CASE REPORT

UNUSUAL PRESENTATION OF RETROPERITONEAL LYMPHANGIOMA IN AN ADULT

Shirish Paul Ganta¹, S. Venkateshwar Rao², V. Sivaleela³, N. Kiran Raju⁴, V. N. Narvekar⁵

HOW TO CITE THIS ARTICLE:

Shirish Paul Ganta, S. Venkateshwar Rao, V. Sivaleela, N. Kiran Raju, V. N. Narvekar. "Unusual Presentation of Retroperitoneal Lymphangioma in an Adult". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 13, February 12; Page: 2231-2234, DOI: 10. 14260/jemds/2015/322

ABSTRACT: Lymphangioma is a rare benign congenital malformation of lymphatic system. We present a case of 32 year old male with rare retroperitoneal lymphangioma extending to mediastinum.

KEYWORDS: Lymphangioma, retroperitoneum, mediastinum, ultrasound, computed tomography, MRI.

CASE REPORT: A 32-years old male presented with lower abdominal pain for 10 days. On clinical examination no abnormality was detected.

Ultrasound showed irregular multiseptated cystic lesion with posterior acoustic enhancement in left iliac fossa (LIF), seen extending along iliac vessels, pre and para-aortic areas upto diaphragmatic hiatus (Fig. 1 A, B). Provisional diagnosis of retroperitoneal lymphangioma was made. CECT abdomen with oral and intravenous (I.V) contrast showed a homogenous hypodense irregular lesion in LIF causing mild displacement of bladder to right. The cystic areas are noted extending along iliac vessels, aorta and superiorly into mediastinum (Fig. 2). The lesion showed cystic density. Post contrast study shows no enhancement (Fig. 3).

MRI was performed and the lesion showed homogeneously hypointense signal on T1-weighted images and hyper intense on T2-weighted images (Fig. 4).

Biopsy of the lesion proved to be lymphangioma.

Our final diagnosis based on imaging & histopathological report is retroperitoneal lymphangioma with extension into mediastinum superiorly.

DISCUSSION: Embryologically the lymphatic system is formed by the 8th week of intrauterine life and arises from six primitive sacs; paired jugular sacs located lateral to the jugular veins, an unpaired retroperitoneal sac at the root of mesentery, an unpaired cysterna chyli dorsal to aorta and adrenal glands and paired posterior sacs located in relation to sciatic veins. The postulated mechanism of formation of lymphangioma is early developmental sequestration of lymphatic vessels, that fail to establish connection with normal draining vessels at about 14-20 weeks of intrauterine life. They become markedly dilated under the pressure effect of accumulating lymph.

Lymphangioma is a benign congenital malformation of lymphatic channels, which has been classified into 3 groups: capillary, cavernous and cystic depending on the size of lymphatic space.^{1,2} Histologically, it is characterized by a flat epithelial endothelium and a wall alternatively containing lymphoid tissue, small lymphatic spaces, smooth muscle, and foam cells.³ Lymphangioma is most commonly found in paediatric patients⁴ with slight male preponderance. The most frequently involved regions are the neck (75%) and axilla (20%).^{5,6} Less than 5% of lymphangiomas are diagnosed intra-abdominally^{4,7} and they have been reported in mesentery,⁸ gastro-intestinal tract,⁹ spleen,¹⁰ liver,¹¹ and pancreas.⁴ They are infrequently encountered in the retro peritoneum.

CASE REPORT

Patients with retroperitoneal lymphangioma are usually asymptomatic and the masses are often found incidentally by imaging technique or surgery for other purposes.^{1,2} Occasionally patient will present with symptoms when the tumour is large, or when infection, haemorrhage, torsion, or rupture occur.²

On ultrasound imaging, retroperitoneal lymphangioma may appear as a multi loculated hypoechoic or anechoic fluid filled mass with internal septations. CT shows a well circumscribed, multi loculated, homogenous cystic mass. Internal septations and thin wall may enhance after intravenous contrast injection. CT has the advantage of providing more information on the size, extent, composition, and relationship with important adjacent structures. Any compression onto these organs can also be well demonstrated.

On MR imaging, lymphangioma displays homogenous hyper intense signals in T2- weighted images and low intensity signals in T1-weighted images.

In our case, the lesion was not only at a rare site that is retroperitonem but also crossed midline to involve whole abdomen and also extending superiorly into mediastinum, hence it is one of the very uncommon presentation.

Differential diagnosis include benign lesion such as simple cyst and pseudocyst,⁴ and necrotic neoplastic growth such as liposarcoma, leiomyosarcoma, fibrosarcoma, teratoma,² or metastatic lymphadenopathy and in Indian context multi loculated ascitis due to high prevalence of tubercular abdomen. The final diagnosis requires histopathological examination, usually achieved by excision or biopsy. Complete surgical excision is the treatment of choice but may be difficult in some patients due to local invasiveness. Some surgeons may advocate internal peritoneal marsupialisation.⁵ Aspiration, drainage, and irradiation of the lymphangioma give a poor result.⁵ Treatment by argon beam ablation and sclerotherapy have also been reported in a patient with a life-threatening total abdominal lymphangiomatosis.¹²

REFERENCES:

1. IYF Chan, J Khoo. Retroperitoneal lymphangioma in an adult. J HK Coll Radiol 2003; 6: 94-96.
2. Bonhomme A, Broeders A, Oyen R. H, Stas M, DeWever I, Baert A. Cystic lymphangioma of the retroperitoneum. Clin Radiol 2001; 56: 156-158.
3. Takiff H, Calabria R, Yin L, Stabile BE. Mesenteric cysts and intraabdominal lymphangioma. Arch Surg 1985; 120: 1266-1269.
4. Koenig TR, Loyer EM, Whitman GJ, Raymond AK, Charnsangavej C. Cystic lymphangioma of the pancreas. AJR 2001; 177: 1090.
5. De Perrot M, Rostan O, Morel P, Le Coultre C. Abdominal lymphangioma in adults and children. Br J Surg 1998; 85: 395-397.
6. Kosir MA, Sonnino RE, Gauderer MWL. Paediatric abdominal lymphangioma: a plea for early diagnosis. J Pediatr Surg 1991; 26: 1309-1313.
7. Yang CS, Wu MS, Wang HP, Shun CT, Lin JT. Disseminated cystic lymphangiomatosis presenting with acute abdomen: report of a case and review of the literature. Hepatogastroenterology 1999; 46: 196-198.
8. Bliss DP Jr, Coffin CM, Bower RJ, Stockmann PT, Ternberg JL. Mesenteric cysts in children. Surgery 1994; 115: 571-577.

CASE REPORT

9. Shigematsu A, Iida M, Hatanaka M, et al. Endoscopic diagnosis of lymphangioma of the small intestine. *Am J Gastroenterol* 1988; 83: 1289-1293.
10. Pistoia F, Markowitz SK. Splenic lymphangiomatosis: CT diagnosis. *AJR* 1988; 150: 121-122.
11. Van steenberg W, Joosten E, Marchal G, et al. Hepatic lymphangiomatosis. Report of a case and review of the literature. *Gastroenterology* 1985; 88: 1968-1972.
12. Rothenberg SS, Pokorny WJ. Use of argon beam ablation and sclerotherapy in the treatment of a case of life-threatening total abdominal lymphangiomatosis. *J Pediatr Surg* 1994; 29: 322-323.

Fig. 1A, Ultrasound LIF showing irregular multiseptated cystic lesion along external iliac vessels. Fig. 1B, colour Doppler showing the same lesion with iliac vessels

Fig 2A: Axial NECT scan demonstrating irregular hyodense lesion adjacent to bladder. Fig. 2B, showing extension to mediastinum along aorta

Fig. 3: Axial CECT demonstrating non enhancement of lesion

CASE REPORT

Fig. 4: Axial MRI images A, T1-weighted shows homogeneously hypointense signal of lesion adjacent to bladder. B, T2-weighted shows hyperintense signal

AUTHORS:

1. Shirish Paul Ganta
2. S. Venkateshwar Rao
3. V. Sivaleela
4. N. Kiran Raju
5. V. N. Narvekar

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Radiology, Asram Medical College.
2. Associate Professor, Department of Radiology, Asram Medical College.
3. Professor, Department of Radiology, Asram Medical College.
4. Assistant Professor, Department of Radiology, Asram Medical College.

FINANCIAL OR OTHER

COMPETING INTERESTS: None

5. Professor and HOD, Department of Radiology, Asram Medical College.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Shirish Paul Ganta,
Assistant Professor,
Department of Radiodiagnosis,
Asram Medical College,
Eluru-534005, West Godvari District,
Andhra Pradesh.
E-mail: shirishpaul24@gmail.com

Date of Submission: 03/01/2015.
Date of Peer Review: 05/01/2015.
Date of Acceptance: 04/02/2015.
Date of Publishing: 12/02/2015.