

Case Report

LYMPHANGIOMATOSIS OF SPLEEN AND MEDIASTINUM

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ABSTRACT

Lymphangiomas are uncommon neoplasms with clinical manifestations ranging from insignificant incidental findings to large, symptomatic cystic masses. Though they can involve any organ of the body, but common sites include soft tissues of neck, axilla, mediastinum and retroperitoneum. A rare association of lymphangiomatosis of spleen and mediastinum in a 13 year old girl presenting with pain abdomen is being reported.

Key Words: *Lymphangiomatosis, Spleen, Mediastinum, Splenectomy*

INTRODUCTION

Lymphangiomatosis is a condition in which there are lymphangiomas in more than one organ. Lymphangiomas may be found in soft tissue of neck, liver, mediastinum, lungs, spleen, or retroperitoneal organs (Dan *et al.*, 2005). Lymphangiomatosis involving spleen and mediastinum in the same patient is very rare (Harada *et al.*, 1994). Although literature is flooded with lymphangiomas of various organs cited above but association of spleen and mediastinum in a single case is very rare. Keeping in view the rarity of this association this case is being reported.

CASES

A 13 years old female presented with pain in left hypochondrium for last 6 months. There was no history of fever or any respiratory difficulty. On physical examination spleen was palpable by 4 fingers below the left costal margin. Hematological investigations were within normal limits. Ultrasonography of abdomen revealed an enlarged spleen, riddled with cysts of various sizes. X-ray chest showed a soft tissue mass in superior mediastinum. CT scan thorax revealed a multicystic lesion measuring 12x6cm in the anterior part of superior mediastinum (Figure 1). CT abdomen showed enlarged spleen with multiple cystic lesions (Figure 2). As patient was asymptomatic for mediastinal lesion, splenectomy was carried out in view of large splenic size and pain abdomen. Excised spleen weighed 1500 gram and measured 16x10x6cm. Cut section showed numerous unilocular to multilocular cysts of varying sizes involving entire parenchyma of spleen. Cysts contained clear pale yellowish fluid. Microscopic examination revealed fibrous wall lined by endothelial cells. Patient had an uneventful postoperative period. She is on follow up for her mediastinal lesion, surgery for which was refused by her parents.

DISCUSSION

Lymphangiomatosis is an uncommon condition that may affect any organ diffusely or in the form of solitary cysts. These are classified as lymphangioma simplex, cavernosum, cysticum and mixtum (Brown *et al.*, 1986). Majority of cases are young and may remain asymptomatic. The present case however was symptomatic because of splenic mass.

Lymphangiomas in mediastinum can present as restrictive and obstructive impairment of pulmonary function. These can be found in upper, middle and posterior mediastinum and have to be differentiated from other lesions in respective mediastinum (Harada *et al.*, 1994). Lymphangiomas of spleen are infrequent hamartomas of spleen with clinical manifestations ranging from insignificant incidental findings to large symptomatic mass requiring surgical intervention (Morgenstern *et al.*, 1992).

Lymphangiomatosis pose a challenge in diagnosis as they largely remain asymptomatic and tissue diagnosis reveals varied pictures. Diagnosis of mediastinal or splenic lymphangiomas is based on X-rays, ultrasonography, CT scan, and MRI. Magnetic resonance combined with magnetic resonance angiography

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is the diagnostic method of choice in the generalized lymphangiomatosis extending to more organs (Ikezoe *et al.*, 1992).

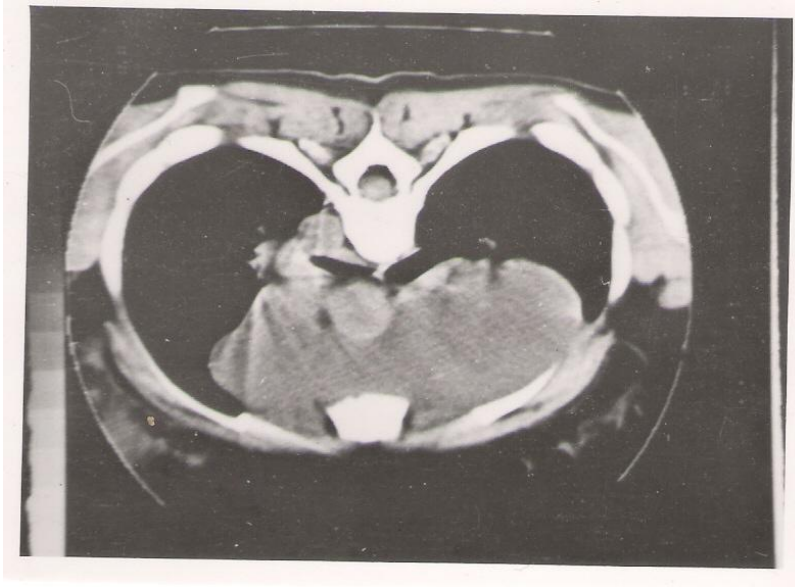


Figure 1: CT scan of thorax showing a large low density nonenhancing cystic area in the anterior mediastinum abutting great vessels

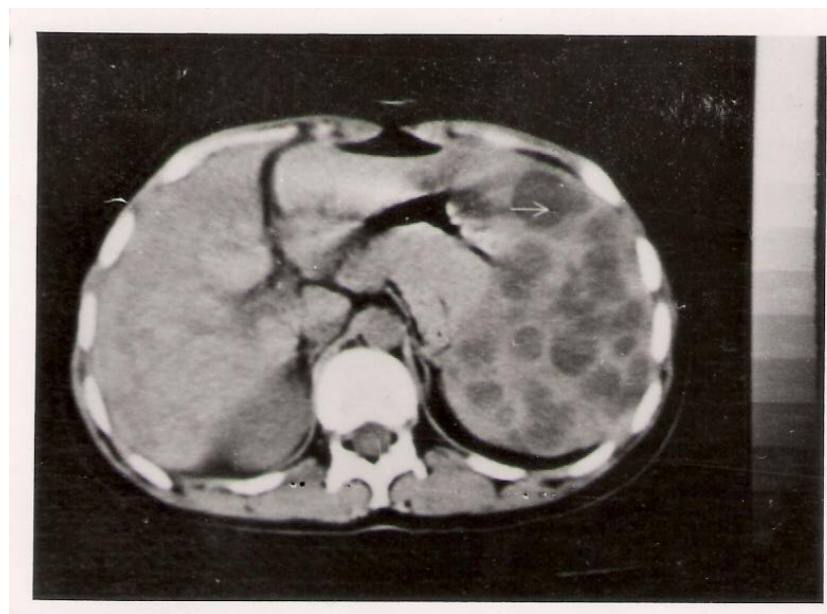


Figure 2: CT scan of abdomen showing multiple cystic lesions in spleen

Large cystic, symptomatic masses need excision. Extensive involvement of spleen may cause symptomatic splenomegaly necessitating splenectomy as in the present case. However, complete excision of lymphangiomas at other sites like neck, mediastinum may not be possible without jeopardizing vital structures because of their tendency towards infiltration. In such cases conservative measures like sclerotherapy with bleomycin, OK-432, laser therapy and radiotherapy have been reported to give promising results (Johson *et al.*, 1986; Landthaler *et al.*, 1990; Ogita *et al.*, 1994).

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