A Case of Idiopathic Effusive-Constrictive Pericarditis

To the Editor:

Effusive-constrictive pericarditis (ECP) is a rare form of pericardial syndrome, where cardiac constriction occurs in the presence of significant pericardial effusion. Patients present cardiac tamponade, although post-pericardiocentesis symptoms persist, thus revealing cardiac constriction which is resolved only after pericardiectomy. Although the condition was first described some years ago, only a few cases have been reported in the medical literature. We present a case of ECP in a patient with a history of mitral valve replacement.

A 55-year-old man with mitral valve replacement in 1998 and postpericardiectomy syndrome, characterized by fever and tiredness with minimal effort; following this treatment, the patient remained asymptomatic. In April 2004, he was seen for severe pericardial effusion and cardiac tamponade that required pericardiocentesis. Afterwards, he initially remained asymptomatic. He was readmitted in July 2004 and reported that he felt well after hospital discharge, but a few weeks later, began to feel tired after considerable exertion, and then with only moderate effort. At the time of readmission, the patient’s blood pressure was 110/70 mm Hg; there was no paradoxical pulse, but the jugular vein was enlarged. Cardiac auscultation was rhythmic at 70 bpm
and prosthetic sounds were normal. He presented slight hepatomegaly and edema with fovea in the lower extremities.

The electrocardiogram showed sinus rhythm. The echocardiogram disclosed a protodiastolic septal notch, restrictive transmitral flow, and respiratory variation in mitral E wave velocity. The artificial valve was functioning normally.

The chest computed tomography scan showed moderate, predominantly posterolateral, pericardial effusion with pericardial thickening (Figure 1).

We suspected pericardial constriction and requested right and left catheterization. We also observed equalization of end-diastolic pressures in the right and left chambers. The diastolic pressure curve of the left ventricle showed the typical "square root" or "dip-plateau" morphology (Figure 2).

Based on a diagnosis of ECP (probably idiopathic), pericardiectomy was performed. The histologic study showed hyalinized fibrous tissue and lymphoplasmocytic infiltrate, consistent with a chronic inflammatory response. Clinical progress was favorable.

Whenever a patient presents right heart failure that is not explained by systolic dysfunction, valve disease, or right heart disease, ECP should be suspected. A differential diagnosis with cardiomyopathy is essential, since ECP presents relief of postpericardiectomy symptoms. Visualization of pericardial thickening or calcification on imaging studies is extremely helpful in the diagnosis.5

On echocardiography, ECP shows reciprocal respiratory changes between both ventricles.6,7 Cardiac catheterization reveals an increase and equalization of end-diastolic pressures, reciprocal postinspiratory changes6,7 (Figure 2, arrow), and left ventricular dip-plateau morphology.

Effusive-constrictive pericarditis simultaneously presents pericardial effusion with hemodynamic compromise and pericardial constriction. The diagnosis is established postpericardiocentesis, if the patient does not have complete symptomatic relief and manifests constriction,2 as in the case we present. The importance of the diagnosis lies in the need for resection of the visceral pericardium. Pericardiectomy is the treatment of choice in symptomatic patients.

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Figure 1. Chest computed tomography: moderate, primarily posterolateral, pericardial effusion (asterisk), and pericardial thickening (arrow).

Figure 2. Catheterization. A: LV curve with diastolic dip-plateau and right atrial curve (equalized diastolic pressures). B: left (LV) and right (RV) ventricular pressure curve (equalized diastolic pressures). LV-RV pressure discordance during inspiration (arrow).
REFERENCES


Pericardial Cyst: All’s Well That Ends Well

To the Editor:

Pleuropericardial cyst is a rare lesion, with a benign clinical course in most cases, that is detected incidentally on chest x-ray and may be confused with a neoformative process of the lung. We present a case of pleuropericardial cyst in a female smoker.

A 55-year-old woman came to the emergency room for chest pain. Relevant history included smoking (15 pack-years) and diabetes treated with oral hypoglycemic agents. She had not presented respiratory or cardiovascular symptoms of interest to date. Because the medical history and the initial physical examination revealed no relevant data, a hemogram and biochemistry were requested, with all results within normal limits. Electrocardiography was normal and abdominal sonography ruled out acute bile disease. The posteroanterior chest x-ray showed a rounded, right paracardiac image of approximately 5 cm (Figure). The image was not seen in a previous x-ray brought by the patient (from 6 years earlier) and therefore, she was admitted by our department for further study of the lesion.

The next test requested was a chest computed tomography (CT), which disclosed a lesion measuring 6 cm at its largest diameter adjacent to the right side of the pericardium, with no change observed after administration of intravenous contrast material. The lesion had cystic characteristics and no definable wall, corresponding radiologically to an uncomplicated pleuropericardial cyst; the rest of the study was normal.

The patient was discharged and an outpatient echocardiography was scheduled, which confirmed the diagnosis of uncomplicated pleuropericardial cyst. She remained asymptomatic, and clinical follow-up was carried out by the outpatient clinic, with echocardiograms performed regularly.

The incidence of pleuropericardial cyst has been estimated at 1 per 100,000 cases, accounting for 5%-10% of all mediastinal tumors. Most are congenital, although several acquired cases with an infectious, inflammatory, or traumatic etiology have been described. They are usually identified in the fourth or fifth decade of life. The most frequent site is the right costophrenic angle (70%), followed by the left costophrenic angle (10%-40%), although other unusual sites have also been reported, for instance, the vascular hila, the superior mediastinum, or the left heart border. There are usually no symptoms, with the cyst being an incidental finding on conventional chest x-ray, identified as a homogeneous, radio-opaque oval lesion. However, episodes of chest pain, tachycardia, persistent cough, and cardiac arrhythmias have been described. These symptoms can result from the pressure of the cyst on adjacent organs.

This cyst generally presents a favorable prognosis, although its natural history is not completely known. The literature reports cases ranging from spontaneous disappearance to recurrence after surgical resection, as well as serious, even fatal complications such as cardiac tamponade due to rupture, cardiogenic shock, erosion of the vena cava, torsion, cardiac compression, or infection of the cyst. The differential diagnosis is extensive and includes, among others, Morgagni hernia, pericardial fat, ventricular aneurysm, cardiac or bronchogenic tumors, and evidently all.