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Case report - Vascular thoracic

Cystic lymphangioma: report of two atypical cases

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Abstract

Cystic lymphangioma is an uncommon congenital benign neoplasm, which frequently occurs to children and young adults and acquired form may be detected in middle-aged adults. Cystic lymphangioma usually appears in the neck, the axillary region, and the mediastinum. In the first case, cystic lymphangioma located in the posterior mediastinum extending over the vertebral column to the both hemi thoraces. In the second one, it was palpated on the posterior chest wall at the level of T6–8 vertebrae. In the view of literature, these cystic lymphangioma are accepted to be atypical because of their locations.

Keywords: Lymphangioma; Cystic; Atypical locations

1. Introduction

One of the developmental malformations of lymphatic tissue is cystic lymphangioma. Cystic lymphangioma usually occur in the neck, axillary region, and rarely mediastinum [1]. This study aims to present two cases with cystic lymphangioma because of their atypical locations.

2. Case reports and results

2.1. Case 1

A 48 year-old man had symptoms of palpitation and dyspnea with minimal effort during the recent 4 months. The echocardiogram demonstrated a giant cystic lesion adjacent to the heart mimicking pericardial cyst. Physical examination revealed normal findings. Laboratory studies were insignificant. Chest X-ray image (CXR) showed a silhouette along the right cardiac border. Computerized tomography (CT) of the chest showed a posterior mediastinal cystic lesion bulging towards both hemi thoraces (Figs. 1a,b). Cystic lesion was located in front of both right pulmonary artery and left atrium. It circumscribed the descending aorta, oesophagus and azygos vein on the anterior and thereafter extended to the left paravertebral sulcus. Upon right posterolateral thoracotomy, cystic lesion was mobilized and freed bluntly from descending aorta by finger dissection. The cyst was totally excised. The lesion was measured 13.5 × 5 cm in dimensions with 0.7 cm thickness wall, macroscopically. Pathohistological examination revealed cystic lymphangioma. The patient reported no complaints postoperatively.

2.2. Case 2

A 14 year-old male came with a problem of mass lesion on the posterior chest wall. The patient reported it had grown gradually in 4 months before hospitalized. It was painful on palpation. The hard and fixed mass lesion was located at the level of T6–8 thoracic vertebrae. Lateral CXR showed the lesion posterior to vertebral column. CT study defined a 3.0 × 5.0 × 6.0 cm mass with calcified wall. The mass with centrally cystic region was detected as hypo echoic solid image on magnetic resonance images (MRI) (Figs. 2a,b). Whole body bone scintigraphy scanning showed increased activity at the vertebral corpuses of T8–9. At the operation, it was dissected bluntly and freed from paraspinal muscles. There were some fibrous adhesions and a bony stalk to the spinal processes of the T6–8. The fibrous bands and bony stalk were removed sharply. The mass was excised totally. It was measured 6 × 5 × 2 cm

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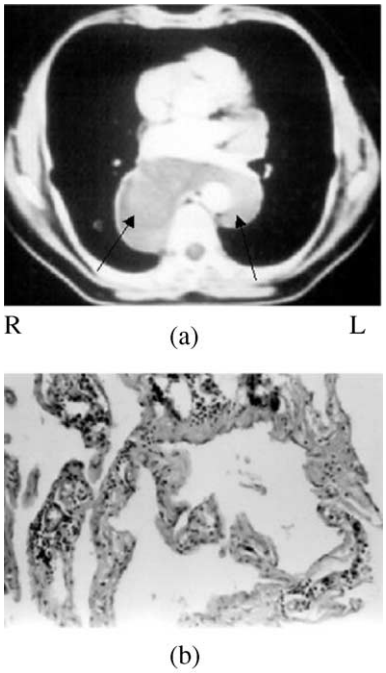

Fig. 1. (a) CT image, cystic lesion bulging towards the both hemi thoracic cavities (arrows). CT, computerized tomography; R, right; and L, left. (b) Histological micrograph of the lesion. Call attention to cavernous structures keeping lymphocyte groups under the endothelial layer with small amount of erythrocytes in the lumen.

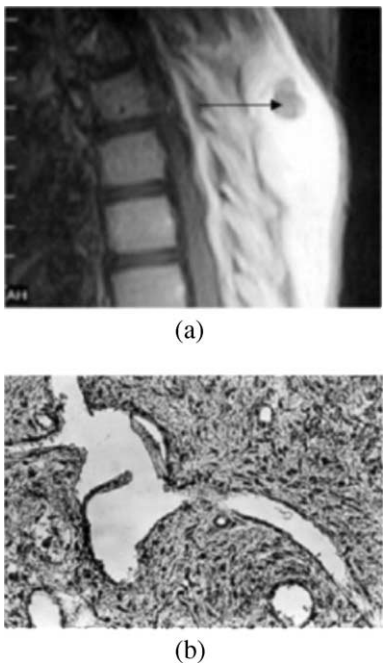

Fig. 2. (a) Thoracic MRI, centrally cystic mass lesion (level of T6–8), posterior chest wall (arrow). MRI, magnetic resonance imaging. (b) Histological micrograph of the lesion. In the fibro vascular stroma, lymph vessels covered with short single layer endothelial cells seemed to become cystic forms.

in dimensions with 2 mm thickness calcified wall. Two separated cystic cavities of 2 × 2 cm diameter were determined inside the lesion upon dissected it. It was also reported to be a ‘cystic lymphangioma’. The patient was uneventful postoperatively.

3. Discussion

Lymphangioma has been classified into three groups: (1) lymphangioma simplex; (2) cavernous lymphangioma; and (3) cystic lymphangioma or cystic hygroma [2,3]. ‘Cystic lymphangioma’ was determined in the two cases included in this study.

Cystic hygroma presents as a soft tissue mass in the posterior triangle of the neck and only rarely does it extend into the mediastinum [2,4]. Most of the cystic hygromas are not diagnosed during childhood until growing to be very large [2,5,6]. In the first case, the patient was adult and had a gradually increasing symptom during the past 4 months before admission. The cyst became very large and probably compressed anatomical structures adjacent to it. The second case expressed pain only on palpation 2 weeks before hospitalized.

Usually cystic lymphangioma appear in the neck or the axillary region and the rest in the mediastinum, retroperitoneum, pelvis, and chest wall [1,5,6]. In the first case, the lesion was located in the posterior mediastinum and in the other; it was on the posterior chest wall, which is a very rare location. First case of chest wall lymphangioma was reported as early as 1973 [7].

Intrathoracic lymphangioma located in paravertebral sulcus was reported. The one atypical lesion in the anterior mediastinum enveloped the heart and great vessels [2]. In the first case, the posterior mediastinal lesion was extending to both hemi thoraces. There is no reported case determined as having such bilateral bulging property in the literature.

CT and lymphangiography were helpful in determining the extent of the disease, the cystic, and the lymphatic nature of the mass. MRI can also be used to demonstrate the relationship of the mass and surrounding structures [2–4]. The CT features of the mediastinal lymphangioma are: (1) well-circumscribed lesion; (2) natural structures that have been enveloped or displaced; (3) absence of calcifications; and (4) varied attenuation values within the lesion [8]. The posterior mediastinal lesion shown by thoracic CT was a well-circumscribed lesion without invasive characteristics. The second case obtained thoracic CT and MRI as well. Both the calcified wall of the mass and the fibrous vertebral connection were well depicted.

Castleman’s lymphoma or thymic cyst [9], pericardial cyst, bronchogenic cyst, cystic teratoma and cystic thymoma [10] all should be kept in mind in the differential diagnosis of cystic lymphangioma.

Most surgeons agree that the cystic hygroma should be excised when the diagnosis is made because of the danger of severe complication. Meticulous surgical excision, in one or more stages is the most accepted treatment. Other types of treatment have been proposed as adjuvant such as radiotherapy and injection of sclerosant agents, but they are controversial [4]. Treated by surgery, the prognosis remains good. However, a few local recurrences, fistula malformation or infection have been reported [1,5]. In the two cases, the lesions were totally resected. But, it should be mentioned that it was challenging to dissect the lesion bluntly by the index finger in the opposite hemithorax close to the descending aorta in the first case. However, there were no morbidity or recurrence in the third postoperative month in both cases.

In conclusion, lymphangioma is a benign tumour. Provided that the tumour is completely resected, a good prognosis could be achieved. In the current study, we have not come across morbidity as reported in the literature; perhaps this is because of the limited number of lymphangioma cases encountered in our clinic. In both cases, we managed to perform complete resection without any complications. The findings of this study are worth reporting due to atypical locations and rareness of the cystic lymphangioma.

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Appendix A. ICVTS on-line discussion

Author: ***Dr. Sameh Sersar***, *Assistant Lecturer, Mansoura University, Department of Cardiothoracic Surgery, Mansoura, 123 Egypt*

Date: 22-Oct-2003

Message: How were you able to remove the left sided lesion enveloping the descending aorta? What about the infiltrative nature well known to cystic lymphangioma? Was it evident intraoperatively?

Response

Author: ***Dr. Erkan Yildirim***, *Ankara Numune Education and Research Hospital, Thoracic Division, Talatpasa Bulv., Samanpazari, Ankara, Turkey*

Date: 03-Nov-2003

Message: Although it was reported in the literature [1,2] that the cystic lymphangioma may have an infiltrative nature, in our case the lesion was removed bluntly by finger dissection completely without rupturing the cyst. The descending aorta was detected to be uninfiltrated by the lymphangioma.

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