Giant left-sided pleuropericardial cyst, mimicking a heart disease

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Abstract

Mediastinal cysts (MC) mainly have an embryonic origin, are benign and frequently discovered thanks to tomodensitometry, sometimes by magnetic resonance imaging. Rarely symptomatic, excepted in cases of very large cysts, they are mainly pleuropericardic cysts (PPC) that represent 30% of MC.
Surgery is commonly performed by videothoracoscopy or by video-assisted mini-thoracotomy, mainly for PPC.
We report the case of a 62-year-old woman, smoker (30 packs years), who is hospitalized in Constanta Pneumology Hospital in June 2011 for slight shortness of breath, sweating, pain in the left hemi thorax, minor hemoptysis, recurrent. In her medical history, there are to be noticed a blood transfusion after hysterectomy for uterine fibroma (1995), arterial hypertension (2006). After admission, X-ray exam of the chest shows cardiomegaly and a few lung nodular lesions in the right upper lobe. An initial differential diagnosis includes congestive heart failure, dilated cardiomyopathy, valvular heart disease, left pleurisy, pericarditis, paracardiac tumor mass, tuberculosis +/- HIV. Following laboratory tests imaging (chest CT and ultrasound performed in June 26th 2011 and 27th) a possible pleuropericardic cyst was suspected. Exploratory thoracentesis was not performed and, a month later, in the Institute of Pulmonology "Marius Nasta", Bucharest, a left open thoracotomy revealed a cystic formation about 10 cm in diameter. Histopathologic exam confirmed the diagnosis of cyst pleuropericardic. The prognosis after surgery was favorable. As a feature of the case are worth mentioning: the large size of pericardial cyst at the upper limit of the data reported in the literature, which mimics cardiomegaly, the hemoptoic onset in a hypertensive patient, heavy smoker; the late suspicion of pleuropericardic cyst through pleural echographic exam; the atypical localization; the facilitated certain diagnosis by surgery and hystological exam; the favorable postoperative prognosis; and all morbidities cofound (Pulmonary Tuberculosis, bronchiectasis, COPD).
Keywords: *giant pleuropericardial cyst, open thoracotomy, chest ultrasound, hrmoptysis*

Rezumat

Chist pleuropericardic gigant stâng, mimând o boală cardiacă
Chisturile mediastinale (CM) au, în principal, origine embrionică, sunt benigne și frecvent descoperite prin tomografie computerizată sau rezonanță magnetică. Arareori simptomatice, cu excepția chisturilor de dimensiuni mari, sunt preponderent chisturi pleuropericardice (30% din CM). Raportăm cazul unei femei de 62 de ani, fumătoare (30 pachete/an), internată de urgență în iunie 2011, în Spitalul Clinic de Pneumoftiziologie Constanța, pentru dispnee ușoară, transpirații, durere în baza hemitoracelui stâng, hemoptizii minore, recurente. În istoricul medical remarcăm transfuzie de sânge după histerectomie pentru fibrom uterin (1995) și hipertensiune arterială (2006). Radiografia pulmonară, la internare, evidențiază cardiomegalie și leziuni pulmonare nodulare apical lob superior drept. Un prim diagnostic diferențial include următoarele entități clinice: insuficiența cardiacă congestivă, cardiomiopatia dilatativă, valvulopatia, pleurezia stângă, pericardita, o posibilă masă tumorală paracardiacă, tuberculoză +/- HIV. Ca urmare a examenelor paraclinice imagistice (tomografia computerizată a toracelui efectuată în 27.06.2011 și echografia pleurală), se suspectează un posibil chist pleuropericardic, e temporizată toracenteza exploratorie, iar o lună mai târziu, în iulie 2011, în Institutul de Pneumologie „Marius Nasta" București, se intervine chirurgical și, prin toracotomie stângă, este descoperită o formațiune chistică de 10 cm în diametru. Examenul histopatologic confirmă diagnosticul de chist pleuropericardic. Prognosticul postoperator al pacientei a fost bun, dar s-a impus monitorizare. Ca particularități de caz, merită menționate: dimensiunea mare a chistului pericardic, la limita superioară a datelor raportate în literatură, care imită cardiomegalia; debutul hemoptoic la o pacientă hipertensivă, mare fumătoare; suspicionarea chistului pleuropericardic prin intermediul ecografiei pleurale datorită localizării atipice; confirmarea tardivă postoperatorie; prognosticul favorabil după înlăturarea unui chist pleuropericardic gigant, precum și comorbiditățile bronhopulmonare descoperite consecutiv (tuberculoza pulmonară sechelară, bronșiectaziile, BPOC).
Cuvinte-cheie: *chist pleuropericardic gigant, toracotomie, echografie pleurală, hemoptizie*

Introduction

Cysts of the mediastinum, which are benign masses, are usually detected by chance, and constitute a small but important diagnose group, representing 12 to 18% of all primary mediastinal tumors[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 [1]. Mesothelial cysts, including pericardial and pleural cysts, are clinically silent, being estimated to occur in approximately 1 in 100,000 persons[1] and affect females more than males with a sex ratio of 8:4[2].

Case report

We describe a case of a left sided pleuropericardial cyst diagnosed in a 62- year white woman, heavy current smoker (30 pack years), whose medical records included 2U of whole blood transfusion after radical hysterectomy for uterine fibrom in 1995, hypertension since 2006, gastroesophageal reflux syndrom since 2010, who was admitted in Constanta Clinical Pulmonology Hospital with progressive shortness of breath on exertion, night sweats, left chest pain and minor reccurent hemoptysis.

Physical examination revealed obesity (body mass index, BMI=32,4kg/m^2), an increased rate of pulse (109 beats/min) and high blood pressure (190/120 mm Hg).

A routine chest roentgenogram revealed in first day of hospitalization, June 23rd 2011, an enlarged cardiac silhouette and a few homogenous well defined small nodular opacities in the apical zone of right upper lobe (Figure 1).

An initial differential diagnosis included congestive heart failure dilated, cardiomiopathy, valvular heart disease, peri-

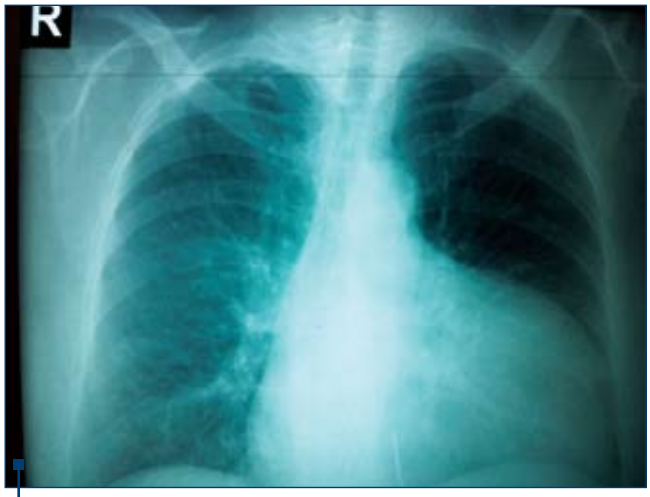

Figure 1. Chest X-ray performed in 23rd June 2011 in an adult female, 62 years old, revealed an important enlargement of cardiac opacity, high suspicion of cardiomiopathy; lung round nodular opacities, 0.6-1.2 cm, in the apical area of the right upper lobe, well defined, homogenous, suspicion of Pulmonary Secondary Nodular Tuberculosis

carditis, left pleural effusion, cardiac tumor, cardiac hydatid cyst disease and, even, HIV related tuberculosis.

In addition to plain chest radiographs, the evaluation included computed tomography (CT) which was performed in 26th of June 2011 and showed sequelae of Pulmonary Secondary Nodulary Tuberculosis localized in apical segment of right upper lobe, bronchiectasis in lower right lobe and a smooth ovoid thin-walled 10 cm x 11 cm, sharply defined, homogeneous mass (without tear drop configuration) in the left cardiophrenic angle (Figure 2). Because of its fluid content with a density of 4 Hounsfield units (HU), a pleural collection of fluid was suspected. The chest ultrasound revealed, one day later, a pericardial effusion 10 cm x 11 cm consistent with a potential pericardial cyst (Figure 3).

Surgery was carried out a month later, in July 2011, in The Pulmonology Institute "Marius Nasta", Bucharest. A left open thoracotomy was attempted and revealed a round cystic formation of about 10 cm in diameter in the lower left thorax, that was filled with fluid. Histopathological exam confirmed the diagnosis of a cyst pleuropericardic. The patient was discharged a week later. Prognosis was good, requiring further clinical and radiology monitoring. In September

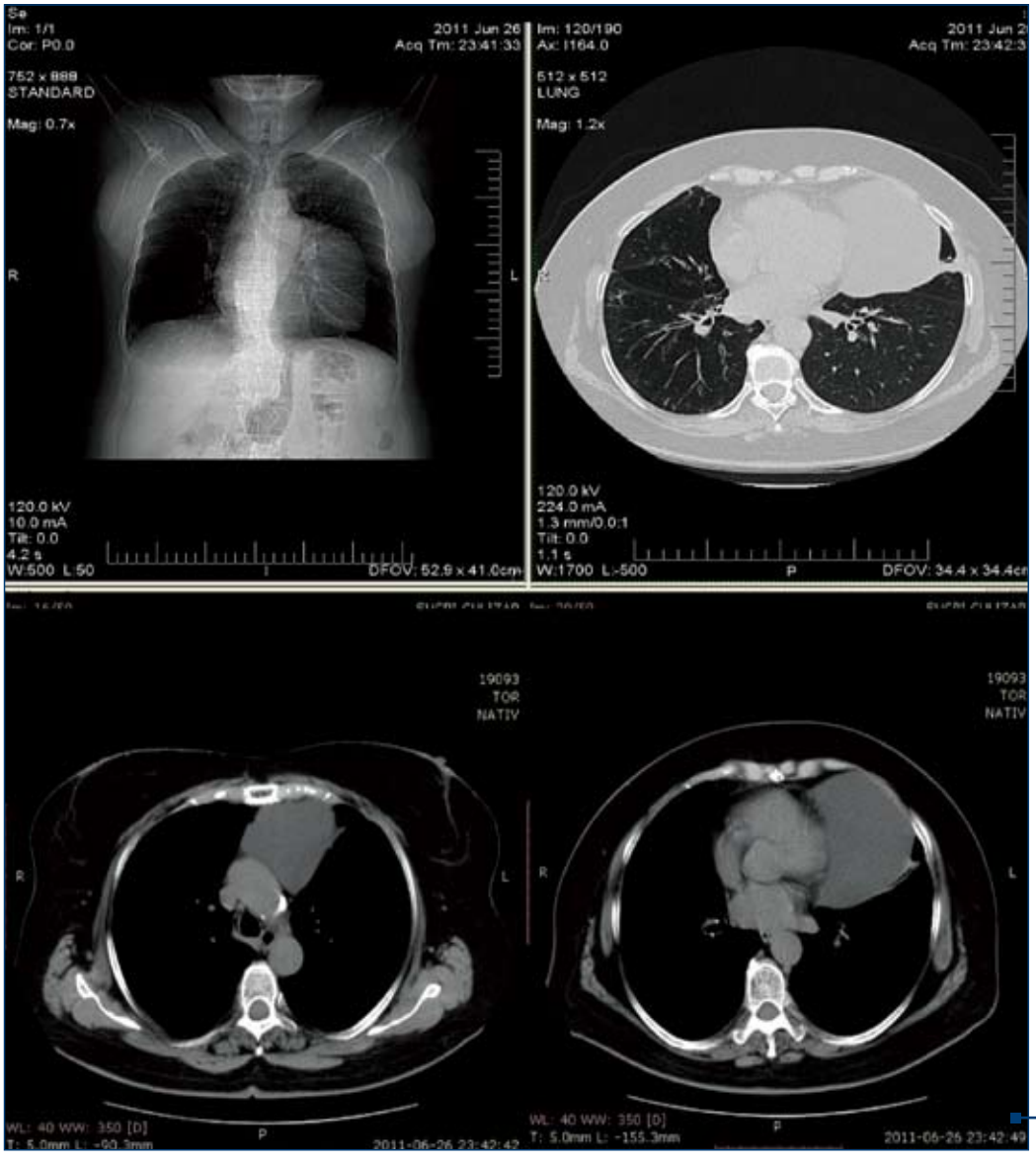

Figure 2. CT scan of the chest performed in 26th of July, 2011 reveals an effusion localized in the left pleural space, left lower lobe bronchiectasis and fibronodular lesions in the apical segment of the right upper lobe

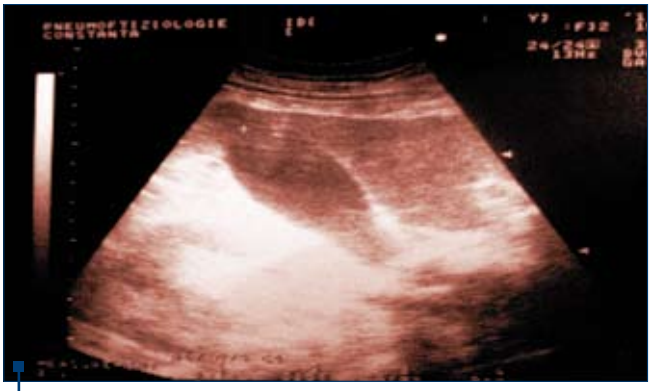

Figure 3. Chest Ultrasound reveals, in June 27th 2011, a pericardial effusion 10 cm x 11 cm consistent with a potential pericardial cyst

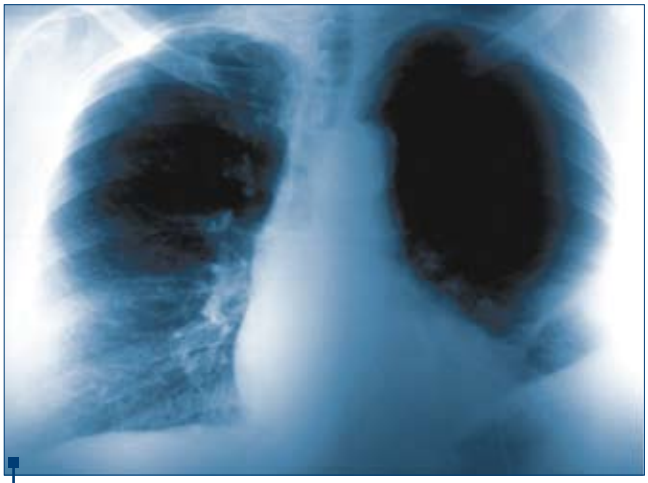

Figure 4. Chest X-ray performed in September 2011, one month after surgical intervention, revealed an ascended left hemidiafragm, left ventricular hypertrophy

2011, the patient was admitted in our respiratory clinic for evaluation. Chest X-ray showed a left ventricular hypertrophy (Figure 4). COPD was diagnosed with 56% reduction of FEV1 and FVC/FEV1 <70%.

Discussion

Anterior mediastinal masses comprise a diverse group of tumors and occasionally manifest as a cystic lesion, including mesothelial cysts. Congenital cysts of the mediastinum are an uncommon but important diagnostic group, representing 12 to 30% of all mediastinal masses[1]. Pleuropericardial cysts (PPC) are congenital coelomic cysts, which originate during embryogenesis of the pericardium and pleura[3]. PPC are identified in the fourth or fifth decade of life and represent 5-7% of all the mediastimum tumors. Although most are congenital, a few cases of acquired pericardial cysts do exist and are, frequently, located in the right hemithorax in the costophrenic angle (70%) and are asymptomatic. Most of them have a diameter of 3 to 6 cm, but Leigh and Weens indicated they may be as large as a grapefruit[3].

Our patient has a left paracardiac mass, mimicking a cardiomegaly, and we established its cystic nature by ultrasound examination. CT should be the initial procedure for evaluating most patients with mediastinal abnormalities detected by plain chest radiography. Currently, mediastinal cysts can be accurately diagnosed with imaging modalities such as MRI and ultrasonography.

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. Although CT is considered a better investigation than chest radiography in determining the pathologic diagnosis of an anterior mediastinal mass, in our case report CT proves still poor at making that prediction with confidence. However, several anterior mediastinal masses could be diagnosed accurately by ultrasonography.

The final differential diagnosis must be established with Morgagni hernia, pericardial fat pad and bronchogenic cysts, which reveals quite similar conventional X-ray findings, although their occurrence in the left cardiophrenic space is quite atypical.

When pleuropericardial cysts are symptomatic, ussually when they are large, the symptoms are generally dominated by respiratory signs, such as dyspnea. In some cases, the manifestations of inferior vena cava compression may be present.

Diagnosis is usually done by Roentgenography of the thorax or CT scan.

In the past, many presumed pericardial cysts were not surgically removed because of a characteristic appearance and benign behavior[3]. 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[2].

Indications for surgery, in the presented case report, included the presence of symptoms and uncertain radiographic diagnosis. 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 sugestive for pleuropericardial cyst.

Conclusions

Some feature of the case are worth mentioning: the large size of pericardial cyst at the upper limit of the data reported in the literature, which mimics cardiomegaly, the hemoptoic onset in a hypertensive patient, heavy smoker; the late suspicion of pleuropericardic cyst through pleural echographic exam; the atypical localization; the facilitated certain diagnosis by surgery and hystological exam; the favorable postoperative prognosis; and all morbidities cofound (Pulmonary Tuberculosis, bronchiectasis, COPD).

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