

Solitary Lymphangioma Of The Spleen: A Case Report

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Citation

S Bhorawal, P Garg, S Singla, V Rathee, S Verma, S Mittal, S Aggarwal, A Narang. *Solitary Lymphangioma Of The Spleen: A Case Report*. The Internet Journal of Gastroenterology. 2012 Volume 11 Number 2.

Abstract

Solitary Lymphangioma of spleen are rare benign tumors and found in pediatric or young population. They are commonly seen in neck and axillary region. Abdominal Lymphangioma are found in less than 5% of cases and less than 100 cases of lymphangioma spleen are reported. It is rare to found these tumors in elderly. Here in a case of Lymphangioma of spleen in 60 years old female is reported with a review of literature.

INTRODUCTION

Lymphangioma are congenital malformations of vascular origin.¹ They are commonly seen in pediatric age group and 95% of lymphangioma occur in cervical and axillary region. Splenic lymphangioma are rare and can be found as isolated splenic lymphangioma or associated with lymphangiomatosis with multi organ involvement, this condition was first described by Redenber in 1828¹. These are benign lesions and formed by either blocking or agenesis of lymphatic channels. Treatment remains surgical in form of complete excision by total splenectomy.

CASE REPORT

A 60 years old female presented to surgical clinic of Pt. B.D.Sharma P.G.I.M.S. Rohtak with complaints of progressively enlarging swelling in the abdomen for past 10 years along with on and off pain in the abdomen for the past 6 years. On examining the abdomen was distended with an intra-peritoneal lump of size 17 x 18 centimeters. It was firm in consistency and the lump could be moved in all directions. Her routine investigations were within normal limits and ultrasound scan showed a large cystic lesion of size 16x15 centimeters in the lower abdomen and pelvis with necrotic component in it. CECT abdomen revealed a well-defined soft tissue density lesion with large soft tissue component measuring 12x14.8x17.3 centimeters with heterogeneous enhancement. As the mass was moving in all directions and considering the age of the patient a diagnosis of ovarian cyst was made and the patient was planned for elective laparotomy. Intraoperative bilateral ovaries were found to be normal and a huge mass arising from the spleen

was noted. Splenectomy was done and histopathology revealed a rare diagnosis of Lymphangioma of the spleen.

Figure 1

Figure 1 Huge splenic cyst found during laparotomy



Figure 2

Figure 2 Cutting the specimen revealed the cystic nature of the spleen



Figure 3

Figure 3 CECT cross section showing enlarged cystic spleen



Figure 4

Figure 4 CECT recon image showing extent of splenic cyst



DISCUSSION

Splenic cysts are rare with a reported incidence of 0.07% in a 42327 autopsy series.² 75% of them are parasitic while rest are non-parasitic.² Parasitic cysts are formed in hydatid disease due to *Echinococcus granulosus* infestation, which itself is uncommon as spleen is affected in only 3.5% of cases.³ Non parasitic cysts are further classified as primary or true cysts and secondary or false cysts as they lack any cellular wall. They can result from trauma or hemorrhage into the organ. True cysts have a definite cellular lining of epithelium i.e. epidermoid, dermoid or mesothelial or lined by endothelium i.e. lymphangioma or angioma.⁴

Lymphangioma of spleen is rare benign malformations of vascular origin composed of endothelial lined cysts containing lymph.⁵ This was first described by Fink in 1885.⁵ These are rare and less than 100 cases are described in literature.⁶ 75% of lymphangioma are found in cervical region, 20% in axillary region and rest 5% can be found at various sites like mediastinum, kidney and bones.^{7,8} Abdominal lymphangiomas rarely occur in pancreas, liver, spleen, retroperitoneum, mesentery and Gastrointestinal tract.^{5,9-13} Lymphangioma of spleen can occur sporadically or as a part of generalized lymphangiomatosis, a condition in which lymphangiomas develops in multiple organs.⁷ Splenic lymphangioma are more common in females and 80% to 90% are discovered till second decade of life.¹⁴ Splenic

involvement occurs in solitary nodular, multiple nodular or diffuse forms.¹⁴ They are classified as capillary, cavernous or cystic and are supposed to arise after congenital obstruction of lymph flow and subsequent dilatation of lymphatic channels or as a result of agenesis of lymphatics resulting in lymphangiectasis from lack of communication to lymphatic channels.^{5,14,15} They arise from subcapsular location which is the site of distribution of splenic lymphatics. The cysts are lined by endothelium and are filled with a proteinaceous fluid.^{7&14}

Clinically the presentation depends upon the size of the lesion. Small lymphangioma are asymptomatic but bigger ones produce symptoms by compressing the adjacent structures like stomach, kidney or duodenum.¹³ They can present with acute pain abdomen due to rupture or intracystic hemorrhage. Hypertension due to compression of renal artery has also been reported.¹⁶ Symptoms can be due to isolated splenic involvement or due to other organs involved in generalized lymphangiomatosis.⁸ Although considered benign they can be locally invasive.¹⁴ On CT or MRI they are thin walled cystic lesions and are low signal intensity on T₁ weighted images and high signal intensity on T₂ weighted images however intracystic hemorrhage or high protein content of the fluid can result in High signal intensity in T₁ weighted image.¹⁷ They are considered benign but treatment remains surgical in form of splenectomy as they have a high recurrence rate if excision is not complete.^{18,19}

CONCLUSION

Cystic lymphangioma are to be considered in the differentials of massive splenomegaly

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