Pericardial mesothelial inclusion cyst leading to right heart failure

Perikardiyal mezotelyal inklüzyon kisti neden olma sağ kalp yetmezliğine

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Pericardial mesothelial inclusion cysts are usually benign intrathoracic lesions, which locate aberrantly and originate from the coelomic space. Despite asymptomatic nature, they may infect or compress to the adjacent structures in larger sizes. The diagnosis is usually made coincidentally by chest X-ray. In this article, we present a 57-year-old female case who was admitted with the complaints of dyspnea and swollen legs and in whom a paracardiac mass was detected by chest X-ray radiography in the light of literature data.

Key words: Inclusion cyst; paracardiac mass; thoracotomy.

Perikardiyal mezotelyal inklüzyon kistleri, aberran yerleşme olmuş ve çölomik boşluktan köken alan genellikle benign toraks içi lezyonlardır. Asemptomatik olmalarına karşın, büyük boyutlara ulaşıp komşu yapıları enfekte edebilir veya bu yapılara bası yapabilir. Tanı sıklıkla göğüs röntgeni ile tesadüfen konur. Bu yazıda, nefes darlığı ve bacaklarda şişlik yakınması ile başvuran ve çekilen göğüs röntgeninde parakardiyak kitle tespit edilen 57 yaşında bir kadın hasta, literatür verileri eşliğinde sunuldu.

Anahtar sözcükler: İnkluzyon kisti; parakardiyak kitle; torakotomi.

Pericardial mesothelial inclusion cysts are generally benign intrathoracic lesions which locate aberrantly and originate from the coelomic space. They may be acquired or congenital and can be found in any localization, varying from the upper mediastinum to the diaphragm. However, they are especially found at the right front cardiophrenic angle. These cysts are rare, occurring at a rate of 1/100,000, and they constitute 7% of all mediastinal tumors.

There are no findings on a physical examination that suggest the presence of pericardial cysts. They are usually detected on routine radiological studies or incidentally in the operating room. In general, they are asymptomatic, unless they infect or compress on the surrounding structures when they increase in size. Pericardial cysts are commonly unilocular, but they may appear to be multilocular due to constrictive fibrous bands. The variety in location of mesothelial cysts is related to embryological reasons. They can occur anywhere but are only rarely found in the mediastinum between the anterior chest wall and the right heart. Fusion of the mesenchymal coelomic lacunae originates from the pleural and pericardial cavity on one side and the peritoneal cavity on the other, and they are divided by the anteroposterior development of the septum transversum. Incomplete fusion of a lacunae, especially at the level of the pericardial coelom, may result in the formation of a mesothelial cyst, and this kind of fusion or the secondary migration of an isolated element can also occur at the level of the parietal pleura, mediastinal pleura, or septum transversum, which might explain the unusual locations of mesothelial cysts.

These cysts are histologically lined with a single layer of mesothelial cells, with the remainder of the wall being composed of connective tissue with collagen and elastic fibers. Additionally, they contain a clear, water-like fluid.

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Controversy exists regarding the optimal treatment of pericardial mesothelial inclusion cysts, but the defining factors are the qualification of the lesions and whether or not there are symptoms related to compression. Many authors have suggested surgical excision of the mass, but this is a complicated procedure and has the risk of malignancy. Endoscopic resection or percutaneous aspiration of the cyst are also treatment options.

CASE REPORT

A 57-year-old female patient was admitted to our facility with a one-year history of dyspnea and leg swelling. Her heart rate was 96 bpm, and her blood pressure was 150/90 mmHg. In addition, jugular venous distention, hepatomegaly via palpation over the right subcostal region, and 2+ lower extremity edema were detected on a physical examination. The New York Heart Association (NYHA) functional capacity was class II, and there was no cardiac murmur. Furthermore, the results of an electrocardiogram (ECG) were normal. However, a mass with opacification was detected and visualized near the right side of the heart on chest X-ray radiography (Figure 1), and contrast-enhanced thoracic tomography found a well-restricted, heterogeneous, multilocular mass of approximately 5x5 cm in size located beside the right atrium (Figure 2). In addition, a hyperechogenic mass lesion measuring approximately 5x5 cm was seen near the right atrium on transthoracic echocardiography (TTE). This cystic, paracardiac mass, which was located in front of the
right phrenic nerve and connected to the pericardium via a pedicle, was then successfully removed by a right lateral thoracotomy (Figure 3). The pericardium was adherent to the right atrium, so the mass was excised along with the pericardium. The surgical specimen was examined, a single-layer, smooth epithelium was found in the cyst structure, which led to the conclusion by a pathologist that it was a mesothelial inclusion cyst (Figure 4). After the surgical procedure, the patient had an uneventful recovery and was discharged from the hospital on the seventh postoperative day.

**DISCUSSION**

A pericardial cyst is typically filled with fluid, has a thin wall, and contains no solid components in the mediastinal structure. It is usually located in the immediate proximity of the heart or can possibly originate from it.

An isolated cystic mass located adjacent to the heart primarily should raise the possibility of a neurenteric cyst, a bronchogenic, esophageal cyst, lymphangioma, or a pericardial cyst.

Symptoms of atypical chest pain, dyspnea, and persistent cough are indicated in about one-third of the patients with pericardial mesothelial inclusion cysts. Other complications that have been reported in the literature include rupture, cardiac tamponade, mitral valve prolapse, obstruction of the right main stem bronchus, atrial fibrillation, and erosion into adjacent structures, for example the right ventricular wall or superior vena cava (SVC).

Myxomas are the most common primary cardiac neoplasms, constituting about half of all cardiac tumors, followed by lipomas. Cardiac lipomas are distributed throughout the heart but are usually located in the subepicardial region.

With the exception of myomas, lesions in the right side of the heart should raise the suspicion of malignancy, and metastases through venous invasion are more common with these lesions than with primary malignant cardiac tumors.[7] Beyond hematogenous and lymphatic spread, direct continuous extension to the myocardium or pericardium may also happen. Among neoplasms, melanoma has the highest frequency of metastases to the heart, followed by malignant germ cell tumors, leukemia, lymphoma, lung cancer, and other various sarcomas.[8]

Additionally, lymphomas also have a very high frequency of metastases to the heart. Although primary cardiac lymphoma is very rare, some patients with disseminated lymphoma have cardiac metastases. In most cases, however, lymphomas typically infiltrate the myocardium and pericardium.

We preferred to treat our patient with surgical excision because he was symptomatic, and we believe this should be the treatment of choice for masses which cause complicated, severe symptoms and for those that have reached huge dimensions or have the risk of malignancy. There are some studies which have recommended excision with cardiopulmonary bypass (CPB),[5,6] but we were able to excise the mass in our patient successfully via a lateral thoracotomy without performing CPB because of its location.

In conclusion, we believe that surgical resection of a pericardial cyst should be performed when malignancy is suspected, when the diagnosis is uncertain, or when the patient has symptoms due to complications and is unresponsive to other treatment options.

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