Case Report

Left Pleuropericardial Cyst

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ABSTRACT: Introduction: Cysts of the mediastinum, which are benign masses, are usually detected by chance, and constitute a small but important diagnose group, representing 7 to 18% of all primary mediastinal tumors. Pleuropericardial cysts, located most frequently in the left anterior and inferior mediastinum, are identified in the fourth or fifth decade of life affecting females more than males with a sex ratio of 8:4. Material and method: We present the case of a 52 year old woman diagnosed with pericardial cyst located in the left cardiophrenic angle. CPP was rather big and was initially diagnosed as encysted pleurisy. Ultrasound guided exploratory thoracentesis was performed evacuating 300 ml "water spring" like fluid very scarce in cells. We further investigated our patient for differential diagnosis but the patient refused surgery and she is now managed conservatively with a close follow-up. Conclusions: A pericardial cyst should always be suspected when a cystic lesion is detected in the mediastinum. Pleuropericardial cysts are usually suspected after an abnormal chest X-ray is obtained. If the patient is asymptomatic and the information provided by CT indicates a benign tumoral process suggestive for a pleuropericardial cyst, conservative management with careful follow-up is justified.

KEYWORDS: *Pleuropericardial cyst, mediastinal tumor, cardiophrenic angle.*

Introduction

Pleuropericardial cysts usually are congenital anomalies. After the third week of gestation, the mesoderm separates itself to form pleural, pericardial and peritoneal spaces. Incomplete partitioning can result in a pleuropericardial cyst.

They are found in the literature under various names: le kyste pleuropericardique (Jeaubert de Beaujeu, 1945; Roche, 1954), pleural cyst, pericardial cyst, pericardial coelomic cyst (Lambert, 1940), springwater cyst (Greenfield, Steinberg, and Touroff, 1943), mesothelial cyst (Churchill and Mallory, 1937), and thin-walled cyst [1].

The classification of mediastinal cysts is based on their localization and etiology, encompassing bronchogenic, esophageal duplication cysts of foregut origin, mesothelial derived pericardial/pleural cysts, thymic cysts, and other miscellaneous cysts. Lambert (1940) has described the origin of these cysts. The pleura and the pericardium develop from the same coelomic cavity: that is why Herbig, Ganz, and Vieten (1952) believe that the term 'pleuro-pericardial cyst' gives the most accurate description of the tumor [1].

Mesothelial cysts, including pericardial and pleural cysts, are clinically silent. They usually manifest themselves as an asymptomatic disease in the case of well-limited lesions, in contrast to bronchogenic cysts which often become symptomatic or complicated [2]. Of the pericardial cysts, 70-80% is located in the right cardiophrenic angle of which the incidence is estimated to be 1/100,000 [2,4].

Pleuropericardial cysts are identified in the fourth or fifth decade of life and represent 7-18% of all the mediastimum tumors. CPP affects females more than males with a sex ratio of 8:4 and are typically located at the right cardiophrenic angle (51-70%), or left cardiophrenic angle (28-38%).

The cyst is unilocular, round, or ellipsoid. It may or may not be pedunculated and varies in size. The weight ranges between 100g and 300g. Most of them have a diameter of 3 to 6cm, but Leigh and Weens indicated they may be as large as a grapefruit. Lam (1947) reported one case where the cyst contained 1 liter of fluid and measured 25 x 37-5cm.

The fluid is a transudate. The 'water' is clear and has been compared to rock-water, whence the name 'spring water cyst'. In some cases, however, the fluid is yellow (Roche, 1954) [1]. Microscopical exam showed that the wall of the cyst contained a single layers of mesothelial cells and a loose stroma of fibrous tissue, which is suggestive for pleuropericardial cyst [4].

Case

We report the case of a 52 year old woman with history of high blood pressure that was admitted in January 2013 for chronic cough, night sweats, progressive shortness of breath on exertion and weight loss worsening in the last 3 weeks.

Physical exam revealed rather good clinical condition but pale and sweated skin, diminished breath sounds on the left lower lung area and BP=150/80mmHg.

Sputum smears for acid fast bacilli were negative.

Chest X-ray evidenced an oval shaped opacity of 5/8cm, high density, situated in the left paracardiac area with anterior projection.

Laboratory exams were normal. IgG antibodies for Echinoccocus were negative.

CT scan of the thorax described the presence of encysted pleural effusion in the left cardiophrenic angle with a fluid layer of 5-8cm.

Ultrasound exam of the thorax revealed a pleuropericardial cyst tangent to the left chest wall. Ultrasound guided exploratory thoracentesis was performed evacuating 300ml "water spring" like fluid; lab exams revealed a transudate very scarce in cells.

Control chest X ray evidenced downsizing of the initial cyst.

Final diagnosis was: Left pleuropericardial cyst, hypertension grade II.

We recommended surgery because of the initial size of the cyst and because of possible complications but our patient refused.

Even if she did not have any symptoms she was reexamined after 6 months. Thorax computed tomography didn't evidence the growth of the remaining cyst (Fig.1).

We consider her case is particular because the initial size and location of the CPP made possible the ultrasound guided exploratory thoracentesis. Prognosis was good, requiring further clinical and radiology monitoring.

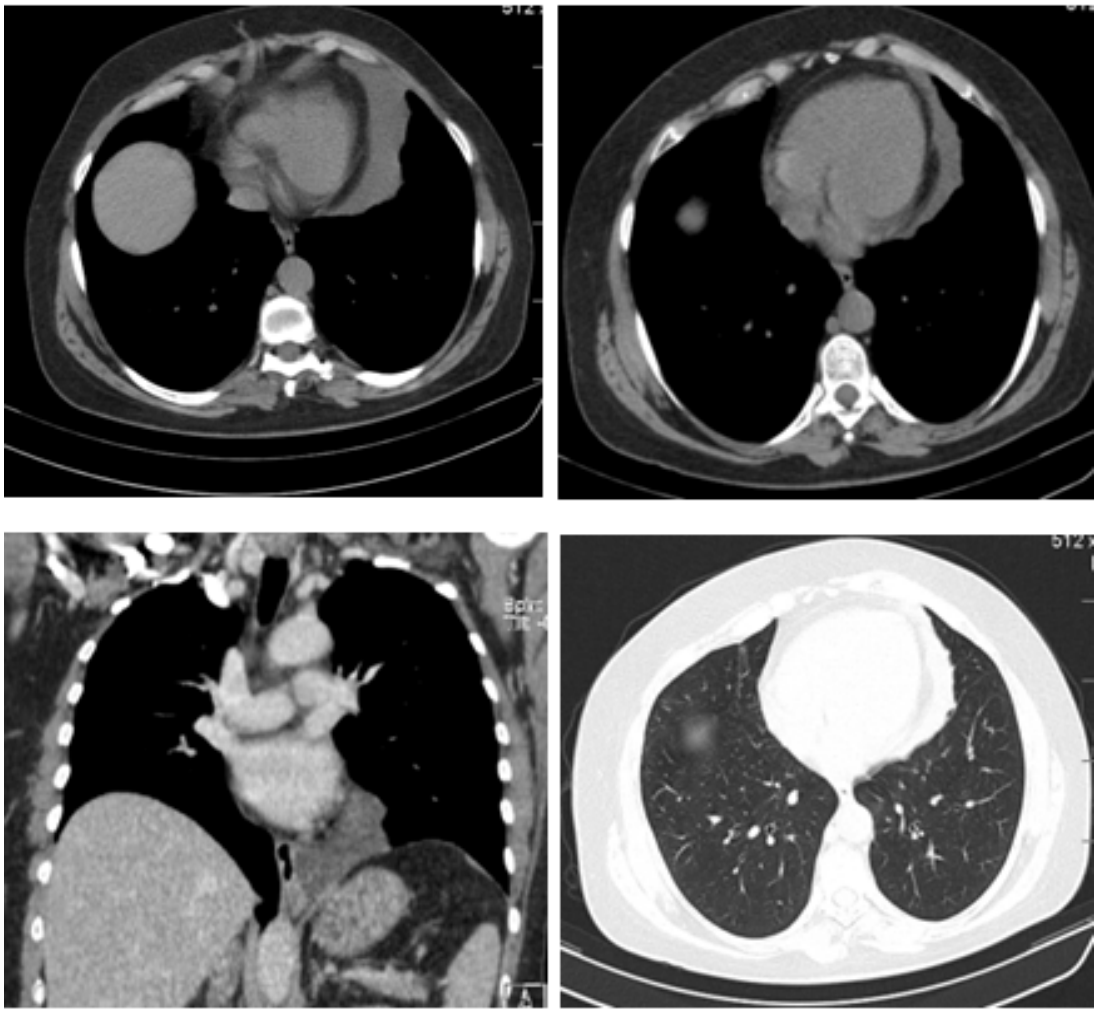

Fig. 1. Thorax CT scan after 6 months from the thoracentesis

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Discussions

Pericardial cysts are mostly congenital but can also be acquired after cardiothoracic surgery.

These cysts frequently occur in the right cardiophrenic angle and their diagnosis is usually suspected after an abnormal chest X-ray is obtained.

The size of these cysts varies from 2 to 3cm to as large as 28cm.

Although most pericardial cysts are asymptomatic, patients may present with chest pain or dyspnea. In addition, life-threatening complications such as pericardial tamponade have been reported [6,7,8].

Most cysts are asymptomatic and can be left untreated, as these cysts do not tend to malignant degeneration [7,8,9].

However, sometimes, especially in children, these cysts can give complaints of chest pain or dyspnea [1].

Some danger lies in the pressure disorders which occur when the tumor grows in size: pressure on the heart which is affected directly or through the vagus, and pressure on the bronchi with resulting atelectasis of the pulmonary lobes.

Following aspiration there is a danger of infection, so that this should be avoided as a general rule.

As to the benign nature of these cysts, all authors reviewed by us are in complete agreement: no degeneration recurrence of simple cysts has ever been observed [1,7].

In the past, many presumed pericardial cysts were not surgically removed because of a characteristic appearance and benign behavior.

Pericardial cysts are usually asymptomatic, although in few cases multiple complications have been reported which include cyst rupture, cardiac tamponade, mitral valve prolapse, hoarseness atrial fibrillation right ventricular outflow tract obstruction, spontaneous internal haemorrhage, pulmonary stenosis related to extrinsec compression, and even sudden death.

There are no reports of malignant transformation.

Pleuropericardial cysts could be treated by a thin needle puncture under scanography.

Since 1993, videothoracoscopic surgical removal of pericardial cysts is an excellent surgical intervention without serious morbidity and mortality [9].

Indications for surgery, in the presented case report, included the presence of symptoms and uncertain radiographic diagnosis [9,10].

About 75% of the cysts are asymptomatic, and they are incidentally found in chest radiographs and echocardiograms [9,10].

There are reports in the literature of giant cysts, atypical location cysts (anterior and posterior mediastinum), in different shapes, associated to other diseases-Fanconi anemia-and hemorrhagic cysts. Bava et al. described a case of torsion of the cyst intrapericardial pedicle with consequent ischemia and tamponade [5].

Apart from the possibility of congenital cysts in the thorax, it will be necessary to eliminate hernias of the fissure pericardiacoperitonealis (Morgagni) and the trigonum sternocostale diaphragmatic (Larrey).

A more difficult differential diagnosis is between pleuro-pericardial cyst and cystic pericardial lymphangioma [9,10].

Aspiration will reveal lymphocytes in a lymphangioma, and histologically the cyst wall is thicker, contains blood vessels, lymphocytes, and very thin fibers. In addition, the cyst is multilocular.

Malignant tumors are situated higher up in the mediastinum and are most common in the posterior mediastinum.

Thyroid or thymus cysts are found in the antero-superior mediastinum [1].

In these cases treatment is indicated. Several forms of treatment have been described in the literature: complete resection by means of thoracotomy, videoassisted thoracoscopic surgery and percutaneous aspiration under ultrasound guidance [1].

Aspiration of the cyst is usually safe but carries the risk of anaphylaxis and dissemination in the rare case of this being a hydatid cyst.

Surgery has been advocated, only in symptomatic patients.

Several modalities of treatment have been described in the literature; complete resection by means of thoracotomy, median sternotomy, video assisted thoracoscopic surgery and percutaneous aspiration under ultrasound guidance.

The prognosis after complete excision is excellent and morbidity and mortality rates are low [10,11].

Asymptomatic cases are managed conservatively with a close follow-up [6,11].

Conclusions

A pericardial cyst should always be suspected when a cystic lesion is detected in the mediastinum.

Diagnosis is not always easy, though most important with regard to subsequent therapy.

The most useful imaging studies for diagnosis are echocardiography, CT, and MRI. Asymptomatic cases are managed conservatively with a close follow-up.

If treatment of a pleuropericardial cyst is mandatory thoracoscopic intervention is a safe and effective method for both diagnosis and treatment with good prognosis after complete excision.

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