Case Report

DOI: 10.7860/JCDR/2016/19989.8142

A Giant Retroperitoneal Lymphangioma: A Case Report

Surgery Section

PRABHU HUBLI[1], MUDDASETTY ROHITH[2], BASAVARAJU M SACHIN[3]

ABSTRACT

Lymphangioma is a congenital lesion of childhood. They usually present in head and neck region. Intra-abdominal lymphangioma in an adult is a rare lesion which poses diagnostic difficulty. Although asymptomatic, they may present with an acute abdomen. Here, we report a rare case of huge asymptomatic retro-peritoneal lymphangioma who underwent complete surgical excision with an uneventful postoperative period.

Keywords: Lymphatic cysts, OK-432, Sclerotherapy

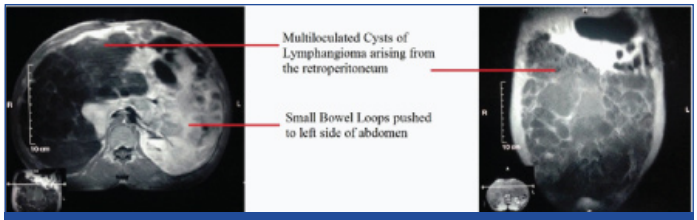
CASE REPORT

A 36-year-old male patient presented with a mass in abdomen since a month which had come to his notice when he had visited a physician for upper respiratory tract infection. He was a farmer by occupation and belonged to lower middle socioeconomic status of north east Karnataka region. The patient had no other symptoms of gastrointestinal obstruction in form of vomiting, constipation, distension of abdomen or pain abdomen; lower limb oedema or urinary symptoms.

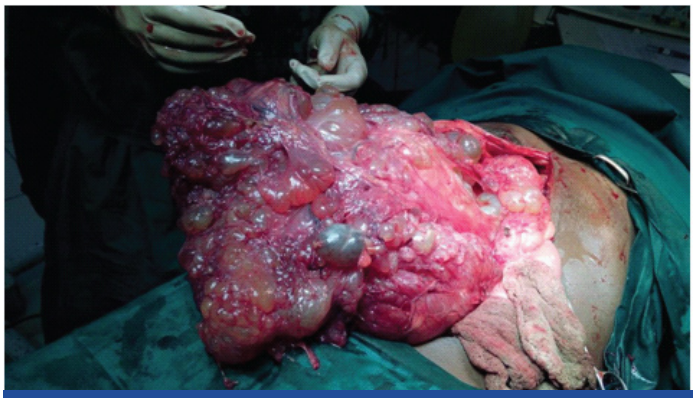
After obtaining an informed written consent from the patient for publishing/reporting the case including the pictures, he was examined. Abdomen appeared flat on inspection. A mass was noted in the right side of the abdomen approximately 30X40cm in size, nodular surface, well defined irregular borders and firm in consistency and non-tender. There was no hepatosplenomagaly, no ascites, bowel sounds were normal. Other systemic examination was normal.

Ultrasonography of abdomen showed a large, multicystic lesion in the right side of abdomen, displacing the bowel to the left. MR imaging of abdomen revealed, a large multiloculated cystic lesion in the right side of abdomen and in the mid line scalloping the under surface of the right lobe of liver [Table/Fig-1].

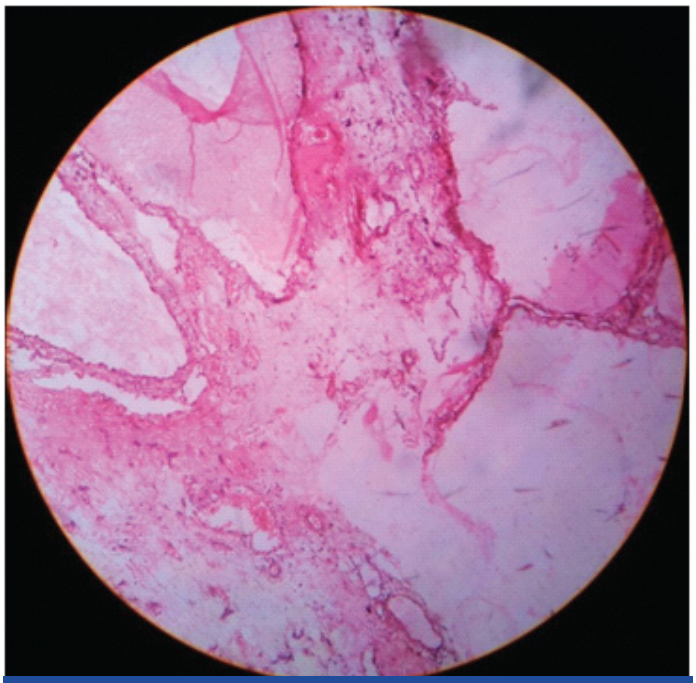
Patient was posted for an exploratory laparotomy. It was a multiloculated cystic swelling likely to be lymphangioma, occupying the right side of the abdomen [Table/Fig-2]. The bowel loops were shifted to the other side. The lesion was found to be arising from the retro peritoneum on right side. The kidney and ureter were normal. The lesion was completely excised. The lesion was around 2.5 kilograms in weight. The histopathological examination of the lesion showed large dilated cysts lined by flattened endothelial cells suggestive of cystic lymphangioma [Table/Fig-3]. Postoperative stay in hospital was uneventful. Patient came for follow-up at 3, 6 and 9 months post-surgery and had no clinical or radiological recurrence.



[Table/Fig-1]: MR imaging of abdomen showing multiloculated lesion occupying the right side of abdomen and shifting the bowel to left side.



[Table/Fig-2]: Intraoperative picture of lesion.



[Table/Fig-3]: Histopathological image (Haematoxylin and eosin staining with 10x magnification) of specimen showing endothelial lined cystic spaces.

DISCUSSION

Lymphangioma are rare benign congenital lesions. They are commonly located in the head and neck region, in paediatric population. Lymphangioma in an adult is an uncommon presentation.

Intra-abdominal lymphangioma is a rare entity. In the abdomen, lymphangioma occur most commonly in the mesentery, followed by the omentum, mesocolon and retroperitoneum [1]. Retro-peritoneal lymphangioma form less than 1% of the cases [2]. Thus being a rare case that we came across, reporting it would be helpful in enriching our knowledge and others.

Lymphangioma is thought to be congenital in origin, arising from the lymphatic sequestrations which fail to communicate with the normal lymphatic channels [1]. Fibrosis, trauma and neoplasms also being postulated to cause lymphangioma [2]. The incidence of these lymphangioma is said to be up to 5% in paediatric population. Among them 3-9% constitute intra-abdominal lymphangioma [3].

Most of these present with asymptomatic mass in the abdomen. Rarely they may present as acute abdomen with intestinal obstruction, haemorrhage into the cyst, torsion, inflammation [4]. Magnetic resonance imaging is the investigation of choice. Ultrasonography, computed tomography and magnetic resonance imaging show thin walled, multiseptate, cystic lesion displacing the intestines. Imaging also help to rule out other cystic lesions arising from kidney or liver [5,6]. Final diagnosis is by pathological examination which shows dilated lymphatic spaces. The spaces are filled with eosinophilic material and lined by endothelial cells [5,7].

Definitive treatment is complete excision [8]. Resection of the involved bowel may be needed. With complete excision, results are excellent. However, incomplete excision has high risk of recurrence [9].

Gayen R et al., Fanaei SA et al., Tripathi M et al., have reported similar cases in which complete excision was supported as the preferred treatment [4,9,10]. Fanaei SA et al., also experienced recurrence of the lesion following doubtful complete excision [9].

Other modality of treatment includes injection sclerotherapy using alcohol or acetone [11]. The risk of damage to normal tissue and high chance of recurrence are problem with this modality. Residual lesions after surgery are being tried with OK-432, a biological response modifier with variable results [12].

CONCLUSION

Retro peritoneal lymphangioma is a rare slowly growing lesion, usually asymptomatic at presentation. It requires a thorough evaluation with CT/MRI for diagnosis. Complete surgical excision is the ideal treatment of choice. Definitive diagnosis is only pathological examination.

REFERENCES

[1] Wani I. "Mesenteric Lymphangioma in Adult": A case series with a review of the Literature. *Dig Dis Sci*. 2009;54(12):2758-62.
[2] F. M. Enzinger and S. W. Weis, "Tumors of lymph vessels," in Soft Tissue Tumors, pp. 679–700, Mosby-Years Book, St. Louis, Mo, USA, 1995.View at Google Scholar
[3] Geoff AJ, Sohn TR, Cameron JL, Frederic AB, Ralph HH, Kurt AC: Abdominal lymphangiomas in adults: *J Gastrointest Surg*. 2006;10(5):746-51.
[4] Gayen R, Mahata M, Dasgupta S, Dasgupta J. Giant retroperitoneal cystic lymphangioma- a case report with review of literature. *Journal of Dental and Medical Sciences*. 2015;14(2):69-71.
[5] Davidson AJ, Hartman DS. Lymphangioma of the retroperitoneum. CT and sonographic characteristics. *Radiology*. 1990;175:507-10.
[6] McDonald N, Choe K, Wang J, Sussman J. Radiological Case: cystic lymphangioma. *The Journal of Practical Medical Imaging and Management*. 2014.
[7] Kurtz MD, Heimann TM, Beck AR, Holt J. Mesenteric and retroperitoneal cysts. *Ann Surg*. 1986;203:109-12.
[8] Bezzola T, Buhler L, Chardot C, Morel P. Surgical therapy of abdominal cystic lymphangioma in adults and children. *J Chir (Paris)*. 2008;145(3):238-43.
[9] Fanaei SA, Ziaee SA. Retroperitoneal cystic lymphangioma: Case Report. *Surgical Science*. 2011;2:209-11.
[10] Tripathi M, Parshad S, Karwasra RK, Gupta A, Srivastva S, Sarwal A. Retroperitoneal Lymphangioma in an adult: A case report of a rare clinical entity. *Case Reports in Surgery*. 2015;2015:732531, 1-4.
[11] Stein M, Hsu R, Schneider P, Ruebner B, Mina Y. Alcohol ablation of a mesenteric lymphangioma. *J Vasc Radiol*. 2000;11(2):247-50.
[12] Tokuhara K, Hamada Y, Watanabe K, Tanano A, Takada K, Sato M. A case of huge mesenteric lymphangioma managed by laparoscopy-assisted subtotal resection with sclerosing therapy. *Japanese Society of Pediatric Surgeons*. 2003;39:90–95.

PARTICULARS OF CONTRIBUTORS:
1. Professor, Department of Surgery, Vijayanagar Institute of Medical Sciences, Bellary, India.
2. Post Graduate, Department of Surgery, Vijayanagar Institute of Medical Sciences, Bellary, India.
3. Post Graduate, Department of Surgery, Vijayanagar Institute of Medical Sciences, Bellary, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Muddasetty Rohith,
Room Number 4, New PG Boys Hostel, VIMS Campus, Bellary-583104, India.
E-mail: rohith.m.setty@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Mar 10, 2016**
Date of Peer Review: **Apr 15, 2016**
Date of Acceptance: **Jun 10, 2016**
Date of Publishing: **Jul 01, 2016**