

Lymphangioma Of The Spleen In An Adult

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Citation

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Abstract

Lymphangioma of the spleen is an extremely rare, benign vasoformative tumor which occurs mainly in childhood. We report a case of 46-year-old female who presented with abdominal discomfort and fullness in the upper abdomen for one year. A large splenic cyst was diagnosed by ultrasonography and contrast-enhanced computerized tomography of the abdomen. A splenectomy was carried out for this cyst and the histopathological examination revealed a lymphangioma of the spleen.

INTRODUCTION

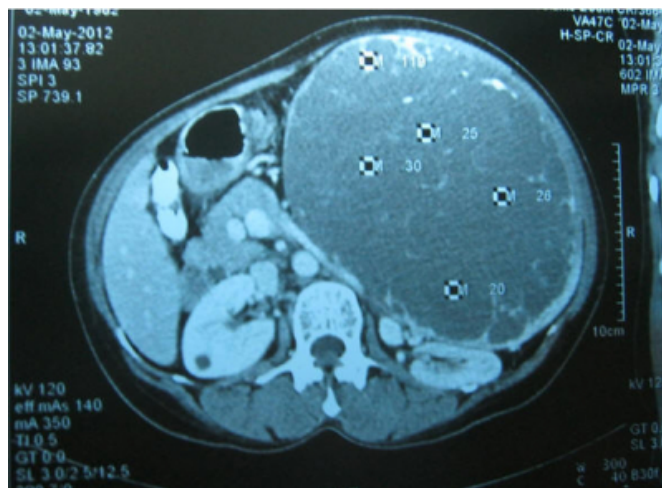
The most frequent splenic malignant tumor is lymphoma, while hemangioma is the most common benign neoplasm.¹ Splenic lymphangioma is a rare benign vascular neoplasm that occurs mainly in childhood.² Involvement of the visceral organs is less common.³ Splenic involvement is quite rare.⁴ The rarity of this condition, especially at an advanced age, prompted us to report this case.

CASE REPORT

A 46-year-old female presented to the surgery department of our institute with a one-year history of abdominal discomfort and a sensation of fullness in her left upper abdomen. Physical examination revealed a large, smooth, non-tender mass occupying the left hypochondrium. The spleen could not be made out separately. The routine hematological and biochemical tests and the chest radiograph were normal. Ultrasonography and computerized tomography of the abdomen showed a well defined splenic cyst of 20x18X16cm, displacing the stomach to the right. The lesion revealed enhancing septae which were suggestive of lymphangioma. Surrounding organs were displaced but appeared normal. A simple cyst was seen involving the mid-polar region of the right kidney. Liver, left kidney and pancreas appeared normal (Figure 1).

Figure 1

CT scan showing a large multiseptated cystic lesion measuring 20x18X16cm involving the spleen. A simple cyst involving the mid pole region of right kidney can also be seen.



Furthermore, no accessory spleens were discovered. The diagnosis of a splenic cyst was confirmed and an exploratory laparotomy was planned. The patient received Pneumovac™ and was scheduled for splenectomy in two weeks. On exploration, a very large cyst of the spleen occupying the entire left upper abdomen was found. The large size and little splenic parenchyma made preservation of the spleen impossible. A splenectomy was performed and the specimen was sent for histopathological examination. Accessory spleens were searched. The excised organ measured 21x19x16cm and weighed 5.2kg with intact surface (Figure 2).

Figure 2

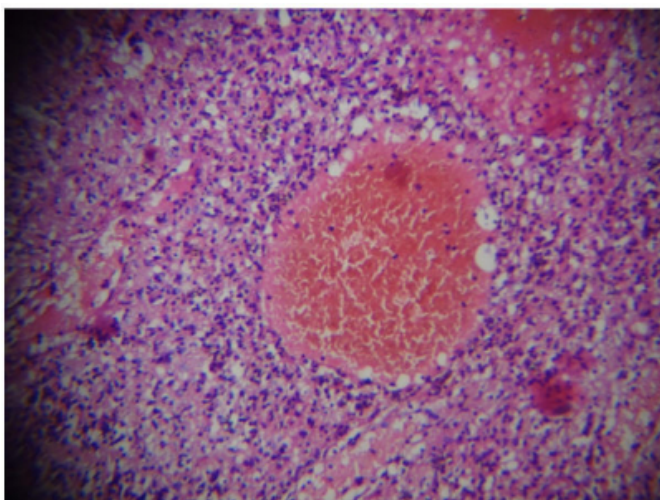
Fig. 2 showing the excised spleen along with the lymphangioma measuring 21x19x16cm and weighing 5.2kg with intact surface.



Histopathological examination revealed dilated lymphatic spaces lined with endothelial cells and filled with proteinaceous eosinophilic material. The supporting stroma was composed of collagen containing lymphocytes and lymphoid aggregates. The histopathological findings were consistent with the diagnosis of lymphangioma of the spleen (Figure 3).

Figure 3

Histology of the excised specimen showing dilated lymphatic spaces lined with endothelial cells which are filled with proteinaceous eosinophilic material. The supporting stroma is composed of collagen containing lymphocyte and lymphoid aggregates (Hematoxylin-Eosin, x10).



The postoperative course was uneventful and the patient was discharged 5 days after the operation.

DISCUSSION

Lymphangiomas, first described by Rodenber in 1828, are benign malformations composed of endothelial lined cysts containing lymph.⁵ They are believed to have a malformative origin in which obstruction or agenesis of lymphatic tissue results in lymphangectasia secondary to lack of normal communication of the lymphatic system.⁶ Lymphangioma is usually classified as simple, cavernous or cystic according to the size of the dilated lymphatics. It may involve many sites and organs such as the soft tissues, bones, mediastinum, liver, kidney, adrenals, testis, lymph nodes and intestines.⁷ Most lymphangiomas (95%) occur in the neck and axillary regions; the remaining 5% are located in the mesentery, retroperitoneum, abdominal viscera, lung, mediastinum, etc. Splenic lymphangioma occurs mainly in childhood and only few cases have been described in adults.²

The clinical presentation may vary according to the size of the cyst and the involved organ. The disease may be discovered by chance or in cases of a considerably enlarged spleen. It may cause left upper abdominal quadrant pain, nausea, vomiting and weight loss due to gastric or left colic flexure compression, or in some cases breathing difficulties due to a raised diaphragm.⁸ Ultrasonography usually shows anechoic or hypoechoic cysts with internal debris which appear on CT scan as multiple non-enhanced, low-density lesions, sometimes with mural calcifications.⁹ Total splenectomy is the most common surgical option, but in small or localized lymphangiomas, partial resection of the spleen can be a valid alternative.¹⁰ It is very important to search for accessory spleens and remove them because lymphangiomatosis can be also found in accessory spleens.¹¹

The presented case is being reported to highlight the following points:

- Though lymphangioma of spleen is an uncommon entity, it should be considered in the differential diagnosis of a splenic cystic mass.
- It is very important to search for accessory spleens because lymphangiomatosis can also be found in accessory spleens.

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