

# Cystic Lymphangioma of Spleen: A Case Report

**Nisha Marwah<sup>1</sup>,**  
**Monika Sangwan<sup>2</sup>,**  
**Megha Ralli<sup>3</sup>,**  
**Renuka Verma<sup>4</sup>,**  
**Swagatika Samal<sup>5</sup>**

<sup>1</sup>MD Professor, <sup>2</sup>MD Senior Resident, <sup>3</sup>PG Junior Resident, <sup>4</sup>PG Junior Resident, <sup>5</sup>PG Junior Resident,  
Department of Pathology, Pt. B.D. Sharma PGIMS Rohtak, Haryana

**Correspondence Author:** Dr. Megha Ralli, 30/32 West Patel Nagar, New Delhi - 110008,  
Phone: 07206191047. E-mail: drmegharalli@gmail.com

## Abstract

Lymphangioma, a very uncommon benign neoplasm, is seen in children and rarely in adults. It most commonly involves the neck (75%) and axilla (20%). It can occur sporadically in mediastinum, retroperitoneum, and internal organs. Splenic lymphangioma is a very rare condition and is usually found incidentally. The rate of malignant transformation is low, its prognosis is good. We report a case of cystic lymphangioma of spleen in a 47 year-old female presenting with abdominal pain. This case emphasizes on rarity of the case at this age and the differential diagnosis with other cystic proliferation of spleen in particular hydatid disease.

**Keywords:** Cystic lymphangioma, Benign neoplasm, Spleen

## INTRODUCTION

Cystic lymphangioma is benign neoplasm composed of malformation of lymphatic system.<sup>1</sup> They generally occur under the age of two years with no difference in incidence between males and females.<sup>2</sup> It most commonly involves the neck (75%) and axilla (20%). They can occur sporadically in mediastinum, retroperitoneum, and internal organs. Splenic lymphangioma is a very rare condition and is usually found incidentally.<sup>3</sup> Parasitic cysts are most common cystic proliferations of spleen. Non parasitic cysts are classified as primary or true cysts and pseudocysts. Amongst the true cysts, hemangioma are most common ones.<sup>4</sup> In the majority of cases, lymphangiomas have an asymptomatic course and despite the use of modern imaging techniques, often makes preoperative diagnosis difficult. Lymphangioma of spleen is extremely rare. To prevent complications such as infections, torsion, enlargement etc., total resection of tumor is done. The rate of malignant transformation is low, its prognosis is good. Here, we report a case of 47 year-old female presenting with abdominal pain.

## CASE REPORT

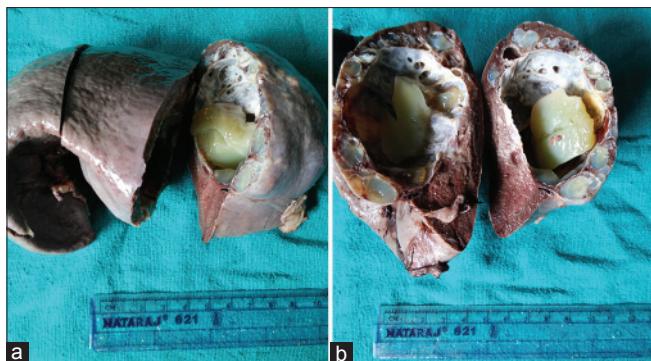
A forty seven years old female presented with pain in abdomen since one month. Physical examination revealed enlarged liver till L5 subcostal region. Peripheral blood count, coagulation studies, liver and kidney function tests were all within normal limits. USG abdomen revealed

large multilocular cystic mass in spleen. Clinically and radiologically it was diagnosed as hydatid cyst of spleen. A splenectomy was done and sent for histopathological examination.

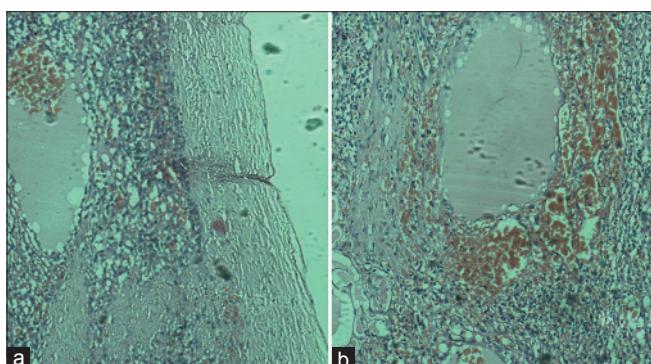
Grossly, the spleen weighed 500 gm and measured 13 × 8 × 2 cm. Cut section revealed multiple variable sized cystic cavities involving almost the whole spleen. The largest cavity measuring 8 × 6 × 4 cm. The cavities were filled with gelatinous mucoid like material (Figure 1). On microscopic examination, the cysts were lined by endothelial cells and filled with acellular eosinophilic fluid (Figure 2). The cyst wall consisted of fibrous tissue with occasional calcification. Immunohistochemistry revealed D2-40 (Figure 3) and CD 31 positivity in endothelial lined cells and no or weak positivity with CD34. So the final diagnosis of cystic lymphangioma was made. The postoperative course was uneventful and the patient was discharged on the seventh day after the splenectomy. The patient made complete recovery and free of disease two months postoperatively.

## DISCUSSION

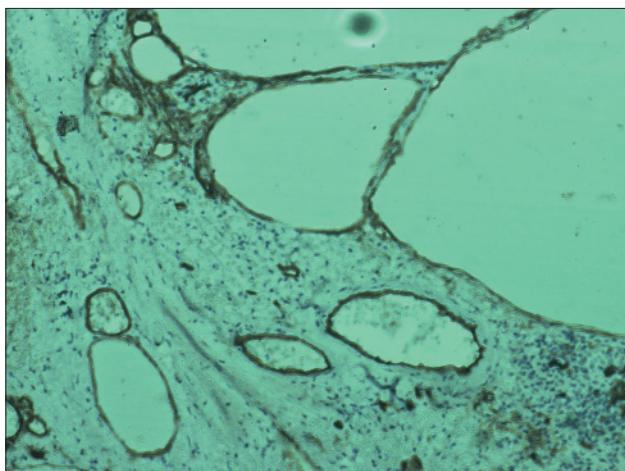
In 1885, Frink reported the first lymphangioma in the spleen.<sup>5</sup> Cystic lesions of spleen include parasitic and non parasitic cysts. Among parasitic ones, echinococcal disease represent 50-80% of the cases.<sup>6</sup> Non parasitic cysts are classified as primary or true cysts and pseudocysts. Most



**Figure 1: Spleen showing multiple cystic cavities filled with gelatinous mucoid like material**



**Figure 2: (a) Section showing fibrous splenic capsule over the cystic cavities (H&E, 100X), (b) Section showing cysts lined by flat endothelial cells and filled with acellular proteinaceous fluid (H&E, 200X)**



**Figure 3: Section showing D2-40 positivity in endothelial lined cells (IHC, 200X)**

cysts are post traumatic pseudocysts and true cysts are rare including hemangioma, lymphangioma, epidermoid and dermoid cysts.<sup>4</sup>

Lymphangioma is infrequently seen in mediastinum, adrenal gland, kidney, bone, omentum, gastrointestinal tract, retroperitoneum, spleen, liver and pancreas.<sup>2,5</sup>

Opinions regarding histogenesis vary and a conclusive consensus has not been achieved.<sup>7,8</sup> They are considered to be developmental abnormalities due to either obstruction or due to obstruction leading to lymphangiectasias. The cause of obstruction of lymphatic system could also be due to bleeding or inflammation resulting in lymphangioma.<sup>9</sup>

Lymphangioma can be seen in the spleen alone, or it can be associated with multivisceral involvement; when diffuse it is termed systemic cystic angiomyomatosis.<sup>10</sup> Generally, lymphangioma is divided into capillary, cavernous and cystic types. The cystic type is the most common type.<sup>1</sup>

Patients with splenic lymphangioma present with upper left quadrant pain along with fever, nausea, vomiting and weight loss. Pain in left hypochondriac region was the presenting feature in our patient. Because of similarity of signs and symptoms clinically it is often confused with hydatid disease. Radiological findings are also not conclusive. Hence, histopathological examination is important in making diagnosis.<sup>11</sup> Hemangiomas are also an important differential diagnosis. Lymphangiomas show presence of flat endothelial lined spaces filled with eosinophilic proteinaceous material instead of blood and located in subcapsular area or larger trabeculae of spleen where lymphatics are normally concentrated while random localization is seen in case of hemangiomas. Immunohistochemically, the endothelial cells of lymphatic tissue show positivity for endothelial receptor-1, vascular endothelial growth factor-3, prox-1, and monoclonal antibody D2-40.<sup>3</sup>

Complications of splenic cysts include rupture with peritonitis as well as invasive hemorrhage, infection, abscess formation, pleural effusion or empyema.<sup>4</sup> Splenectomy is choice of treatment. Conservative management like aspiration, drainage and sclerosis are associated with high risk of recurrence,<sup>1</sup> while the prognosis is good.<sup>12</sup>

## CONCLUSION

Though cystic lymphangiomas are uncommon entity, they should be considered in differential diagnosis of various types of splenic cystic masses in patients of any age presenting with abdominal lump, pain, nausea and fever.

## REFERENCES

- Chung SH, Park YS, Jo YJ, Kim SH, Jun DW, Son BK. Asymptomatic lymphangioma involving the spleen and retroperitoneum in adults. *World J Gastroenterol*. 2009;15:5620-3.
- Kim DH, Byun JN, Jang JY. Cystic lymphangioma involving the mesentery and the retroperitoneum: A case report. *J Korean Radiol Soc*. 2005;52: 347-50.

3. Kim MJ, Cho KJ, Han EM and Lee YJ. Splenic Lymphangioma- A Report of Three Cases. *The Korean J Pathol.* 2002;36:416-9.
4. Wahab MA, Elenin AA, Sultan A, Ghawalpy NE, Ezzat F. Lymphangiomatous cysts of the spleen. Report of 3 cases and review of the literature. *Hepatogastroenterology.* 1998; 45: 2101-4.
5. Chang CH, Hsieh CB, Yu JC, Jan CI. A case of lymphangioma of the spleen. *J Med Sci* 2004; 24: 109-12.
6. Gidaro GS. Cystic splenic disease of surgical interest. *G Chir.* 1997; 18:555-9.
7. Silverman ML, Livolsi VA. Splenic hamartoma. *Am J Clin Pathol* 1978; 70: 224-9.
8. Sakuma T. Lymphangiomatous hamartoma of the spleen. *Critical Comentary. Pathol Res Pract* 1995; 191:1168.
9. Vergheese BG, Kashinath SK, Kanth RR. Lymphangioma of the spleen-A rare tumor rarely seen in adult: A case report and a comprehensive literature review. *Euroasian J Hepato- Gastroenterol* 2013;3:64-9.
10. Seckler SG, Rubin H, Rabinowitz JC. Systemic cystic angiomas. *Am J Med* 1964; 37:976-86.
11. Vezzoli M, Ottini E, Montagna M, Fianza AL, Paulli M, Rosso R et al. Lymphangioma of the spleen in an elderly patient. *Haematologica.* 2000;85:314-7.
12. Witzel K, Kronsbein H, Pleser M, Hunfeld H, Rumpf KD. Intraabdominal cystic lymphangioma in childhood. Report of 2 cases. *Zentralbl Chir.* 1999; 124: 159-62.

**How to cite this article:** Nisha Marwah, Monika Sangwan, Megha Ralli, Renuka Verma, Swagatika Samal "Cystic Lymphangioma of Spleen: A Rare Report". *Int J Sci Stud.* 2014;2(1):40-42.

**Source of Support:** Nil, **Conflict of Interest:** None declared.