

Case Report

Splenic Lymphangioma a Rare Tumor Rarely In Adult Patient Case Report

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Abstract

Lymphangiomas are benign tumors originated from a congenital malformation of the lymphatic system. Most lymphangiomas are located in the neck and armpit; however, they can appear in other locations (5% of all cases), rarely they are found in the spleen. Generally, lymphangiomas are found in children (80%-90%) in which they are diagnosed incidentally. They are more frequent in women, 80-90% are diagnosed during childhood and the mostly, they are detected before 2 years of age.

We present a case report of this rare tumor rarely in a 57-year-old married woman with a surgical history of pleomorphic leiomyosarcoma of the thigh. On follow-up, contrast-enhanced Computed Tomography scan was carried out which revealed two splenic tumors, 7×5 cm and 1 x 1 cm, well-defined intrasplenichypodense lesions, without internal septations. With a preoperative diagnosis of a splenic tumor, the patient was subjected to hand-assisted laparoscopic splenectomy.

Gross examination of the spleen showed a mass of brown-red tissue, rubbery consistency, measuring 7.5 x 6 x 5 cm, with a white-yellow center, and a yellow tumor of 7 mm. On microscopic examination, multiple dilated lymph sacs filled with lymph were observed. The diagnosis of a cystic lymphangioma of spleen was histologically made.

INTRODUCTION

Lymphangiomas are benign tumors originated from a congenital malformation of the lymphatic system, in which due to an obstruction or agenesis of lymphoid tissue, a secondary lymphectasia is developed from the absence of a normal communication between the lymphatic ducts, which result in the formation of an empty sac, progressively dilating until a cyst is formed [1,2].

Lymphangiomas may present as a part of a systemic cystic lymphangiomatosis or may occur as a solitary lesion. Most lymphangiomas are located in the neck (75%) and armpit (20%), where it is known as cystic hygroma; however, they can appear in other locations (5% of all cases) [2-5], rarely they are found in the spleen [2,6-8]. Gohn'sserie [2], published in 2005, informed about 14 cases of intra abdominal lymphangiomas during a period of 15 years, in which one of the patients has an splenic lymphangioma; and the Allen's serie [9], published in 2006, informed about 6 cases of intraperitoneal lymphangiomas during an 18 years period, none of them where splenic. In total, there have been published 180 cases of splenic lymphangiomas during 1939 and 1990 and only 9 cases since that time until 2010 [4], confirming the rarity of these tumors.

CASE PRESENTATION

A 57-year-old married woman with a surgical history of pleomorphic leiomyosarcoma of the thigh. On follow-up, contrast-enhanced Computed Tomography scan was carried out which revealed two splenic tumors, 7×5 cm and 1 x 1 cm, well-defined intrasplenichypodense lesions, without internal septations (Figure 1). The wall was thin and had a sharp demarcation to splenic parenchyma. There was no rim or internal enhancement.

There was no history of previous abdominal trauma and she had no history of fever, weight loss, and loss of appetite as well. History was negative for infection by malaria or any hematological disorder. Her bladder and bowel habits were normal. Physical examination showed good general condition. The blood and urine investigations were within normal limits. Differential diagnosis included a cystic lymphangioma of spleen, hydatid cyst, haemangioma, pseudocysts of spleen and splenic metastasis.

With a preoperative diagnosis of a splenic tumor, the patient was subjected to hand-assisted laparoscopic splenectomy. Preoperative vaccination against capsulated organisms was done 4 weeks prior to surgery. The patient was placed on a right lateral decubitus position. Under general anaesthesia, a hand

port was inserted in the periumbilical position. Two additional ports were placed under direct vision. The splenectomy was then carried out in a standard fashion: ligation of the short gastric vessels in the gastrosplenic ligament, dissection of the hilum (hilar dissection was performed with extreme caution to avoid injury to the tail of the pancreas), use of the endo-GIA for the splenic vessels, dissection of the peritoneal attachments (phrenicosplenic, splenocolic and lienorenal ligaments), removal of the intact spleen through the hand port, and closure of ports after meticulous haemostasis. An elastic drainage tube was left in left subphrenic space.

The spleen was sent for histopathologic examination. The weight of the spleen was about 375 g and it measured 13.5 x 8.5 x 5 cm. The cut section showed a mass of brown-red tissue, rubbery consistency, measuring 7.5 x 6 x 5 cm, with a white-yellow center, and a yellow tumor of 7 mm at a distance of 1 cm. On microscopic examination, multiple dilated lymph sacs filled with lymph were observed (Figure 2). Final diagnosis of cystic lymphangioma of spleen was made. There was no sarcoma. The postoperative

course of patient was uneventful and was discharged on 5th postoperative day after the drain was removed.

DISCUSSION

Generally, lymphangiomas are found in children (80%-90%) in which they are diagnosed incidentally [4,8-10]. They are more frequent in women, 80-90% are diagnosed during childhood and the mostly, they are detected before 2 years of age [12]. Histologically, lymphangiomas are classified in three subtypes according to the final size of the lymphatic ducts: capillary (super-microcystic), cavernous (microcystic) or cystic (macrocytic) [1,5,12,13].

The most common form of presentation of lymphangiomas is as a one, small and subcapsular lesion; however, they can also be multicystic. This last form is commonly associated with other pathologies [1,14]. A splenic lymphangioma can be asymptomatic and be diagnosed during an abdominal surgery performed for another reason, it can be detected while the pathological study of the spleen, or it can cause splenomegaly or

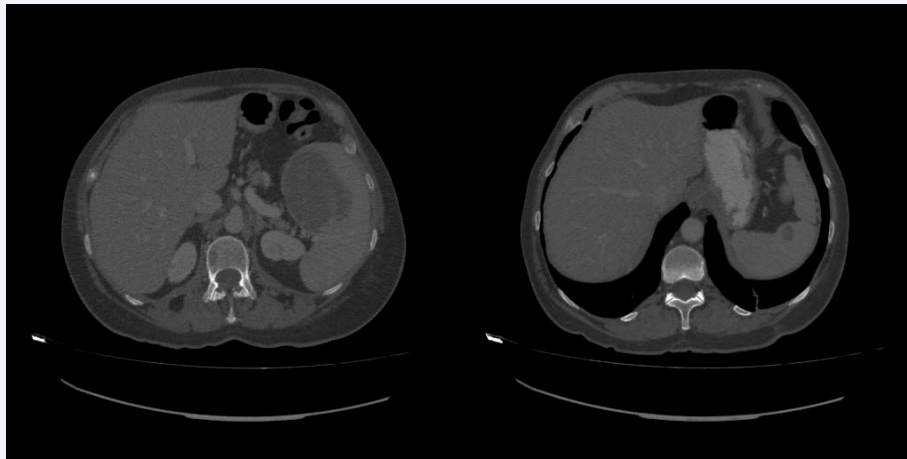


Figure 1 Contrast-Enhanced Computed Tomography: two solid splenic mass.

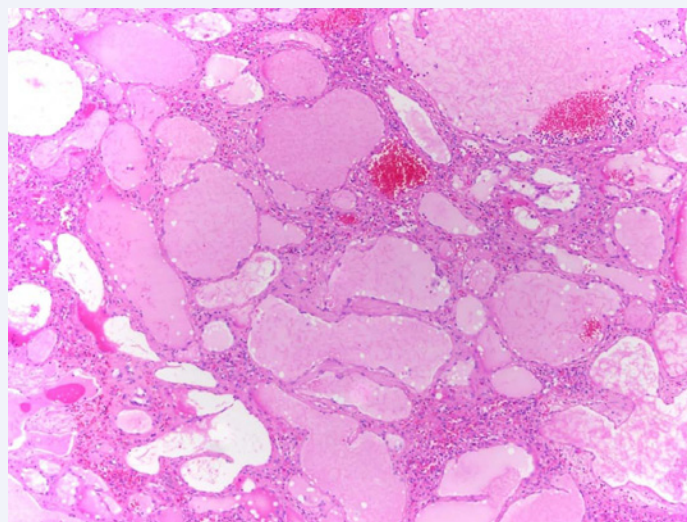


Figure 2 Multiple micro cystic structures lined by a flat layer of attenuated endothelial cells and filled with eosinophilic amorphous proteinaceous material (H&E, 40x).

have huge splenic nodules. In this last case, it can be complicated with consumptive coagulopathy, hypersplenism and portal hypertension [15,16]. Symptomatic cases can be presented with pain over the left hypochondriac region due to splenomegaly and the impingement of the cyst on neighboring organs [16]. It is commonly followed by fever, nausea, vomits, anorexia and loss of weight [4], however, this opinion is not shared by everyone [6]; pain in the left shoulder and constipation are infrequent. Due to the similarity with the signs and symptoms of the hydatid disease, it is commonly [6,17] mistaken with this one, in which a negative result in the agglutination test for *Echinococcus granulosus* does not always rule out this diagnosis. Sometimes, an asymptomatic patient consults about a painless abdominal mass, with recent growth and easily palpable on physical examination [6,17]. Occasionally, the abdominal mass effect caused when the spleen reaches 3-4 kg of weight, can cause diaphragmatic immobility with the following atelectasis or pneumonia [6,17]. If the lymphangioma partially affects the spleen, hematologic disturbances will not be presented; if the affection is diffuse and includes all the spleen, it causes an arrest phenomenon which is expressed with anemia, granulocytopenia and thrombocytopenia [1,17].

US, CT and MR imaging are complementary tools for noninvasive characterization and evaluation of splenic diseases. Ultrasounds are useful as an initial exam for cystic splenic diagnosis; however, it does not limit the bounds of the lesion [12,16,17] although, contrast ultrasounds allows a better study of these lesions. CT scanning, on the other hand, not only delimits the topography of the lesion, but also it shows its size, nature and anatomical relationships [2,14,18]. The MRI does not have more advantages than the TC except that it avoids the radiation to the patient. Ultrasounds and TC are the tests of choice for diagnosis and planning the surgical strategy [9]. Radiological studies may reveal splenomegaly or a normal-sized spleen, curvilinear calcifications in the cystic wall or mass effect on the adjacent viscera. Ultrasound studies will show well-defined hypoechoic cystic lesions with occasional internal septation and some echogenic calcifications. Color Doppler can show the vascularization of the cyst, including the intrasplenic arteries and veins along the walls of the cyst [14,18]. In CT studies, lymphangiomas appear as single or multiples masses with well-defined margins, typically subcapsular, which do not enhance after intravenous contrast administration; sometimes, around the biggest cyst there are small satellite lesions which suggest splenic lymphangioma when accompanied by peripheral wall calcifications [5,12]. On MRI imaging, the mass is shown as hyperintense areas on T2 and isointense areas on T1. The diagnostic differential of the splenic lymphangioma includes other solid and cystic lesions as hemangiomas, splenic infarction, septic embolism, chronic infection, lymphoma and metastasis. According to Pearl-Nassar classification, there are only three types of cystic lesions: parasites cysts; the first with epithelial covering (dermoid, epidermoid or transitional) or endothelial covering (hemangiomas or lymphangioma) and the second, which do not have endothelial covering [19].

Splenic lymphangiomas are thin-walled cysts with a trabeculated and fibromuscular internal morphology, covered in endothelium and full of eosinophilic aprotinaceous

fluid [1,2,5,17]. On the wall of the lymphangioma there are lymphatic spaces, lymphatic tissue and smooth muscle fibers [9]. The subcapsular location of the lymphangioma is the most common, being the intra-parenchymal location the rarest [5]. Immunohistochemical confirmation of lymphangioma is by reaction of factor VIII and the specific D2-40 endothelial marker [5]. Histological study rules out without difficulty the suspected diagnosis of parasitic cysts and establishes the vascular origin of the lesions.

The treatment for splenic lymphangioma depends on their size. Traditionally, bigger and symptomatic lesions have been treated by splenectomy via median or left subcostal laparotomy [17,20]. However, since the first laparoscopic splenectomy of a splenic lymphangioma performed by Know et al in 2001, several authors propose the laparoscopic surgery as the best approach for these spleen tumors [19,21,22], considering the splenomegaly as a contraindication to this approach [4]. Although many surgeons recommend total splenectomy as standard treatment of splenic tumors [9,2,8,17], the objective of resection benign tumors should be the local excision of the tumor, preserving the majority parenchymal mass as possible [23] or even the whole spleen to prevent the consequences of the asplenia [17,19,24,25]. During surgery, open or laparoscopic, it is mandatory to look for accessory spleens, and they should be removed, as they could form part of the process. The surgery should be indicated without delay, unless the cyst is infected or there are any other circumstances which contraindicate the procedure [26]. Recurrence and malignancy rates are low and the prognosis is good; although, there are some cases of malignancy of lymphangiomas to lymphangiosarcoma. Conservative treatment of the splenic lymphangioma with interferon-alpha was used in a child by Reinhart et al. [23], with success and good tolerance; however, it is not known the time and doses for an optimum treatment to cure this disease.

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