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Case Report



The Insidious Course of Chronic Constrictive Pericarditis: A Case Report

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Abstract:

Chronic constrictive pericarditis is an inflammatory disease of the pericardium, characterized by a fibro-calcareous pericardial thickening. This thickening is responsible for an impairment of ventricular filling, thus defining adiastole (1). This almost always affects both ventricles, increasing interventricular interdependence and leading to equalization of right and left filling pressures. The etiology of the condition has changed dramatically over time (2). Tuberculosis, once a common cause, has given way to a multitude of other causes. The clinical picture is often non-specific. The diagnosis may be difficult in some cases. The diagnosis is made mainly by chest X-ray, cardiac echo-Doppler and cardiac catheterization. Subtotal pericardectomy is necessary in severe forms of the disease (1). We report the medical observation of a 37-year-old patient admitted to our department for management of predominantly right heart failure.

Keywords: chronic constrictive pericarditis; pericardectomy; case report

Introduction:

Chronic constrictive pericarditis is a rare condition characterised by a rigid pericardium limiting myocardial compliance, the main aetiologies of which are infection, thoracic radiotherapy and heart surgery. The clinical presentation is that of heart failure, thus aspecific, often leading to a delay in diagnosis of several years. The diagnostic approach includes echocardiography and cardiac catheterisation. A surgical treatment, pericardiectomy, makes this pathology curable if it is identified in time (**3**). Given the diagnostic difficulty of this condition in this case, we will discuss the need for other imaging methods, including cardiac CT and MRI.

Observation:

The patient was 37 years old, with no notable pathological history and no cardiovascular risk factors; he had been suffering from neglected edema of the lower limbs for 5 years; the evolution was marked by the onset of NYHA stage II dyspnea three months prior to his admission, which progressively became stage IV, associated with alteration of the general condition, with signs of right heart failure on examination, in particular spontaneous turgidity of the jugular veins, medium-sized ascites, and edema of the lower limbs reaching the middle of the thigh.

The electrocardiogram was in sinus rhythm with microvoltage predominantly in the frontal leads. The chest X-ray in profile incidence showed calcification of the pericardium (figure 1). Transthoracic echocardiography revealed a thickened, highly calcified pericardium without pericardial effusion; a disruption of the alignment between the posterior wall of the left ventricle and the posterior wall of the left atrium observed in long axis parasternal incidence (figure 2), the ventricles were non-dilated with preserved systolic function with paradoxical movement of the interventricular septum, and moderate dilatation of the atria; as well as a very dilated and uncomplicated inferior vena cava, with a restrictive mitral flow E/A at 2. 5 with a short deceleration time of 76 ms, significant respiratory variations on the mitral and aortic flow (figure 3), a dip plateau aspect on the pulmonary flow with a PHT shortened to 58 ms, an expiratory inversion of the diastolic flow in the suprahepatic veins.

The thoracic CT scan showed a bilateral pleural effusion of moderate size, pericardial calcification, peritoneal effusion, and dilatation of the inferior vena cava.

The biological workup revealed a microcytic hypochromic anemia with an inflammatory syndrome, notably a CRP of 160 and a hyperleukocytosis of 9600 with a predominance of neutrophils, a negative procalcitonin, and the rest of the workup was unremarkable, notably the search for tuberculosis by the genxpert.

In view of these clinical, radiological, echocardiographic and biological aspects, the diagnosis of chronic constrictive pericarditis (CCP) was retained. The patient was put on furosemide with potassium supplementation, then transferred to the cardiovascular surgery department where he benefited from a subtotal pericardectomy with a simple operative follow-up and good clinical evolution.

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Discussion:

The diagnosis of chronic constrictive pericarditis (CCP) must be evoked in the presence of a picture of heart failure, most often predominantly right-sided, which is not proven (4). As a rule, this is a diagnosis of elimination or guided by specific elements related to the main etiologies: discovery of pericardial calcifications, history of recurrent acute pericarditis, history of tuberculosis, thoracic irradiation, or heart surgery. Frequently, no etiology is found suggesting an idiopathic origin of PCC (5) as in our patient's case. Investigations pointing to the diagnosis of pericardial constriction are as follows:

Electrocardiogram:

Electrical changes are common but nonspecific (bifid P wave, microvoltage, electrical alternation, and diffuse repolarization abnormalities). Atrial fibrillation is common in advanced forms (20% of cases). First-degree atrioventricular block is noted in 8 to 10% of cases (1).

Chest X-ray:

The cardiothoracic index is normal in constrictive pericarditis. Cardiomegaly may be observed in case of significant pericardial thickening and especially in case of associated effusion. It is necessary to look for calcifications, which are more visible on the profile film. It is sometimes necessary to evaluate the kinetics of the heart by scopy, which allows a better view of the pericardial calcifications (1)

Doppler echocardiography:

As illustrated in our case, echocardiography plays a crucial role in the diagnosis of constrictive pericarditis, but it is often insufficient insofar as it is an operator-dependent examination, limited by the echogenicity of the patient, which does not allow analysis of the pericardial structure. The positive diagnosis of chronic constrictive pericarditis with cardiac Doppler echocardiography is of variable difficulty depending on the stage of the constriction. The analysis must be meticulous to look for signs of constriction and adiastole. Ultrasound signs in TM and two-dimensional (2D) mode of chronic constrictive pericarditis are neither sensitive nor specific. In 2D mode, the diagnosis is evoked by the presence of pericardial thickening in association with moderate dilatation of both atria and the inferior vena cava in contrast to normal-sized ventricles (1-4). A disruption of the alignment between the posterior wall of the left ventricle and the posterior wall of the left atrium is sometimes observed in parasternal long axis incidence (1); as found in our patient. Doppler analysis is an essential step in the diagnosis of pericardial constriction. It consists of an analysis of the mitral and tricuspid flows and of the flows in the pulmonary veins and in the suprahepatic veins. This study allows to look for signs of adiastole and especially respiratory variations of the flows. These variations make it possible to differentiate a PCC from a restrictive cardiomyopathy (CMR). They are explained by the phenomenon of ventricular interdependence and by the dissociation between intra-thoracic and intra-cardiac pressure responsible for a leftward septal deviation during inspiration (1).

Cardiac catheterization:

This confirms adiastole by demonstrating positive protodiastolic ventricular pressures, mesoteliastolic dipole and equalization of mean right pressures; and may demonstrate respiratory variation in pressures. In the absence of obvious abnormalities, an intravenous filling test can be performed during catheterization to unmask adiastole (5).

Cardiac CT scan:

In the diagnosis of pericardial constrictions, the CT scan shows a thickened (greater than 4 mm) and calcified pericardium in the presence of signs of adiastole. It should be emphasized that the presence of calcifications does not automatically imply the existence of a cardiac constriction, and that the severity of the constriction is not proportional to the size of the calcifications. On the other hand, a non-thickened pericardium on CT images does not eliminate the diagnosis of chronic constrictive pericarditis. CT is a fairly rapid examination but it exposes the patient to radiation as well as to the nephrotoxic and allergic risk associated with the use of iodinated contrast medium. Therefore, contraindications must be respected, namely renal insufficiency, severe allergy to iodine and pregnancy (6).

Cardiac MRI:

The advantage of MRI resides in the fact that it is a non-invasive exploration since it does not use ionizing radiation and does not require the use of nephrotoxic contrast medium. In PCC, MRI provides a positive and especially topographic diagnosis of the constriction lesions in order to guide the surgical procedure (6). In practice, there are two categories of magnetic resonance imaging abnormalities: morphological abnormalities (pericardial thickening > 4 to 6 mm; pericardial calcifications, vena cava and suprahepatic veins are dilated with reduced inferior vena cava compliance on inspiration, moderate dilation of the atria, ventricles are usually small in volume with typically a tubularized appearance of the VD, pericardial effusion) and functional abnormalities: septal fontanelization and pathologic ventricular coupling (6)

In our patient's case, we did not perform a cardiac MRI because the transthoracic echocardiography as well as the thoracic CT

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scan was in favor of a PCC, the cardiac catheterization was not performed because we do not have the technical platform.

Conclusion:

The diagnosis of PCC can be difficult requiring several examinations in particular echocardiography and cardiac catheterization. Visualization of the pericardium on CT or MRI is necessary in some doubtful cases. However, the most important diagnostic tool remains clinical suspicion in the presence of signs of right heart failure not explained by pulmonary pathology or left heart disease (1).Figures:

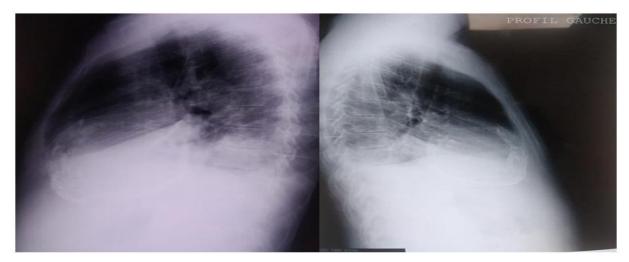


Figure 1: X-ray of the left and right chest showing calcification of the pericardium



Figure 2: Transthoracic echocardiography, long axis parasternal section, showing disruption of the alignment between the posterior wall of the left ventricle and the posterior wall of the left atrium



Figure 3: Cardiac echodoppler showing variations in mitral flow on inspiration as well as the restrictive aspect of mitral flow with a short deceleration time

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