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AUTHORS:

Amarjothi J MV[1], Bennet D [2], Prabakar R [3], Kannan D[4], Naganath Babu O L [5]

AFFILIATIONS:

[1]MS, MRCS, DNB, Registrar, Department of surgical gastroenterology, Madras medical college, Chennai, India,drmosesvikramamarjothi@hotmail.com

[2]MS, McH, Asst, Professor, Department of surgical gastroenterology, Madras Medical College, Chennai, India, drbennet63@gmail.com

[3]MS, McH, Asst, Professor, Department of surgical gastroenterology, Madras medical college, Chennai, India, kalprabha@gmail.com

[4]MS, McH, Professor, Department of surgical gastroenterology, Madras Medical College, Chennai, India, malarkannan08@gmail.com

[5]MS, McH, FRCS, MNAMS, Professor, Department of surgical gastroenterology, Madras medical College, Chennai, India, naganathbabu@gmail.com

CORRESPONDING AUTHOR DETAILS

Amarjothi J M V

ROOM NO 500, 4 TH floor, Tower 2, Department of surgical gastroenterology,

MMC, Chennai-3, Tamil nadu, India

Email: drmosesvikramamarjothi@hotmail.com

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ABSTRACT

Splenic lymphangiomas are rare tumors seen most commonly in children. However very rarely, they may also present as symptomatic multi cystic splenic lesions in adults also. Hence a high index of suspicion is needed to diagnose and treat these lesions when symptomatic and to prevent complications like rupture and infection. Accurate radiological diagnosis of splenic lymphangioma is difficult and histopathology with newer Immunohistochemistry markers like D240 is useful to diagnose splenic lymphangioma. We wish to present this interesting case of symptomatic splenic lymphangioma in a middle aged female patient.

Keywords: Splenic lymphangioma, multi cystic spleen, D240, splenectomy

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INTRODUCTION

Lymphangiomas, in general ,are congenital malformations of the lymphatic system. Splenic involvement is rare in lymphangiomas with less than 1% of lymphangiomas localized intra-abdominally Splenic lymphangiomas are usually benign and predominantly affect children, and only a few cases have been reported in adults.. Most cases of splenic lymphangioma in adults require surgical intervention as they may be symptomatic or lead to complications due to the natural course of the disease .

CASE REPORT

Forty-eight-year year-old woman was admitted with multiple splenic cysts detected by abdominal ultrasonography, done for recurrent pain to the upper abdomen for 2 months .The pain was intermittent and localised to the epigastric and left hypochondrium. Physical examination detected moderate splenomegaly in the left upper quadrant extending for about 4 cm from the costal margin. Peripheral blood count, coagulation studies, and liver and kidney function tests were all within normal limits. CECT showed multiple non-enhancing cysts of variable size throughout the spleen. (Figure 1-A). Due to the presence of symptoms, large splenic size and probability of malignancy, open splenectomy was done. Spleen was found to be enlarged with multiple splenic cysts of variable size, where some contained clear fluid and others were haemorrhagic (Figure 1-B). The histopathology was suggestive of splenic lymphangioma (Figure 2-A) with multiple lymphatic vesicles of variable size between 1-3 cm, lined by a single layer of flat endothelial cells filled with clear mucinous fluid. IHC done was positive for Factor VIII related antigen (Factor VIII R Ag) (Figure 2-B) implying the endothelial origin of the cells. D240, a novel IHC marker specific for lymphangioma was also found to be positive (Figure 2-C)

DISCUSSION

.Most common sites of occurrence of lymphangiomas include the neck, mediastinum and retroperitoneum [1]. Splenic lymphangiomas are benign cysts that commonly affect children, and are rarely reported in adults [2-4]. In 1885, Frink reported the first lymphangioma in the spleen [5]. Most cases (80-90%) are reported in infants and children less than two years of age [6]. Cystic lesions of spleen include parasitic and nonparasitic cysts.

Among parasitic ones, echinococcal disease represent most of the cases [7]. Non parasitic cysts are primary or true cysts and pseudocysts. Endothelial true cysts include lymphangiomas or hemangiomas [1]. Lymphangiomas can be classified as capillary, cavernous, or cystic1 However, the delineation is not universally accepted. [4, 8]. Cystic lymphangiomas are composed of numerous cystic spaces containing amorphous eosinophilic fluid lined by a single layer of flat endothelial cells [7]. Splenic lymphangiomas are mostly asymptomatic and if symptomatic most commonly present with abdominal pain [8]. Diagnosis by radiology is usually not conclusive. In most cases, on CT, splenic lymphangiomas present with thin-walled cystic masses without enhancement or with only slight enhancement of the thin septa. Curvilinear peripheral mural calcifications are suggestive of cystic lymphangiomas but not specific [9]. On T1-weighted magnetic resonance imaging, the cystic lesions appear hypo intense or hyper intense when filled with haemorrhagic or proteinaceous material [8, 9]. On T2-weighted images, they are multiloculated hyper intense corresponding to dilated lymphatic channels [8].

The differential diagnoses of splenic lymphangiomas include other solid and cystic lesions of the spleen, such as haemangioma, chronic infection, lymphoma and metastasis .Immunohistochemistry (IHC) markers including CD 31, CD 34, factor VIII R Ag, D240 and VEGFR-3 may show reactivity for splenic lymphangiomas. As CD34 for endothelial cells is not reliable, FVIII RAg and CD31 are used for identification of endothelial cells lining lymphatic spaces [5]. D240 is a monoclonal antibody initially developed against the M2A antigen or podoplanin, a foetal testis-related antigen [10]. Recently, D240 has been reported to be a selective marker for the lymphatic endothelium, and can be used for diagnosing splenic lymphangioma accurately from

haemangioma also making it a valuable addition for studying benign and malignant vascular disorders in routinely processed tissue specimens [11].

VEGFR-3 is a marker for vascular endothelial growth factor C produced by the tumor cells which stimulate proliferation of adjacent lymphatics [12]. Electron microscopy, in splenic lymphangiomas, show rod-shaped storage granules of endothelial cells (Weibel Palade bodies), which emphasize the endothelial origin of these cells [13] As these cysts are large and present with splenomegaly, they usually require splenectomy in most cases to prevent complications. The main complication is recurrence, which is demonstrated in 9.5% of patients, frequently after incomplete resection [5]. Other complications include rupture, infection, abscess formation, and pleural effusion or empyema, haemorrhage, consumptive coagulopathy, hypersplenism and even portal hypertension [14].

Partial resection techniques, aspiration, drainage and sclerosis are accompanied with high recurrence rates and are avoided. Laparoscopic splenectomy can be tried in patients with a normal size spleen.

Conclusion

Splenic lymphangioma should be considered in the differential diagnosis of a multicystic spleen irrespective of the age of diagnosis and may necessitate surgery including splenectomy for associated symptoms or to prevent future complications. Histopathology and novel IHC markers like D240 are useful to make an accurate diagnosis of splenic lymphangiomas

CONFLICT OF INTEREST

Nil

AUTHOR'S CONTRIBUTIONS

Amarjothi J M V

Group1-Conception and design, Acquisition of data, Analysis and interpretation of data

Group2- Drafting the article, Critical revision of the article

Group3-Final approval of the version to be published

Bennet D

Group1-Conception and design, Acquisition of data, Analysis and interpretation of data

Group2- Drafting the article, Critical revision of the article

Group3-Final approval of the version to be published

Prabakar R

Group1-Conception and design, Acquisition of data, Analysis and interpretation of data

Group2- Drafting the article, Critical revision of the article

Group3-Final approval of the version to be published

Kannan D

Group1-Conception and design, Acquisition of data, Analysis and interpretation of data

Group2- Drafting the article, Critical revision of the article

Group3-Final approval of the version to be published

Naganath Babu O L

Group1-Conception and design, Acquisition of data, Analysis and interpretation of data

Group2- Drafting the article, Critical revision of the article

Group3-Final approval of the version to be published

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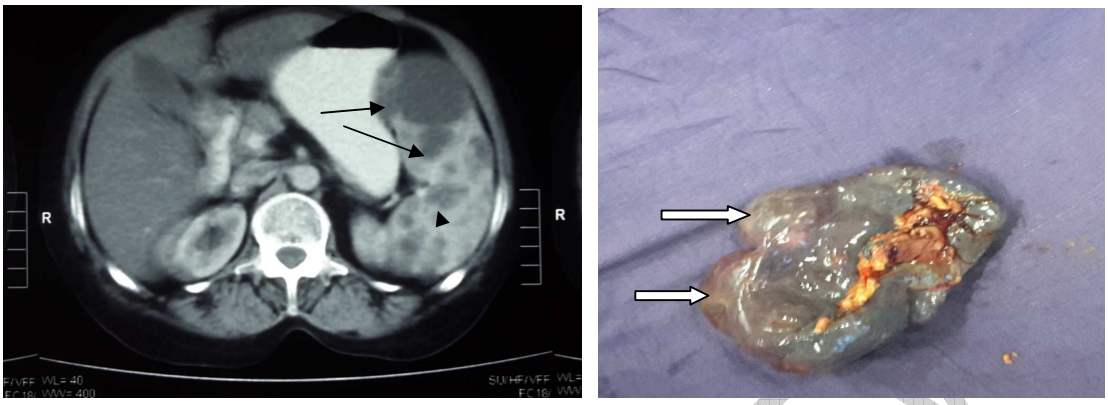
FIGURE LEGENDS

Figure 1: (A) - CECT showing numerous non enhancing cystic lesions in an enlarged spleen (black arrows) (B) - Splenectomy specimen showing numerous cysts in the spleen. (white arrows)

Figure 2 (A) - HPE (HE) 100x (arrows) Showing multiple endothelium lined cystic spaces in splenic architecture (B) - HPE 200x -IHC - endothelial cells showing factor 8 related antigen positivity and granular reaction (arrows) (C) -.IHC- D240 positivity conclusive of splenic lymphangioma (arrows)

FIGURES



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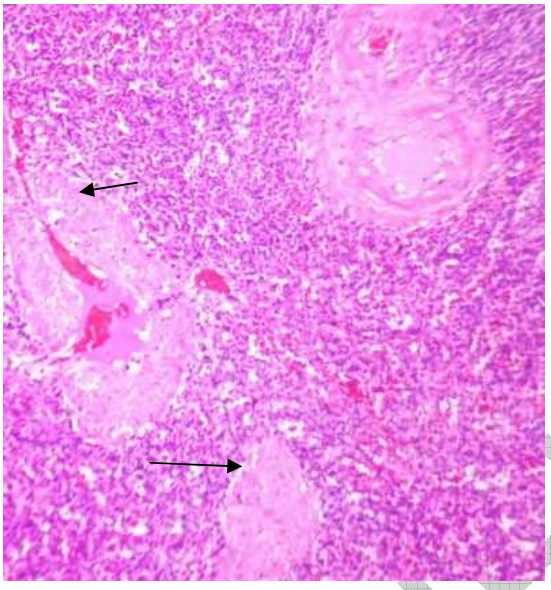

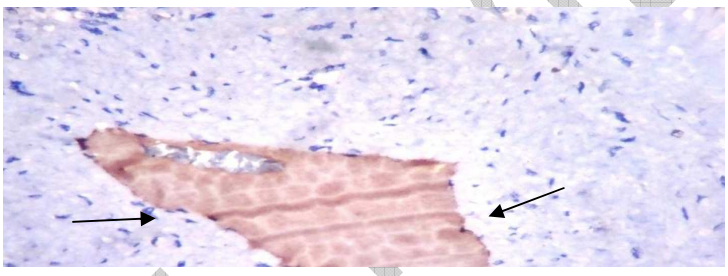

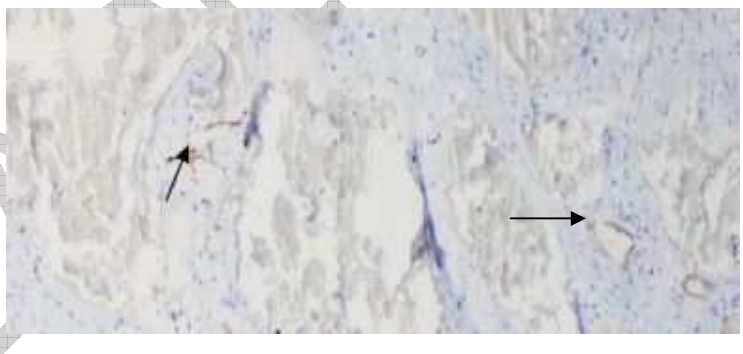

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