

Splenic Cystic lymphangiomas presenting with massive splenomegaly

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Citation

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Abstract

Cystic lymphangiomas of the spleen is an extremely rare pathologic entity. It is considered to be the result of a developmental malformation of the lymphatic system and can involve the spleen alone or may be a part of multiorgan disease. It is usually seen in children. We report a case of cystic lymphangiomas of the spleen occurring in an elderly woman presenting with massive splenomegaly with emphasis on the rarity of the case in old age and on the problems in the differential diagnosis.

INTRODUCTION

Lymphangiomas are benign lymphatic system malformation of obscure aetiology. It can be either solitary or multiple. They principally occur in children or young adults and are rarely seen in the elderly. The most common locations of lymphangiomas are the neck and the axillary region¹. Cystic lymphangiomas of the spleen is a very rare entity and is usually solitary and asymptomatic. We report 51 year old woman who presented with massive splenomegaly which turned out to be cystic lymphangiomas. Even though rare, cystic lymphangiomas should be considered in the differential diagnosis of multiple focal lesions of the spleen even among elderly.

CASE REPORT

A 51 year-old woman presented to us with history of vague pain in the left side of the abdomen. There was no fever, dyspepsia or weight loss. Physical examination showed no pallor or lymphadenopathy. Spleen was grossly enlarged- 20 cms below the costal margin. Initial diagnostic possibilities considered were myelofibrosis or low grade lymphoma. Complete haemogram results were within normal limits. Bone marrow aspirate and biopsy was normal. Ultrasonography and CT scan of the abdomen showed splenomegaly with focal lesions which were highly suggestive of lymphoma or granulomatous disease. As a diagnostic as well as therapeutic option she underwent elective splenectomy without any complications. Spleen on gross pathologic examination was measuring 18 x 13 x 6 cms and weighing 700grams. On cut section it showed

multiloculated cysts replacing the entire splenic parenchyma. These cysts were of varying sizes ranging from 0.1 cm to 2.5 cms in diameter (Figure.1). Histologically, multiple cystic spaces lined by flat endothelial cells were seen (Figure.2). On immunohistochemistry the cells were found to be positive for CD 31 and negative for AE1/AE3 (pan cytokeratin); confirming the endothelial nature.

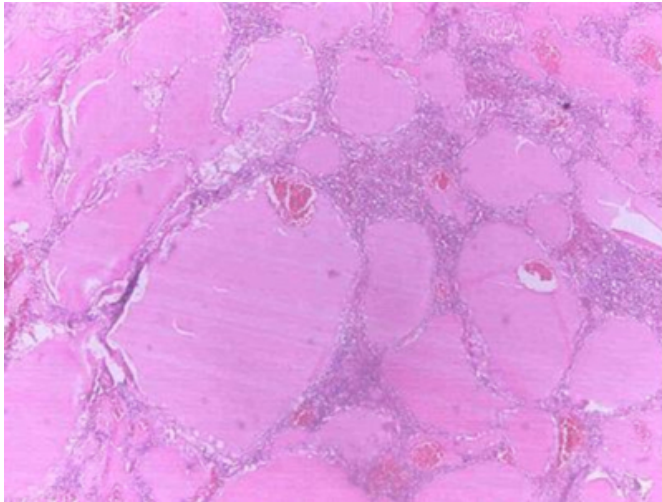
Figure 1

Figure 1: Cut section of spleen showing cysts of varying sizes ranging from 0.1 cm to 2.5 cms in diameter



Figure 2

Figure 2: H & E x 400 showing multiple cystic spaces lined by flat endothelial cells and filled with eosinophilic material



DISCUSSION

Lymphangiomas are rare, sporadic disorders characterized by cystic lymphangiomas involving multiple organs^{2,3}. The most common sites involved are skin of the face, neck and the axillary region. Involvement of visceral organs like kidney, bones, liver and soft tissues are extremely rare. When multiple organs are involved it is termed as systemic cystic lymphangiomas². In contrast to single lymphangiomas, this form has been reported to be progressive and to have poor prognosis⁴. The cystic cavities vary in size, from microscopic to several centimetres in diameter³. The formation of cystic lymphangiomas is thought to be due to sequestration or obstruction of lymphatic vessels during embryogenesis. The exact cause is unknown.

Lymphangiomas of the spleen are rare and can present as solitary nodules, multiple nodules or diffuse growth. More often they consist of multiple cysts of varying sizes lined by flat endothelial cells, which contain eosinophilic serous fluid. The septations are made of fibrous connective tissue with rare vessels⁵.

Clinically, the disorder can be classified as systemic, hepatosplenic, or purely splenic or hepatic.

Lymphangiomas with isolated involvement of the spleen are very rare^{6,7,8}. It is seen equally among both sexes. It has been described to occur from birth to the seventh decade of life; however, isolated solid organ involvement seems to occur in patients older than 25 years of age. Initial symptoms are usually due to the mass effect caused by the enlarged organs.

At ultrasonography the cysts are hypoechoic with internal debris. The septa are hyperechoic and may show detectable vasculature at colour doppler study⁶. If the cystic spaces are small and below the resolution of the ultrasound technique, then portions of the mass may appear hyperechoic due to the existence of numerous reflecting interfaces. This may erroneously lead to misdiagnosis of lymphoma as in our case.

The differential diagnosis of cystic lymphangiomas includes true splenic cysts, mesothelial cysts, echinococcosis and very rarely cystic hamartoma¹. True splenic cysts will have definite epithelial lining. They may present with septations but usually the multicystic cavities are larger than in our case. Parasitic cysts most commonly due to *Echinococcus granulosus* may be indistinguishable from any other multicystic mass; therefore they may resemble cystic lymphangiomas. Any patient with a cystic splenic mass should have a serologic test for echinococcosis. Cystic hamartomas are very rare in the spleen, and occur more frequently in the liver in the paediatric age group⁷. Although cystic lymphangiomas of spleen are a benign entity, one case of malignant lymphangioma was also reported⁹. The malignant degeneration was suggested by the increased proportion of solid areas as in ovarian epithelial neoplasms. In their patient, the endothelial cells had formed papillary excrescences within some of the cystic spaces, and solid foci in other regions of the mass.

The normal evolution of splenic lymphangioma is variable. Small lesions are often incidentally detected. Larger lesions may increase in size and cause the compression of other organs or rupture even after minor trauma. Treatment typically consists of fenestration of large cysts or surgical resection of cystic organs. For larger splenic cysts, splenectomy is advantageous, as the risk of rupture is very high⁸. Splenectomy tends to result in relief of symptoms and in resolution of anaemia and thrombocytopenia in patients with hypersplenism. Cystic lymphangiomas of the spleen, even though rare, should be considered in the differential diagnosis of massive splenomegaly even among the elderly as it is a fully treatable disorder.

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