Case report

Cystic lymphangioma of the heart mimicking a mediastinal tumor

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Abstract

Primary cardiac tumors are rare and myxomas are the most frequent. Cystic lymphangiomas are most often of cervicomediatinal locations and are exceedingly rare in the mediastinum. Cystic lymphangioma involving the heart has never been reported. We report a case of cystic lymphangioma involving the posterior wall of left atrium mimicking a mediastinal tumor. Complete resection was performed via a median sternotomy under cardiopulmonary bypass and cardiac arrest after transection of the aorta and pulmonary trunk. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

Primary cardiac tumors are rare with an incidence ranging from 0.0017% to 0.28% in autopsy series. More than 90% are benign tumors with myxomas representing 70–80% of all cardiac neoplasms [1]. Hemolymphangiomas of the heart have been reported [1] but a cystic lymphangioma (CL) has never been reported. CLs are abnormal collection of lymphatics. They may be located everywhere in the body except in the brain, the anterior chamber of the eye, and the bone marrow where no lymphatic system can be found [2]. Localizations in the red pulp of the spleen have recently been reported [3]. CLs are most often located in the neck and less than 2% are located in thoracic area with predilection in the anterior part of the mediastinum. We report here a case of heart localization mimicking a mediastinal tumor and review the literature on its pathogenesis, clinical features and treatment.

2. Case report

A 67-year-old woman was referred to our department for treatment of a mediastinal mass. She suffered from progressive worsening of a dyspnea that had appeared several months before. Past medical history consisted in allergic asthma in childhood, and no tobacco abuse was found. She was first admitted to a cardiology department after a pulmonary embolism had been suspected then ruled out. She weighed 52 kg and was 1.64 m in height, and physical examination was normal. Chest X-ray revealed an abnormal mediastinal shadow. It was a retrocardiac mass which developed against the tracheobronchial bifurcation, shifting the esophagus and in direct contact with the pulmonary infundibulum. On magnetic resonance imaging, a heterogeneous mass was observed under gadolinium-enhanced T1-weighted images with hyperintense signal on T2-weighted images (Fig. 1). The electrocardiogram was normal. Esophagography and esophagoscopy showed an esophageal shift with normal mucosa. Bronchoscopy revealed compression without stenosis. A mediastinal tumor shifting the heart was suspected and resection was decided. The tumor was exposed by a right thoracotomy through the fifth intercostal space after taping the esophagus. Cardiopulmonary bypass was judged necessary to perform the tumor resection but could not be performed safely through the thoracic approach. Three days later, the operation was achieved under cardiopulmonary bypass and cardiac arrest via a median sternotomy. After cardiopulmonary bypass was instituted between the ascending aorta and both venae cava and cardioplectic arrest obtained, the aorta and pulmonary trunk were transected. This allowed exposition of the left atrium in its upper part. Extent of tumor spread
concerned all of the posterior wall of the atrium. After opening of roof of the left auricle, tumor resection was carried out downwards along the roots of the pulmonary veins in safe margin tissue. The tumor was totally excised. A horse pericardial patch was inserted to replace the posterior wall of the left atrium, fixed by a running suture around the atrium’s defect (Fig. 2). Intraoperative pathologic evaluation did not show any malignant aspect and confirmed that surgical margins were negative. Final pathology results revealed a CL (Fig. 2). The postoperative course was uneventful. The patient is asymptomatic and without recurrence 8 months postoperatively.

3. Discussion

A review of reported cases of CL [3–5] shows locations in the neck in 75% of cases, in axilla and shoulder joint in 15%, and in other sites (cervicmediastinal location, retroperitoneal area, in the spleen or the colon, esophagus and chest wall) in 10%. In the thorax they represent 0.7–4.5% of all mediastinal tumors in the adult population [4]. They can be located in all mediastinal compartments [3,4]. Pulmonary localization [6] has been reported but to our knowledge, this is the first reported case of heart localization of CL. One case of hemolymphangiomatosis has been reported in Miralles et al.’s series of cardiac tumors [1]; however, it was localized in the inferior pulmonary artery and resected by thoracotomy without cardiopulmonary bypass.

Histologically, the absence of red blood cells in the cyst contents eliminates hemangiomia or lymphangiohemangiomia [7]. The microscopical aspect was typical of CL in our case (Fig. 2).

Mediastinal CLs are most often asymptomatic masses unexpectedly discovered on chest X-ray [4,5] whereas cardiac tumors are most commonly revealed by congestive heart failure, syncopal or embolic pathology [1]. When mediastinal CLs are symptomatic, symptoms arise from compression: vocal cord paralysis, arm paresthesia, venous compression, and stridor [4].

Radiological aspects of CL have been described [4,8,9]. It is typically a multiloculated lesion with cystic cavities divided by septa of variable thickness [8]. An evocative imaging feature is the spread of the tumor around adjacent structures or displacement of major mediastinal vessels [4]. Transesophageal echocardiography, which was not performed in our case, could have shown the atrial origin of the tumor.

The pathogenesis of these tumors is not well understood. They may be developmental, hamartomatous, or neoplastic [4]. Hilliard et al. [2] suggested a common pathogenesis of developmental lymphatic disorders leading to lymphangioma (localized lesions), lymphangiomatosis (multifocal lesions), lymphangiectasis (dilated lymphatics), mixed...
vascular lymphatic angiomas, and combinations of lymphatic and other tissues (including lymphangiomyomatosis). This theory does not account for all clinical forms. Unlike cases involving infants or young patients where congenital pathogenesis seems clear, in adults and elderly patients neoplastic or obstructive causes have been discussed [3,5]. Acquired causes seem more likely in our case. Surgical resection is always advised to avoid the risk of compression. CLs have a potential risk of recurrence [3,5], especially if there has been incomplete resection. Therefore, extensive resection is recommended.

Finally, the approach used in our case was inspired by the heart transplantation technique [10]. We preferred this approach to the trans-septal one. This approach offered a larger view on the posterior wall of the left atrium.

References


Fig. 2. (A) Intraoperative view showing the resected portion of left atrium: see aorta (Ao) and pulmonary trunk (Pul) transected and the right pleural cavity (Plc) at the bottom; Med, posterior mediastinum. The posterior wall of left atrium is hanging by a thread (arrow). (B) Photomicrograph: spongy tumor consisting of interconnecting channels lined by flat endothelial cells (arrow) beneath which are bundles of smooth muscle (Sm) and lymphoid nodules (Ln) (H&E, ×100).