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Splenic Lymphangioma in a 35 Year Old Lady: A Rare Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Lymphangioma is a benign cystic neoplasm that results from congenital malformation of the lymphatic system. Among them, splenic lymphangioma is a rare entity accounting for only <0.007% of all tumors. This case highlights the importance of considering splenic lymphangioma in the differential diagnosis of cystic splenic lesions. Most of the patients are symptomless and detected incidentally. A 35-year-old lady presented to a tertiary care hospital of Dhaka, Bangladesh with abdominal pain and fatty food intolerance. On abdominal examination, splenomegaly was noted incidentally. Ultrasonography of whole abdomen showed features of chronic calculus cholecystitis with multiple hypoechoic area in spleen indicating multiple cystic lesion of spleen. Her echinococcal antibody tests were negative. Contrast CT scan of abdomen showed enlarged spleen with multiple cystic lesions indicating polycystic splenic disease associated with gall stones. After proper counseling, she underwent laparoscopic cholecystectomy with open splenectomy. Though she was planned for laparoscopic splenectomy, open conversion was required for huge size of spleen. Her post-operative period was uneventful. She was advised for post splenectomy vaccination and prophylactic antibiotics. Follow up after 1 and 3 months showed no abnormality.

Keywords: Lymphangioma; spleen; splenectomy; abdominal mass.

1. INTRODUCTION

Lymphangioma is a benign cystic neoplasm that results from congenital malformation of the lymphatic system (Ioannidis & Kahn, 2015). This slow-growing tumor is usually seen in paediatric population (Hu et al., 2024). Lymphangioma is rarely seen in adult patients. The commonly involved sites are the neck (75%) and axilla (20%) (Efared et al., 2018). "The abdominal cystic lesions are preferably localized in the mesentery and the omentum" (Perez et al., 2020). "When it involves several sites, it presents as lymphangiomatosis syndrome" (Perez et al., 2020).

Primary neoplasms of the spleen are an extremely rare condition presenting a challenge in diagnosis and management. They can be both benign and malignant. Among them, splenic lymphangioma is an exceptional entity which is a benign tumor resulting from lymphatic malformation (Golmohammadzadeh et al., 2016).

Most of the patients are symptomless and detected incidentally. Sometimes, it can be presented as an abdominal mass with vague symptoms like abdominal pain, discomfort, nausea and vomiting. Rarely, complications like intracystic hemorrhage, coagulopathy, hypersplenism and poral hypertension are observed (Palas et al., 2013). Lymphangioma can be solitary or multiple in number. "The differential diagnosis of splenic lymphangioma includes hydatid cyst, hemangioma and splenic infarction" (Palas et al., 2013). "There are two hypotheses for the formation of lymphangioma of the spleen: (1) congenital malformation of the spleen and (2) inflammation of the lymphatic system causing obstruction and resulting in formation of lymphangioma" (Palas et al., 2013). In the first hypothesis, it is thought that lack of communication between lymphatic ducts result in dilation of terminal ends and formation of large lymphatic sacs. On the contrary, in the second hypothesis it is mentioned that intrauterine or post-partum inflammations of lymphatic ducts may lead to obstruction of lymphatic ducts which resulted in formation of malformed large lymphatic sacs.

"Microscopically, these cysts are composed up of multiple vascular channels lined by single layer of endothelial cells and contain eosinophilic proteinaceous materials" (Al-Shaikh et al., 2017). "Lymphangiomas are classified into three types according to the size of the lymphatic channels: (1) capillary (2) cavernous and (3) cystic".[7] Cystic lymphangiomas are the commonest variant and has a honeycomb appearance of varying sizes of cysts containing lymph lined by a thin endothelium (Al-Shaikh et al., 2017).

Radiological imaging is the main modality of diagnosis of splenic lymphangioma. Cystic lesions of the spleen are difficult to diagnose on radiological findings as they mimic parasitic cysts and other vascular malformations. Imaging tools like ultrasonography, CT scan or MRI are useful to detect splenic lymphangomas. "Ultrasound reveals numerous hypoechoic cysts with hyperechoic septa, as well as visible

calcifications" (Murshid et al., 2024). "CT scan typically reveals subcapsular cysts that are finely defined, thin-walled and have low density" (Murshid et al., 2024). "IMMUNOHISTOCHEMICAL staining includes FVIII-Rag, CD31, CD34 and D2-40 (a selective marker for lymphatic endothelium) for the identification of endothelial cell lining" (Murshid et al., 2024).

"Surgical resection is the mainstay of treatment of splenic lymphangioma to prevent possible complications like splenic rupture, hemorrhage and infection" (Boughdir et al., 2021). "Although total splenectomy is gold standard for the management of splenic lymphangioma, partial splenectomy is becoming popular day by day" (Oubaha et al., 2013). "But it has an increased risk of recurrence in the splenic remnant. Both open and laparoscopic approaches can be made based on patient characteristics, the surgeon's expertise and the extent of splenic enlargement" (Oubaha et al., 2013).

Here we present a rare case of splenic lymphangioma in a 35-year-old lady presented with gall stone disease.

2. PRESENTATION OF CASE

A 35-year-old lady presented to a tertiary care hospital of Dhaka, Bangladesh with abdominal pain and flatulent dyspepsia. Her abdominal pain was in right hypochondriac region which was mild to moderate, dull aching, non-radiating aggravated on taking food and relieved by taking analgesics. On abdominal examination, splenomegaly was noted incidentally. Spleen was non tender, firm in consistency and enlarged about 9 cm from left coastal margin. Her blood picture including complete blood count, liver function and renal function tests were within normal limits. Ultrasonography of whole abdomen showed features of chronic calculus cholecystitis with multiple hypoechoic area in spleen indicating multiple cystic lesion of spleen. Her echinococcal antibody tests were negative. Contrast CT scan of abdomen showed enlarged spleen with multiple cystic lesions indicating polycystic splenic disease associated with gall stones (Fig. 1).

After proper counseling, she underwent laparoscopic cholecystectomy with open splenectomy. Per operatively hugely enlarged spleen with multiple cystic lesions were observed (Fig. 2). Initially she was planned for laparoscopic splenectomy but later converted to open procedure due to huge size of spleen and chance of intraoperative rupture of cyst. . Her post-operative period was uneventful. She was discharged after 5th post-operative day and advised for post splenectomy vaccination for prevention of overwhelming post splenectomy infections (OPSI). Besides, for prophylaxis, she was also advised for taking phenoxymethylpenicillin. Her histopathological report revealed splenic cystic lymphangioma. Follow up visits after 1 and 3 months showed that she was in good health.

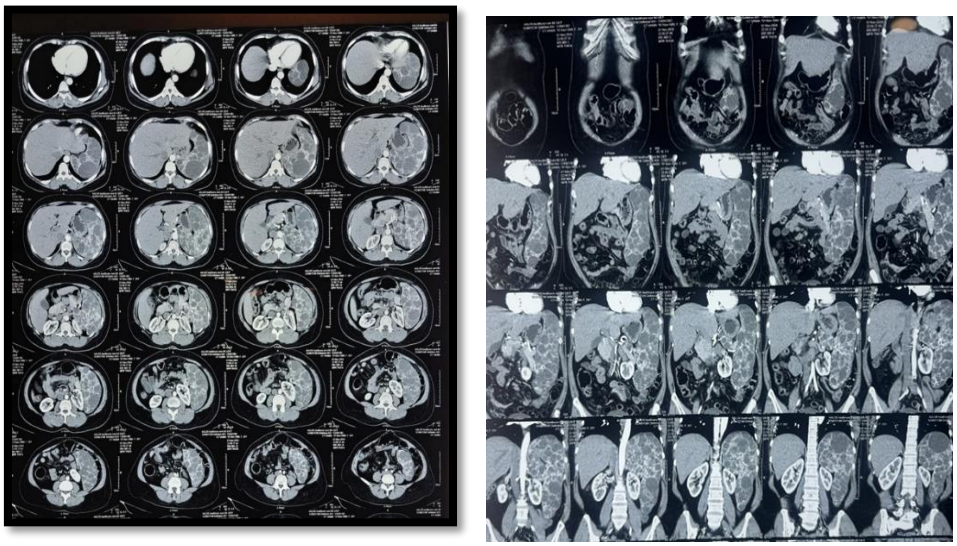

Fig. 1. showing CT scan of the patient

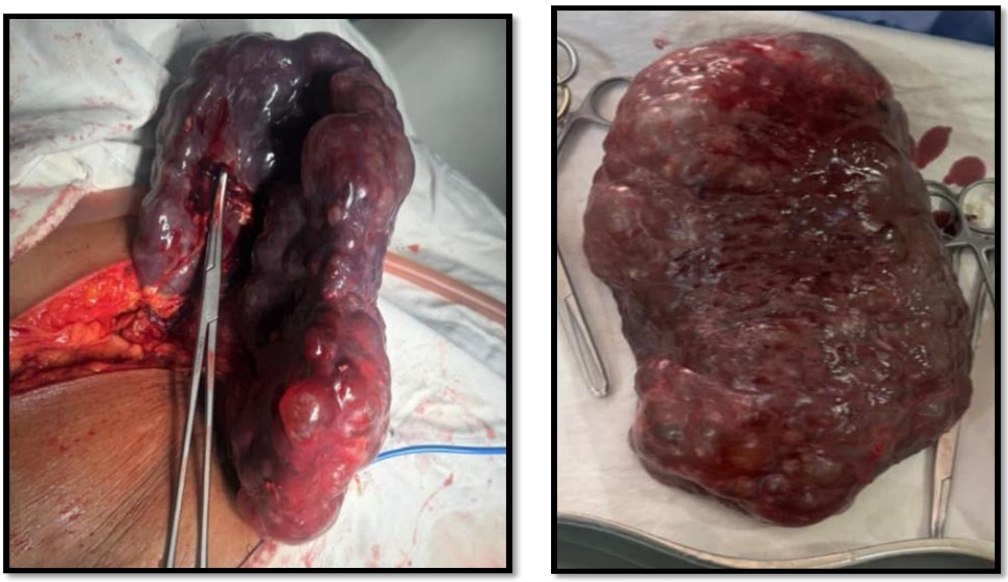

Fig. 2. Showing per-operative view

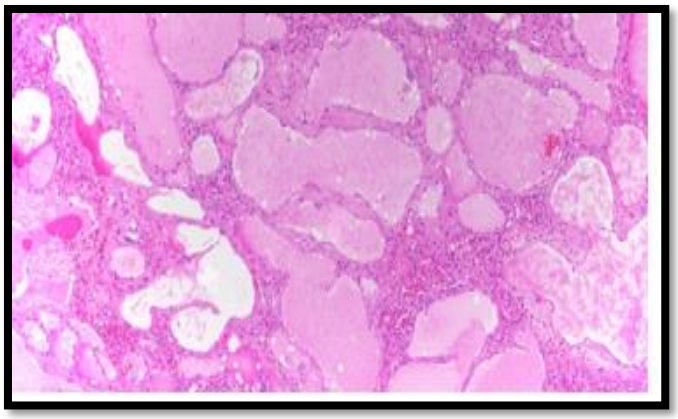

Fig. 3. Histopathology of splenic lymphangioma

3. DISCUSSION

Splenic lymphangioma is a neoplasm of lymphatic system which is rarely seen and accounts for <0.007% of all tumor (Thipphavong et al., 2014). The tumor is benign in nature, usually manifested during childhood and hardly seen after 20 years of age (Thipphavong et al., 2014). During adulthood, the diagnosis is often made incidentally. In our present case, a 35 year old lady presented to us with features of chronic calculus cholecystitis with splenic lymphangioma which was diagnosed incidentally.

In 1885, Frink first reported about splenic lymphangioma (Verghese et al., 2013). So far, very few cases have been reported in the literature regarding splenic lymphangioma. In adults, most of the cases are observed in females above the age of 25. (Shi et al., 2024) In our case, the patient was a 35 year old female showing similarities with previous reports.

Most of the patients of splenic lymphangioma usually do not show any symptoms as they are very slow growing. Occasionally they may present with mechanical discomfort, pain, nausea and vomiting. In our case, the patient originally presented to us for laparoscopic cholecystectomy but an additional diagnosis of splenic lymphangioma was made based on clinical and radiological findings.

Differential diagnoses of splenic lymphangioma include haemangioma, splenic abscess, parasitic cysts, simple cyst and post traumatic pseudocyst of spleen. Imaging modalities and blood picture

are the tools to separate the differentials from lympahngioma. (Thorat & Shaji, 2023).

Among the imaging tolls, ultrasonogram and CT scan of abdomen are most popular ones. These can detect the splenic pathology accurately. Besides, these can also give information about other abdominal pathologies which can help to manage the patient accordingly. In our case, the radiological diagnosis was made by CT scan of abdomen.

"There is a debate regarding splenic lymphangioma whether it is a neoplasm or a hamartoma" (Ousmane et al., 2019). "Most pathologists believe it is a hamartoma due to anomalous congenital dilation of the lymphatic vessels leading to formation of such neoplasm". (Shi et al., 2024). "Macroscopically, splenic lymphangioma can present as solitary, multiple or diffuse lymphangiomatosis" (Shi et al., 2024). It is usually located in subcapsular region consisting of a solitary large cyst with a thick fibrous wall or multiple cysts filled with clear fluid and separated by fibrous septa (Shi et al., 2024). "Microscopically, these cysts are composed of acellular proteinaceous compounds lined by thin wall. The surrounding tissue may be normal or may show congestion, fibrosis infiltrated by macrophages and lymphocytes'' (Thorat & Shaji, 2023). Similar findings were also noted in our case (Fig. 3).

Surgical resection is the treatment of choice in splenic lymphangioma (Ousmane et al., 2019). Surgery is done to avoid complications such as infection and rupture. Though laparoscopic splenectomy is getting popular day by day, open splenectomy is preferred because of its huge size and increased chance of per operative rupture of cyst. Total splenectomy is the surgical choice, as partial splenectomy may give rise to recurrence (Ousmane et al., 2019). In the present case total splenectomy was performed as mentioned in different literature (Ousmane et al., 2019).

4. CONCLUSION

Lymphangiomas are benign neoplasms of the spleen that are infrequently found in adults. Most patients present with no symptoms and diagnosed incidentally. Though being uncommon, lymphangioma should be considered as a differential diagnosis of cystic lesions of the spleen. Splenectomy is the treatment of choice to provide better management to the patients.

DISCALIMER (ARTIFICIAL INTELLEGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

CONSENT

Patient's informed written consent was taken to publish her case for academic purpose.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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