ORIGINAL RESEARCH PAPER

General Surgery

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A RARE CASE OF SPLENIC LYMPHANGIOMA IN ADULT PRESENTING WITH ANEMIA

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ABSTRACT

BACKGROUND: Lymphangiomas are uncommon benign lesions of the lymphatic vessels, very rarely affecting the spleen. Isolated involvement of the spleen is rare. Fewer than 100 cases of splenic lymphangiomas have been reported in literature.
CASE REPORT: 68 year old female, with generalised weakness. Physical examination revealed anemia. Abdominal CECT scan showed two well defined cystic lesions of the spleen. Splenectomy was done and histopathological analysis led to the diagnosis of splenic lymphangioma. Postoperative period was uneventful and she recovered well.
DISCUSSION: This pathology is very rare, diagnosed incidentally before the age of 2, with presentation in adults being extremely rare. They are generally asymptomatic, or can cause splenomegaly.
CONCLUSION: Isolated splenic lymphangioma in adult patients is very rare. The preoperative diagnosis is challenging because imaging techniques are not specific. Pathological analysis is the only effective way to render the definitive diagnosis. It has a benign course after complete surgical resection.

INTRODUCTION

Lymphangiomas are benign tumours of the lymphatic system, occurring as congenital malformations of the same, in which obstruction or agenesis of lymphatic tissue causes a lymphangiectasia secondary to the absence of normal communication between the lymph ducts, which terminate in a cul-de-sac and slowly dilate until a cyst is formed [1,2], presenting predominantly in children, very rarely affecting adults [3], either diagnosed incidentally or sometimes presenting with abdominal complaints such as pain abdomen, abdominal mass or increasing abdominal girth [4]. They are generally located in the neck most commonly, second most commonly in the axilla and less commonly in other regions including the orbit, mediastinum, adrenal gland, kidney, bone, omentum, GIT, retroperitoneum, liver and pancreas [5]. They can occur sporadically in the mediastinum, retroperitoneum and internal organs [6]. The lymphangiomatous process can involve additional sites in a diffuse or multifocal fashion, such as the liver, mediastinum and the lung, called as the lymphangiomatosis syndrome [7]. Only 189 reported cases of splenic lymphangiomas were documented in literature between 1939 and 2010 [8], with only very few reported in adults [7]. Isolated splenic lymphangiomas are much rarer with only 9 cases being reported between 1990 and 2010 [9,10]. We report a case of a splenic lymphangiomas, presenting in an adult, detected for the first time in the said patient.

CASE REPORT

PATIENT INFORMATION

A 68 year old woman, of Indian origin, with no prior co morbidities, presented with history of generalised weakness for 3 months duration, associated with loss of appetite. She gave no history of significant loss of weight. She gave history of increased fatigue and inability to sustain daily activities, but gave no history of abdominal pain, bleeding manifestations, constipation, fever, chronic cough, headache, giddiness or night sweats.

CLINICAL FINDINGS

On examination, she had significant pallor, no icterus and stable vitals. Her abdomen was soft, non tender, without any palpable mass or organomegaly. Her initial blood investigations showed haemoglobin of 10.4g/dL and an elevated platelet count of 6.00 lakh/cumm, with an otherwise normal white cell count. Her liver function tests, electrolyte levels and stool microscopy were within normal limits with a non reactive serology.

DIAGNOSTIC ASSESSMENT

An ultrasound abdomen as part of a routine screening to identify the cause of anemia showed the spleen enlarged and measuring 12.4 cm, with normal echotexture. A well defined thin walled anechoic cystic lesion was noted in the upper pole of the spleen measuring 3.3 X 2.6 cm, without any septations or internal solid components. Another mixed solid and cystic lesion was noted along the medial surface of the spleen, measuring 4.4 X 3.8 cm, with the largest cystic lesion in it measuring 12 X 10 mm, without any evidence of calcifications. A provisional diagnosis of a probable hydatid cyst in the spleen was made and she was planned for a Contrast Enhanced CT Abdomen and Pelvis for further evaluation. CECT scan showed a well defined cystic lesion (+19HU) measuring 3.1 X 2.7 cm in the upper pole of the spleen, without any internal septations, abnormal enhancement on post contrast or enhancing mural nodule (Fig. 1). Another well-defined lobulated cystic lesion (+28HU) measuring 4.0 X 3.2 cm in the mid pole of the spleen with thin enhancing internal septae, without calcification or abnormal enhancement on post contrast or enhancing mural nodule was noted (Fig. 2). A plain x-ray of the chest showed patchy non homogenous opacity in the left upper zone probably suggestive of pneumonitis.

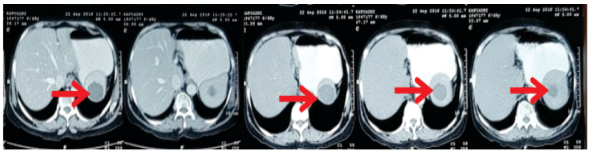


Fig. 1 - Well defined cystic lesion (+19 HU) measuring 3.1 X 2.7 cm in the upper pole

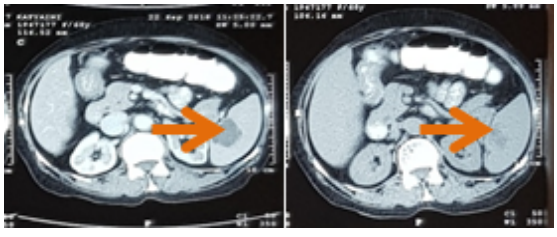


Fig. 2 - Well-defined lobulated cystic lesion (+28 HU) measuring 4 X 3.2 cm in the mid pole

THERAPEUTIC INTERVENTION

A diagnosis of isolated hydatid cyst of the spleen was made and invasive diagnostic modalities, including aspiration of the cyst was not considered in view of a possible anaphylactic reaction. The patient was preoperatively transfused 2 units of packed red blood cells for anemia correction and was started on iron supplementation therapy. She was planned for an elective splenectomy in view of which she received Pneumococcal and Hemophilus vaccines. Her surgery was uneventful.

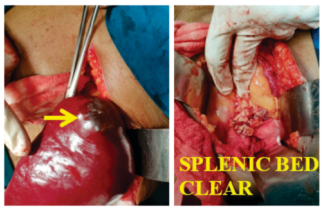


Fig. 3 - Intraoperative view

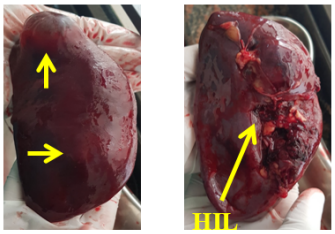
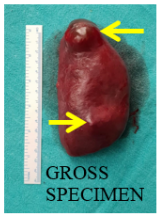


Fig. 4 - Post Splenectomy Specimen

Intraoperatively, two cystic swellings were found, one in the upper pole and another in the mid pole (Fig. 4). Splenectomy was done, with the splenic bed noted to be clear (Fig. 3) and the specimen was sent for histopathological examination. No accessory spleens were found during the surgery.

FOLLOW-UP

Gross examination of the specimen showed the spleen of size measuring 14 X 9 X 4 cm, with two cystic swellings on the anterior surface of the spleen, one in the upper pole and one in the mid pole. Cut sections of the spleen exuded serous fluid. A cyst measuring 2 X 2cm was seen and a multi cystic lesion measuring 3 x 4 cm was seen in the subcapsular area. On microscopic examination, multiple sections showed splenic parenchyma with a lesion in the subcapsular area composed of cystic spaces filled with eosinophilic proteinaceous material and blood cells, with adjacent parenchyma being unremarkable, with normal splenic tissue. A diagnosis of lymphangioma was given.



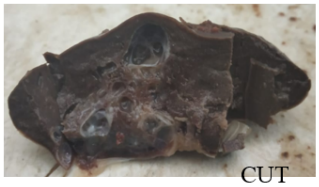


Fig. 5 - Gross Specimen *Fig. 6 - Cut Section*

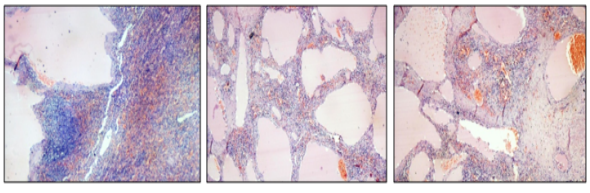


Fig. 7 - Histopathological Appearance

DISCUSSION

Splenic lymphangiomas are predominantly found in children, diagnosed incidentally before the age of 2. They are thin-walled cysts with trabeculated and fibromuscular internal morphology, covered in endothelium and full of eosinophilic proteinaceous fluid. Lymphatic spaces are found on the wall of the lymphangioma containing lymphatic tissue and smooth muscle fibres. The subcapsular location of a lymphangioma is the most common, the intraparenchymatous location being rarer. They are generally asymptomatic, or can cause splenomegaly which can be complicated by haemorrhage, consumptive coagulopathy, hypersplenism and portal hypertension.

Ultrasound is a useful initial test. Colour Doppler can demonstrate vascularisation of the cyst, including the intrasplenic arteries and veins throughout the walls of the cyst. Computed tomography is superior for diagnosis as it shows the topography of the lesion, its size, nature and anatomical relationships. Immunohistochemical confirmation of lymphangioma is by reaction to factor VIII and the specific D240 endothelial marker.

Differential diagnoses of splenic lymphangiomas include haemangiomas, splenic infarction, septic embolism, chronic infection, lymphoma and metastasis.

Treatment modality usually considered is total splenectomy via a median or a left subcostal laparotomy done electively. Accessory spleens should be looked for and removed to prevent recurrence. The rates of recurrence and malignancy are low and the prognosis is good, although there are some cases of lymphangiomas becoming malignant lymphangiosarcomas.

Splenic lymphangiomas are believed to be a hamartoma by most pathologists, due to anomalous congenital expansion of the lymphatic vessels, bleeding or inflammation leading to obstruction and subsequent lymphangiectasia [5, 11]. Macroscopically, splenic lymphangioma can come in solitary nodules, multiple nodules, or as diffuse lymphangiomatosis [12]. It consists of a single large cyst with a thick fibrous wall or multiple variable-size thin-walled cysts filled with clear fluid and separated by fibrous tissue or residual splenic tissue. In cases of diffuse lymphangiomatosis, the spleen is replaced by expanding cysts that result in little remaining normal parenchyma [5, 13]. Microscopically, they are classified into capillary, cavernous or cystic lymphangiomas, with cystic lymphangiomas being the most common [14].

Regarding our patient, the splenic lymphangiomas was asymptomatic, presenting with non specific symptoms of anemia and fatigue and diagnosed incidentally on a work up. The pre operative differentials included hydatid cysts or other parasitic cysts, hemangioma and lymphoma. No serological investigations were carried out to rule out the possibility of parasitic cysts in our patient due to the need for outsourcing of the same to speciality testing centres. No image guided aspiration or biopsy was carried out preoperatively to confirm the diagnosis, due to the possibility and high risks of bleeding, anaphylaxis and inadequacy for diagnosis [3]. The patient was managed with elective splenectomy, to prevent any possible complications.

CONCLUSION

Splenic lymphangioma is very rare, more so in adults. The clinical presentation of splenic lymphangiomas is usually related to the size of the spleen, but they can be asymptomatic or discovered by chance on imaging studies performed for a different reason. If they are large, they can cause abdominal pain, loss of appetite, nausea, vomiting, weight loss, and a palpable mass on physical examination. Contrast ultrasound enables better identification of these tumors. The final diagnosis should be based on the sum of the clinical, radiological and histopathological data. The treatment of choice is splenectomy and the prognosis is good.

ADDITIONAL INFORMATION

This case is being published as a rare presentation of a rare condition, which is splenic lymphangiomas, presenting for the first time in adulthood, with no prior symptoms to suggest its presence since childhood, with appropriate consent taken from the patient and her kin for publication of the same with non disclosure of patient identity and information. The authors have no conflicts of interests in the final diagnosis and management of this case. Hospital expenditure and treatment costs during the course of hospital stay were appropriately borne by the patient.

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