesions of spleen for proper management and better patient care.

KEYWORDS : lymphangioma; splenic tumours; splenic cyst, laparoscopic splenectomy

INTRODUCTION:

Splenic lymphangiomas are extremely rare benign tumours constituting <0.007% of all splenic neoplasms. From the year 1970 to 2017, only 200 cases of splenic lymphangiomas were reported worldwide [1]. Here we report a case of symptomatic splenic lymphangioma in a 30 years female who underwent laparoscopic splenectomy.

Case Report:

A 30 years old female presented with complaints of vague upper abdominal pain for the past 1month. The pain is confined to left upper abdomen- dull aching, intermittent, non-radiating and has no relation to food intake. No history of fever, vomiting or abdominal distension. No history of coffee ground vomitus or black tarry stools. No history of breathlessness, cough with expectoration or chest pain. No history of yellowish discolouration of eyes, clay-coloured stools or generalized itching. No history of abdominal trauma in the recent past. No history of loss of appetite or weight loss. No known comorbidities. History of irregular menstrual cycles with decreased flow for the past 5 years. Patient is currently undergoing ovulation induction therapy and IVF for primary infertility. History of two cycles of failed IVF in the past 1 year. No history of any surgeries in the past. No significant family history.

On examination, vitals were stable. Examination of abdomen did not reveal any specific findings on inspection. On palpation, there was no tenderness/ guarding/ palpable organomegaly. Shifting dullness was absent. Per-vaginal and per-rectal examination were normal.

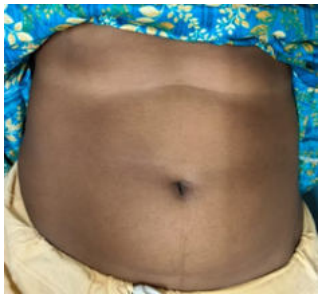

Fig 1: Clinical photograph of abdomen showing no findings on inspection.

Blood investigations were within normal limits. Haemoglobin was 12 gm/dl. Peripheral smear was normal. USG abdomen revealed 11.5cm multiloculated anechoic cyst with internal septations and posterior acoustic enhancement noted in splenic parenchyma adjacent to splenic hilum.

CECT-Abdomen (IV contrast) showed evidence of multiple lobulated multiseptated peripherally enhancing lesion (HU-12) with few specks of calcification noted in the spleen largest of size 6.2 x 8.3cm. Splenic index-630. Possibility of splenic hydatid cyst in background of splenomegaly. The cyst is seen abutting the left dome of diaphragm superiorly.

Patient was prepared for splenectomy. Differential diagnoses were Hydatid cyst disease, cystic lymphangioma of spleen, haemangioma and splenic pseudocyst. One dose of pneumococcal, meningococcal, and pentavalent vaccines was administered two weeks' time before surgery as per protocol. Third generation cephalosporins were started. Laparoscopic splenectomy was performed.

Intraoperatively patient was placed in a supine position with left sided elevation in reverse-Trendelenburg position. Pneumoperitoneum was created using closed method by Veress needle. Port placement was planned as follows. A 10mm camera port placed at umbilicus. Two 10mm working port placed in epigastrium and left anterior axillary line at level of umbilicus following baseball diamond concept. Another 5mmm working port was placed in left midclavicular line at the level of umbilicus. The phrenicocolic ligament was divided to exposure the lower pole of spleen. Lesser sac was entered and splenic hilum visualized by dividing greater omentum and short gastric vessels using Enseal device. Splenic artery and vein dissected, doubly clipped and ligated. Tail of pancreas protected and phrenicosplenic ligament divided. Specimen was retrieved via mini-laparotomy incision via epigastric 10mm port. Strict haemostasis was achieved. Drain tube was placed in splenic bed before rectus sheath closure with 1-prolene. The total duration of procedure was 200 minutes and the estimated blood loss was approximately 75 millilitres.

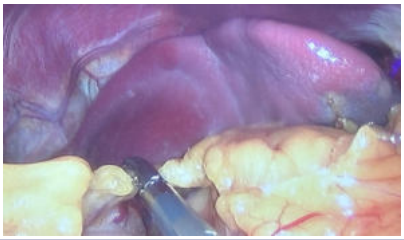

Fig 2: Intraoperative photograph showing division of greater omentum and short gastric vessels to exposure splenic hilum

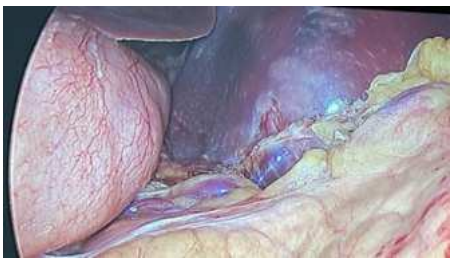

Fig 3: Intraoperative photograph showing tortuous splenic vessels leading to hilum of spleen in lesser sac.

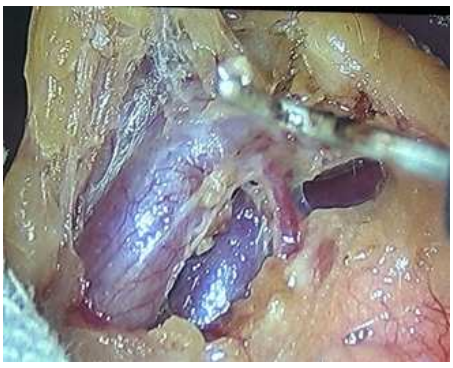

Fig 4: Intraoperative photograph showing dissection of splenic vessels at hilum.

Gross examination of specimen revealed a cyst of size approximately 9 x7 cm (cut-open) and adjacent cyst of size 5 x 5 cm with a wall-thickness of 1-2mm. The cyst is filled with gelatinous material.

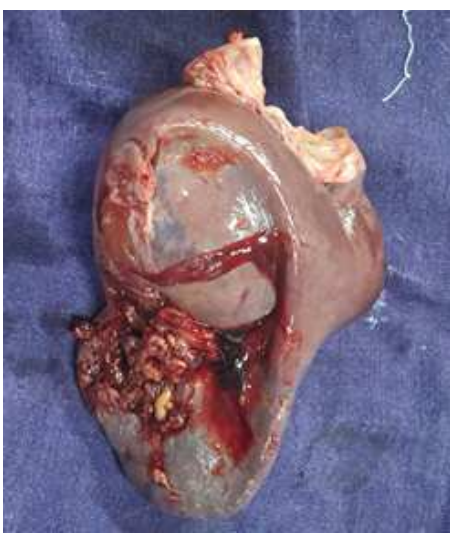

Fig5: Gross specimen of spleen showing multiple cysts of varying sizes.

Postoperative course in hospital: Patient was started on sips of fluid on the 1st post-operative day and soft solid diet on 3rd post-operative day. Surgical wound was healthy with no features of erythema, induration or discharge. Patient did not report any complaints and was discharged on the 5th post-postoperative day. Follow-up in outpatient services were uneventful.

HPE report-Section studied shows splenic parenchyma showing congestion with lesion composed of variably lined spaces lined by flattened endothelial cells filled with eosinophilic fluid material. Intervening stroma shows hemosiderin laden macrophages and areas of haemorrhage. Impression- lymphangioma of spleen.

DISCUSSION:

Lymphangiomas of spleen account for only less than 0.007% of splenic neoplasms in humans. Their aetiology is attributed to the congenital sequestration of lymphatic vessels which are disconnected from the normal lymphatic system. The underlying factors are lymphatic agenesis or obstruction due to inflammation. The majority of lymphangiomas occur in head and neck region (75%) as cystic hygroma presenting in childhood under 2 years of age. The common intra-abdominal sites for lymphangioma are retroperitoneum and mesentery. Less commonly they can present in mediastinum, bone or other intraabdominal organs like liver, kidneys, and pancreas. They can present as isolated lymphangiomas or as a part of systemic lymphangiomatosis syndromes [2,3].

Lymphangiomas are classified into benign vascular cystic neoplasms of the spleen. Other benign neoplasms of spleen are haemangioma, hamartoma, inflammatory pseudotumor and fibromas. Non-Hodgkin lymphoma, metastasis and hemangiosarcoma are classified into malignant splenic neoplasms. These cystic neoplasms are confused for pseudocysts of pancreas(most common variety of pancreatic cysts) and parasitic cysts due to hydatid disease [2].

Radiologically, these cystic lymphangiomas appear in ultrasound as hypoechoic lesions and computed tomography as hypodense cystic lesions. They present as multiloculated, multiseptated cysts of varying sizes predominantly in subcapsular region of spleen. They might rarely present with peripheral rim calcification. In magnetic resonance imaging, these multiloculated cysts possess simple fluid intensity with exception of haemorrhagic cysts/ cysts with proteinaceous debris [4]. Lymphangiomas appear as avascular non-enhancing lesions with the characteristic "Swiss-cheese pattern" on angiography. Preoperative tissue biopsy is contraindicated in view of potential risk of bleeding and inadequate cellular smears in cystic lesions [3].

On histopathological examination, lymphangiomas can be differentiated into capillary, cystic or cavernous types. Cystic lymphangiomas (commonest variety) appear as multiple cystic spaces filled with acellular eosinophilic proteinaceous substance lined by flattened endothelial cells. The background splenic parenchyma might be normal or appear as fibrosed, congested or infiltrated by leucocytes. The lining endothelium is reactive to endothelial markers like CD-31, CD-34, vascular endothelial growth factor receptor-3 and Factor VIII on immunohistochemistry. haemangiomas can be differentiated from lymphangiomas by reactivity to podoplanin- specific lymphatic endothelial marker [5].

The potential complications that are encountered in an untreated cystic lymphangioma are haemorrhage into the cyst, splenic rupture, torsion of cyst, secondary infection resulting in splenic abscess and intestinal obstruction. The surgical options available are total or partial splenectomy via laparoscopic or open approach. Open surgical approach is undertaken when moderate or massive splenomegaly is encountered. However, partial splenectomy is associated with 9.5% risk of development of recurrent lymphangioma from the residual spleen [6].

Conflict Of Interest: None

Informed Consent: Informed consent was obtained for publication of case report and clinical photographs of the patient.

CONCLUSION:

This case report outlines the management of a rare adult cystic neoplasm. The importance lies in differentiating cystic

lymphangiomas from other cystic lesions of spleen. Splenectomy helps to avoids complications of lymphangioma and provides tissue specimen for definitive histopathological examination.

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