

Mediastino-hepato-renal cystic lymphangiomas – diagnostic and surgical considerations

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Abstract: Cystic lymphangiomas or hygromas are rare benign vascular tumours, caused by congenital malformation of the lymphatic vessels. It appears as a progressive swelling in the head or neck of children during 2-5 years of life, yet rarely seen in the mediastinum or abdomen. Symptomatic mediastinal cystic lymphangiomas provide symptoms such as chest pain, breathlessness, cough, and dysphagia, making it difficult to differentiate from other mediastinal tumours. The tumour can become larger due to infections, inflammations, obstructions and bleedings. Chest X-ray, ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI) provide helpful information but the diagnosis appears merely after surgical resection and histological examination. Only a few cases have been reported. Hence, we report the first case of a mediastinal and asymptomatic renal and multiple hepatic cystic lymphangiomas in a 71-year-old male with respiratory symptoms and sever reduction in lung capacity. The symptoms regressed fully after surgical excision and lung diffusions capacity increased significantly.

Keywords: Mediastinum; lymphangioma; congenital; malformation

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Introduction

Lymphangioma is a rare benign tumour due to congenital lymphatic vessels abnormalities. Almost 90% of cases are diagnosed at birth or within the first 2 years of life (1-4). They appear as fluctuant, freely mobile, compressible, painless and slow growing asymptomatic masses mostly in the head and neck but occasionally in the axilla, or groin. The cysts can occur anywhere in the lymphatic channels of the body.

Case report

A 71-year-old man was referred for surgical resection of a mediastinal mass (*Figure 1*). He was diagnosed with an asymptomatic 5 cm mediastinal cyst in 2007 and the decision was follow-up. In 2013, a computed tomography (CT) scan disclosed growth of the mass to 9-10 cm. Lung function test (LFT) was significantly reduced with a

diffusion capacity of 48%. The mass was resected with an uneventful operation and postoperative period. Pathology disclosed a cystic lymphangioma. Seven weeks after surgery, the patient was asymptomatic and with an improved LFT with diffusion capacity 67%, FEV1 95% and FVC 99% of expected, respectively.

Discussion

Embryonic lymphatic vessels fail to communicate to venous channels. Abnormal lymphangiogenesis thus ends as single or multiple cysts in one or multiple organs, growing as unilocular or multilocular masses that contain serous or milky fluid. Khobta *et al.* (2) classified lymphangiomas into: capillary, cavernous and cystic, whose full pathogenesis is not fully established yet (5). Cystic lymphangiomas were first described in 1843 by Wernher, while the first thoracic case of lymphangiomas was reported in 1973 (2). There have been previous

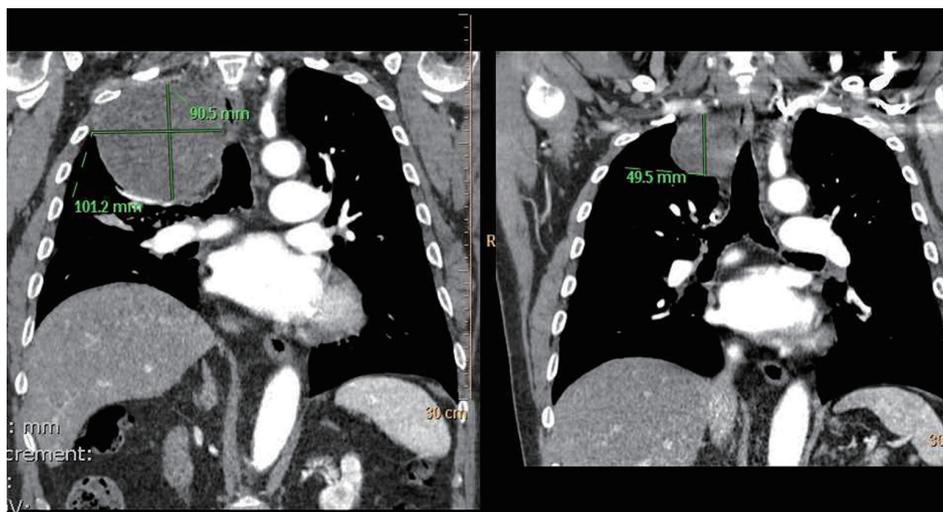


Figure 1 Computed tomography (CT) shows enlargement of cystic lymphangioma during a periode of 5 years (cystic value 10-20 on Hounsfield scale).



Figure 2 Cystic lymphangioma also called hygroma are soft tumours filled with milky fluid within a thin walled sac, vascularised by small tiny vessels from parietal pleurae.



Figure 3 Perioperative picture showing mediastinal cystic lymphangioma.

reported cases of asymptomatic lymphangioma involving the spleen and mediastinum (5). Our case is extraordinary since the patient had a combination of a single cystic lymphangioma in mediastinum causing symptoms and multiple asymptomatic cysts in lever and both kidneys. Differential diagnose include bronchogenic cysts, thymic cysts, thymomas, hydatid cysts, neurofibroma and malignant bronchogenic tumours. Mediastinal cysts form 12-18% of all primary mediastinal tumours (6). Diagnosis is almost always established after surgery despite extensive diagnostic procedures. Complete surgical resection with an intact cyst capsule is recommended in order to minimize risk of recurrences (2,7) (*Figures 2,3*). Other therapies (2,7)

such as aspiration, incision and drainage, irradiation, and sclerotherapy seem to be short time treatment with a high risk for complications like infection, haemorrhage, damage to muscle/nerve, rupture and recurrences.

Conclusions

We report a cystic lymphangioma diagnosed in an elderly patient's mediastinum, lever and both kidneys. The symptoms regressed fully and lung function normalised after surgical intervention. Although cystic lymphangiomas are rare, they must be considered in differential diagnoses of the mediastinal tumours.

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References

1. Kanzaki M, Kikkawa T, Obara T, et al. Successful excision of an isolated mediastinal cystic lymphangioma with bilateral thoracoscopic surgery. *Ann Thorac Cardiovasc Surg* 2011;17:570-2.
2. Khobta N, Tomasini P, Trousse D, et al. Solitary cystic mediastinal lymphangioma. *Eur Respir Rev* 2013;22:91-3.
3. Hunt I, Eaton D, Dalal P, et al. Minimally invasive excision of a mediastinal cystic lymphangioma. *Can J Surg* 2009;52:E201-2.
4. Limmer S, Krokowski M, Kujath P. Pulmonary lymphangioma. *Ann Thorac Surg* 2008;85:336-9.
5. Mohammadi A, Ghasemi-rad M, Abassi F. Asymptomatic lymphangioma involving the spleen and mediastinum in adults. *Med Ultrason* 2013;15:154-6.
6. Aydin Y, Ogul H, Turkyilmaz A, et al. Surgical treatment of mediastinal cysts: report on 29 cases. *Acta Chir Belg* 2012;112:281-6.
7. Wright CC, Cohen DM, Vegunta RK, et al. Intrathoracic cystic hygroma: a report of three cases. *J Pediatr Surg* 1996;31:1430-2.

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