A large thymic mass - a possible cause of unexpected death

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Abstract: Thymic cysts are particularly rare, and because most are asymptomatic, they are usually found incidentally as unsuspected masses on chest radiographs. We report two cases, one congenital and the other neoplasic thymic cysts, both presented initially as idiopathic thymic cysts, and later in association with chronic respiratory or cardiac symptoms developed in relation with widening of the mediastinal shadow confirmed on chest radiographs. Morphological evidence of hemorrhage into thymic cyst was found in a patient and ischemic necrosis to another. These cases emphasize that enlargement of the non-neoplasic and neoplasic thymic cysts may cause acute or chronic symptoms of airway or cardiac compression with widening of the mediastinum. Thymic cysts should be added to the list of thymic abnormalities that may occur in patients with compressive mediastinal masses, which can be sometimes lifethreatening for patient.

Key Words: thymic cyst, cystic thymoma, mesothelial cyst

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The ectopic thymic tissue is a rare finding. Misplaced thymus could be diagnosed as a cervical tumor [1] or intrapleural or pericardial nodular masses [2] but compressive masses has been reported only in individual autopsy cases of thymomas [3].

Case reports

Case 1.

A 23-year-old man was admitted for progressive dyspnea and dysphagia caused by traheal and pharangeal compression. He had no history of pneumonia or pleurisy. A preoperative chest radiograph showed a large left antero-superior mediastinal mass, which was described as an asymptomatic “pericardial cyst” on a chest radiograph taken few months ago.

At actual patient presentation chest radiograph showed a mass effect on the airway and pharynx, as well. Diagnostic possibilities included mediastinal cysts. Because of the great concern, a right parasternal mediastinotomy was performed under general anesthesia.

At surgery, a large mass filled the antero-superior mediastinum, containing a sero-hemorrhagic fluid. The surgery biopsy specimen evidintiated no signs of malignant tumor. Gross examination of the resected specimen revealed a 7 x 4 x 6 cm, slightly lobulated and sharply circumscribed, pinkish mass. The cut surface demonstrated two cystic spaces, the largest measuring 4 cm in the greatest dimension (Figure 1).

Microscopic examination confirmed thymic tissue in the walls of the hemorrhagic cyst. The walled lymphoid tissue consisted of foci of uniformly small, round lymphocytes with normal features. Higher powered examination of this tissue revealed few epithelial structures reminiscent of Hassall's corpuscles. The lining of the cysts was composed focally of columnar epithelium with fibrous supporting tissue, but much of the cyst lining was replaced by numerous cholesterol clefts.

There was no evidence of malignant epithelial change. Also, there was a slight evidence of recent intracapsular hemorrhage and a homogenous, intense eosinophilic intraluminal
content with numerous cholesterol clefts (Figure 1). The findings were consistent with bilateral thymic cyst with both recent and old hemorrhage.

Case 2.

Asymptomatic, 41 years old, male patient, was diagnosed two years ago on a routine chest X-ray with a left pleuro-pericardial mass. Thorax CT scan performed subsequently showed a normal thymus and a well defined cystic tumor of 58/39 mm, located in the antero-inferior mediastinum, being considered at that time, a pleuro-pericardial cyst.

In the mean time, the patient presented signs suggestive of cardiac ischemia, which were highlighted by electrocardiography, and he received specific drugs. Recently, the patient was diagnosed with myasthenia gravis, clinical stage Osserman IIB.

The medical treatment with anticholinesterase drugs and steroids, improved clinical picture. The patient underwent an extended thymectomy through a median longitudinal sternotomy. Hospital stay was of 32 days and on discharge he was in pharmacological complete remission of myasthenia gravis. Gross examination of the surgery specimen revealed an encapsulated thymic tumor presenting on cross section a large cystic cavity. The cyst wall was lined by nodular masses of different size. The adjacent thymus tissue was normal (Figure 2).

Histological exam showed a cellular tumor with a prominent lobular pattern under low-power microscopy.

The lesions showed a dense tumoral tissue and paravascular spaces with a lobular configuration, sharply separated by thick collagenous septa arising from the capsule. With higher magnification, polygonal epithelioid cells admixed with lymphoid tissue were visible. There was no evidence of capsular penetration by tumour.

At the level of the capsular pedicle we saw two severe stenotic vessels (Figure 2). According with histological Bernatz scheme, we identified a mixed tumor, with epithelial-predominant component, including focal spindle-cells. A diagnosis of non-invasive benign mixt thymoma, Masaoka stage I was made.

Discussion

The „ectopic site” of cystic thymus has been a debated subject. It is considered an abnormal development of the thymus gland during its phase of embryonic descent from the skull basis to submandibular region, associated with the misplacement of gland remnants [4]. In the two our cases presented here, the site of the thymus was in the upper and lower part of the anterior mediastinum, respectively.

We can speculate that the gland descended ‘too rapid or too slow’ invaginating near tracheae or sliding between pleuro-pericardial sacks. It is hard to predict the significance in the future. In the first case, the histological diagnosis of thymic cyst wasn’t difficult. It is already

Figure 1. a - cystic mass; b - fibrous wall with thymic tissue; c - lining columnar epithelium; d - intracapsular haemorrhagic focus; e - cholesterol cleft posthemorrhagic lysis; f - pericapsular thymic tissue.
known that, in order to make the thymic cyst diagnosis, the thymic tissue must be identified within the cyst wall [5].

This ectopic thymus cyst was become in time a compressive cystic mass by sero-hemorrhagic intracystic accumulation. Regarding the second case, it is known that thymoma develops in a normal location of the thymus, but it can arise in various ectopic locations: the pleural cavity mimicking mesothelioma, the pericardium giving rise to “cardiomegaly” on chest x-ray films, or cervical locations [6].

“Thymus cystic change” is a rare diagnostic feature. Thymic cysts are believed to represent different entities, congenital, inflammatory and neoplastic cysts [4].

Congenital thymic cysts develop due to induction of cystic transformation of the ductal epithelium of the thymopharyngeal duct system. The acquired thymic cyst is related by a previous inflammatory process [5] of vicinity, which wasn’t demonstrated in our first case. But, the secondary inflammatory process developed here was reactive to intracavitary accumulation of sero-hemorrhagic fluid. Focal intracapsular hemorrhage and intraluminal cholesterol clefts are indicative of recent and chronic thymic cyst hemorrhage.

Cystic changes in thymoma may result by two pathogenetic mechanisms: (a) confluence and dilatation of the perivascular spaces with creation of large cystic cavities devoid of epithelial lining or inflammation, and (b) cystic dilatation of Hassall’s corpuscles [6, 7]. Our second case illustrates a secondary necrotic event developed in a thymoma associated with severe ischemic condition, demonstrated by the presence of the two severe stenotic vessels at the level of the tumoral pedicle enclosed in the cyst wall.

Sometimes, necrosis can obscure the main underlying tumoral lesion, making impossible to determine whether the cystic changes supervened in a preexisting thymoma or whether the thymoma developed secondary in a thymic cyst [8, 9]. In our case, thymoma was found as mural multiple nodules attached to the wall of the thymic cyst, pointing an initial thymoma with secondary cystic transformation.

Thymic cysts are rare causes of mediastinal masses at any age and “the large tumors” are more rare. Generally, mediastinal cysts are asymptomatic and are detected as unexpected anterior mediastinal masses on chest radiographs, although they may occasionally cause respiratory distress or cardiac signs, by compression of adjacent structures. Usually, thymomas present by their mass effect causing cough, dyspnea, chest pain, or via paraneoplastic mechanisms, for example, myasthenia gravis.

As Ağaçkiran Y said, a mediastinal widening in a patient with myasthenia gravis should suggest the diagnosis of hemorrhage into a thymic cyst, in addition to the possibility of an ischemic necrosis [10].
Conclusions
The presented thymic cysts are unusual by ectopic location in the upper or lower part of the anterior mediastinum. These cases, initially small, asymptomatic cystic lesions, were later presented as large symptomatic masses, on radiograms, causing compression of the upper airway and right atrium.

These thymic cysts are rarely definitively diagnosed pre-operatively. The pathologist has to take in account this entity at assessment of surgical resection in patients with previously undiagnosed anterior mediastinal masses.

References