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Splenic Lymphangioma in an Adult: A Case Report

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Abstract **Case Report**

This article presents a rare entity both in terms of occurrence and location: cystic lymphangioma of the spleen. We report a clinical case treated surgically in the visceral surgery department at the MOHAMED V Military Hospital in Rabat.
Keywords: Spleen, Laparoscopy, Splenectomy, Lymphangioma.

INTRODUCTION

Lymphangiomas are benign congenital malformations of the lymphatic vessels [1]. They most commonly occur in the head, neck, and axillary regions. Intra-abdominal lymphangiomas are rare, with a preference for the mesentery [2]. Due to their rarity and unusual location, making an accurate preoperative diagnosis can be challenging for clinicians.

The ultimate diagnosis is usually made by histopathological examination. Splenic lymphangiomas typically affect children and are extremely rare in adults.

We report a case of splenic lymphangioma in an adult patient with hypothyroidism.

CASE REPORT

This is a 62-year-old female patient followed for hypothyroidism treated with levothyroxine, with a history of cholecystectomy 28 years ago and an unexplored jaundice episode that spontaneously resolved 5 years ago. The medical history dates back 2 years with the onset of pain in the left hypochondrium accompanied by a sensation of heaviness worsened by deep inspiration. Physical examination was unremarkable except for mild tenderness in the left hypochondrium. Biological tests showed no abnormalities. An abdominal ultrasound revealed a spleen of normal size measuring 122 x 92 x 80 mm, containing multiple cystic formations, including a central cyst surrounded by smaller cysts. The main lesion had a thin, partially calcified wall measuring 62 x 58 mm and was avascular on color Doppler.

A complementary MRI was requested, showing a spleen of normal size with regular contours, containing multiple cystic lesions with marked T2 hypersignal and T1 hyposignal, with no enhancement after gadolinium injection. The main lesion measured 62 x 58 mm and was surrounded by sentinel microcysts, suggesting a cystic splenic lymphangioma in its polycystic form **(Fig 1).**

A laparoscopy was performed, revealing a spleen of normal size with multiple cystic formations, including a large central cyst. A splenectomy was carried out, and histopathology confirmed the diagnosis of splenic lymphangioma. The postoperative course was uneventful.

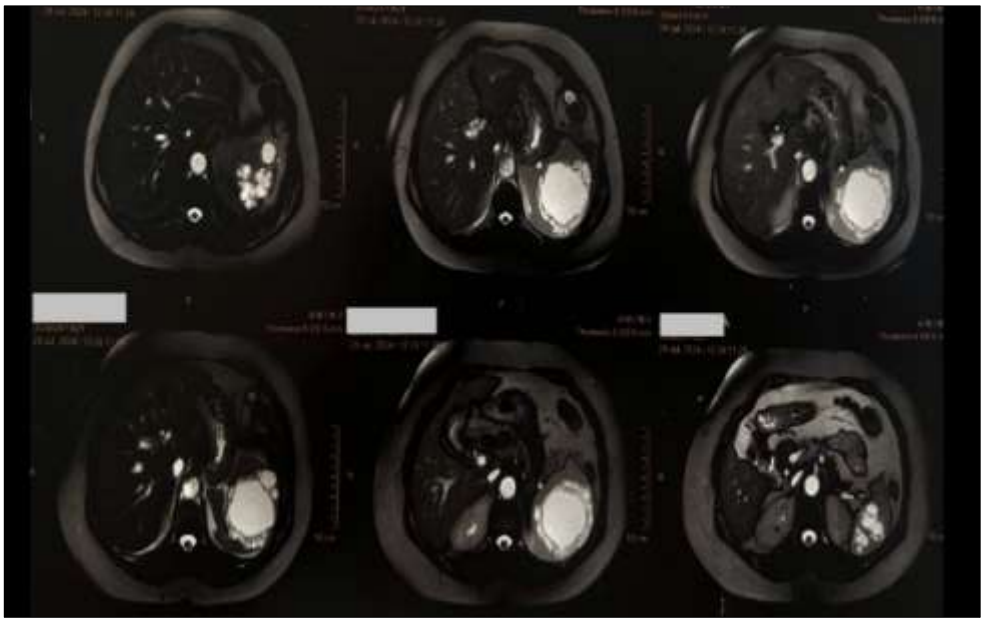

Fig. 1: Abdominal MRI image showing T2 hyperintense cystic lesions in the spleen

DISCUSSION

Splenic lymphangioma is a rare benign cystic tumor, first described by Frink in 1885, and represents less than 0.007% of all tumors [3]. It predominantly occurs in pediatric populations and is seldom observed in adults. Several theories have been proposed to explain its pathogenesis, including sequestration of lymphatic tissue, abnormal budding of lymphatic vessels, failure to integrate with the venous system, and lymphatic outflow obstruction.

Splenic lymphangiomas are considered developmental malformations of the lymphatic system [4]. The majority of reported cases occur in children, with most diagnoses made before the age of 2 years. In contrast, splenic lymphangiomas in adults are extremely rare. While lymphangiomas commonly occur in the neck and axillary regions, involvement of the spleen is exceptionally uncommon [3].

Clinically, these lesions are often asymptomatic. Large and multifocal lesions can cause nonspecific symptoms such as abdominal pain. However, in some cases, they can lead to marked splenomegaly, which may be complicated by hemorrhage, consumptive coagulopathy, hypersplenism, or even portal hypertension [5]. Imaging modalities such as abdominal ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) play an important role in the diagnosis of splenic cystic lesions [6].

The typical ultrasound appearance of the tumour is that of a hypoechoic and heterogeneous lesion, sometimes cystic and partitioned. Doppler shows the absence of flow within the malformation. The CT scan reveals more details on the size and nature of the lesion. MRI remains a second-line examination to assess good mapping of the lesion; the LK of the spleenappears as a hyposignal T1 and a hypersignal T2.

Magnetic resonance has the same advantages as computed tomography. In our case we used Ultrasound and Magnetic resonance. The differential diagnoses of splenic lymphangioma include hydatid cysts, chronic infection, hemangioma, lymphoma, metastasis, and pseudocysts of the spleen. Parasitic infection with echinococcus granulosus is the main cause of cystic lesions of the spleen [7].

In our case, the first clinical and radiological diagnosis was hydatid cysts, but it was ruled out after performing laboratory test.

Definitive proof of the diagnosis of cystic lymphangioma is provided by pathological examination of abiopsy or surgical specimen. The macroscopic appearance is whitish or translucent, unilocular or polycystic with or without communication channels with serous or chylous contents. Microscopically: it is a cystic formationbordered by an endothelial lining with connective tissue partitions whose thickness is related to the age of the cyst [8].

Total splenectomy remains the treatment of choice for splenic lymphangiomas, especially in cases involving large lesions [9].

This approach helps prevent potential complications such as splenic rupture, hemorrhage, or hypersplenism, and also allows for the exclusion of malignant pathology. Alternative treatments, such as aspiration and drainage, have demonstrated limited effectiveness [10].

CONCLUSION

Lymphangiomas are rare benign tumor of the spleen that are especially rare in adults. Splenic localization is incidental occurring in left upper quadrant pain exploration and rarely during a complication. The diagnosis is improved by the ultrasound and CT scan. But the histological exam allows to confirm the splenic lymphangioma diagnosis. The treatment is the total splenectomy under laparotomy or laparoscopy. The conservative medical treatment has not yet shown its effectiveness.

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