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Aortic Bicuspidity and Chronic Constrictive Pericarditis: New Association

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Abstract **Case Report**

***Introduction*:** We report the observation of a patient hospitalized in cardiology for exploration of dyspnea associated with a picture of right heart failure for which a chronic constrictive pericarditis is evoked in front of pericardial calcifications at the trans-thoracic echography with aortic bicuspidia. This didactic observation is the occasion to recall the different complementary examinations allowing to affirm the constriction and thus to eliminate a restrictive cardiopathy.

Keywords: cardiology, heart diseases, the trans-thoracic echography, aortic bicuspidia.

INTRODUCTION

Chronic constrictive pericarditis is a rare and serious disease defined by abnormalities of the pericardium (fibrosis, adhesion, calcifications, thickening), becoming a rigid inextensible shell altering the diastolic expansion of the heart and thus the filling defining adiastole [1]. The frequency of this pathology is estimated to be about 0.5 to 2% of heart diseases. It represents 0.6% of the procedures in a cardiovascular surgery department [2]. The etiology of pericardial constrictions has been modified by prophylaxis and anti-infective treatments. Although tuberculosis remains a classic cause, it is less frequent than constrictions secondary to cardiac surgery or mediastinal radiotherapy, which are clearly increasing in developed countries. Other etiologies are rarer, namely chronic renal failure, systemic disease (lupus, rheumatoid arthritis), bacterial infection or thoracic trauma [3, 4]. The most consistent symptom leading to consultation is exertional dyspnea sometimes associated with asthenia and exertional hepatalgia [2]. On clinical examination, signs of right heart failure are most often observed. Paradoxical arterial pulse (inspiratory decrease in blood pressure of 10 mmHg or more) is more inconsistent and may be absent in patients with pure constriction [5]. TM and two-dimensional ultrasound signs are not sensitive or specific. The main abnormalities reported are abrupt protodiastolic anterior motion of the interventricular septum followed by a posterior protrusion [6]. Cardiac catheterization remains the reference examination showing equalization of ventricular diastolic pressures with a dip-plateau appearance [7]. CT and MRI provide an excellent description of the anatomy of the pericardium [8]. The only curative treatment of pericardial constrictions is surgical, consisting of pericardectomy with decortication of the pericardial leaflets [9].

Aortic bicuspidism (AB) is the most common congenital heart defect. It is of great interest because this pathology, long considered benign, is in fact protean. It can exceptionally be responsible for severe heart failure in newborns and is the most frequent cause of aortic valve replacement in adults. Finally, it can be associated with an aneurysmal dilatation of the ascending aorta at high risk of dissection [10]. The classification recently proposed by Sievers [11] based on the number of raphe (folding of the sigmoid without true commissure) shows that bicuspidia with 1 raphe (long wrongly called "pseudobicuspidia") are the most frequent and often associated with stenosis and/or valve leakage. The presence of an ectasia, a true "aortopathy", puts the patient at risk of aortic dissection. The pathophysiology of the cystic medial necrosis of the aortic wall present in these patients is multifactorial (genetic, structural, hemodynamic factors). The presence of aortic dilatation leads to regular monitoring of the evolution.

MEDICAL OBSERVATION

We report the case of a 38 year old patient, chronic smoker with 35 BP. He was admitted for chronic constrictive pericarditis on aortic biscuspidia, in right heart decompensation at the stage of anasarca made of bilateral pleural effusion of great abundance, ascites of great abundance and minimal pericardial epenches, with at the evacuation puncture a citrine yellow liquid. On trans-thoracic echography, a calcidated pericardium was found, with the LV almost adjacent to the pericardium and a clear relaxation anomaly (Figure 1). A biological workup was performed without any particularity with a protid level of 77 g/L and albumin of 39 g/L, a quantiferon and negative sputum BK. An etiological workup was performed: a cyto-bacterial-chemical study, a thoracic angioscanner finding an aspect of pulmonary embolism with bilateral pleural effusion of great abundance without visualized calcification images of calcifications, an abdominal echo highlighting an ascites of great abundance, a cardiac MRI showing thickening of the pericardium enhancing late after gadolinium, and a cardiac KT showing an equalization of the ventricular diastolic pressures with characteristic aspect in dip-plateau and elevation of the right ventricular diastolic pressures with inspiration while the left ventricular pressures decrease. The initial therapeutic conduct was to control his decompensation and to put him under aspegic and LMWH in curative dose, and of course the treatment of his PCC? decortication.

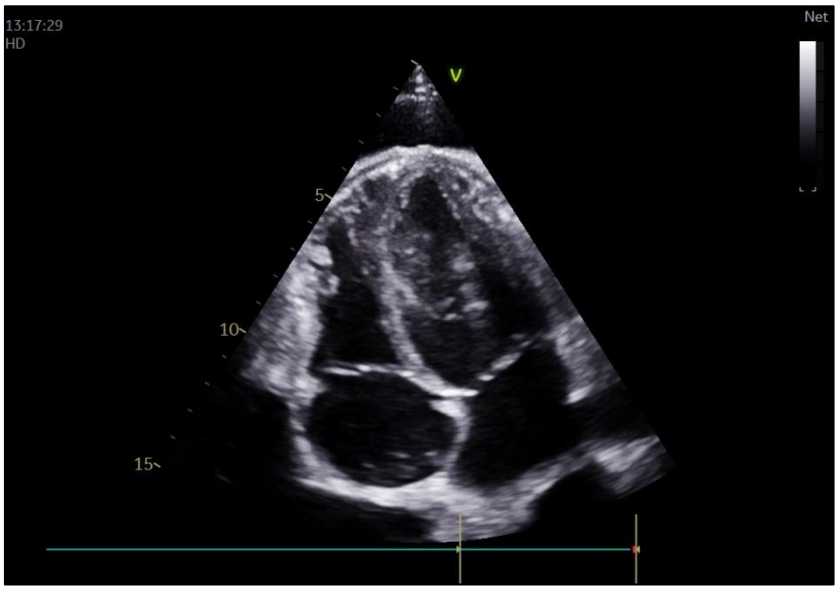

Figure 1: Image showing pericardial calcifications

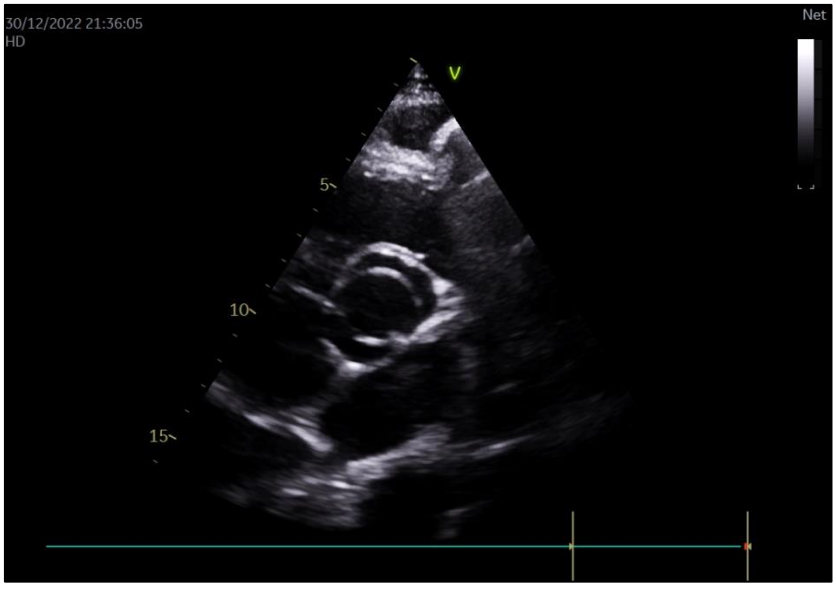

Figure 2: Image showing aortic bicuspidity

DISCUSSION

Pericardial calcifications are part of the healing process of old pericarditis. Tuberculous pericarditis and postoperative pericarditis are two classical etiologies but calcifications may appear in the course of any old pericarditis. On the CT scan, spontaneously hyperdense, linear and/or nodular lesions of the pericardium may be found, more or less extensive, especially anterior.

Echocardiography is an excellent examination to confirm the presence or not of a pericardial effusion and to study its functional repercussions. It also allows the differential diagnosis between constrictive and restrictive pathology. On the other hand, it does not allow a direct measurement of the thickness of the pericardium.

MRI visualizes well the pericardium which appears as a hypointensity line between the epicardial and pericardial fat, with a maximum thickness of less than 4 mm. A thickening of the pericardium that enhances late after gadolinium suggests constrictive pericarditis (present on MRI in 88% of cases). It will be reinforced in the presence of a narrow and elongated right ventricle, dilatation of the right atrium, paradoxical movement of the interventricular septum in cine-MRI sequence and signs of elevated right filling pressures (dilatation of the vena cava, pleural effusion, hepatic congestion, ascites) [12].

Cardiac CT is an excellent examination to judge the thickness of the pericardium and the presence of calcifications. The detection of pericardial effusions is easy. In addition, it can demonstrate the quality of the effusion (liquid vs. hemorrhagic), evaluate the impact on the filling of the right cavities and objectify possible paracardial masses at the origin of the effusion. In addition, cine images allow the assessment of ventricular interference.

Aortic bicuspidism is a frequent pathology, but not well known by anesthesiologists and obstetricians. It is predominantly male with a hereditary factor. The fusion of two of the three aortic valves often has little hemodynamic consequences initially, but exposes the patient to the progressive appearance of degenerative lesions leading to narrowing of the aortic surface or to valvular insufficiency due to coaptation defects [13, 14]. It represents only about 7% of aortic stenoses operated on, whereas it represents 30 to 50% of aortic stenoses operated on in Western countries [15, 16]. It can be stenosing or leaky. It is either complicated by aneurysm of the ascending aorta or infective endocarditis, or associated with a bitronchial coronary lesion requiring consequently a replacement of the ascending aorta in the first 2 cases and a coronary artery bypass graft in the latter. The follow-up of patients shows that aneurysm of the ascending aorta is a serious condition with a high mortality rate and dramatic complications (dissection, rupture) as soon as the diameter exceeds 6 cm. 46% of patients with an aneurysm larger than 6 cm will die within 5 years. The main predictive factor of complication remains the size of the aneurysm. In addition, severe aortic insufficiency is also present [17].

This article has tried to detail 2 major pathologies, one congenital and the other aquise. An attempt was made to investigate the different evolution of aortic bicuspidism, in particular chronic constrictive pericarditis. Except that no correlation was found between the two.

CONCLUSION

In conclusion, PCC is a rare but serious disease most often revealed by a picture of advanced right heart failure. It is sometimes difficult in the absence of pericardial calcifications to differentiate this condition from restrictive cardiomyopathies which are the essential differential diagnosis.

On the other hand, aortic bicuspidism remains a frequent but unrecognized disease. Its association with chronic constrictive pericarditis is very rare. Their management implies a multidisciplinary approach in a specialized center. This observation raises the question of the management of patients with chronic constrictive pericarditis on aortic bicuspidia. It is necessary to treat effectively and appropriately in order to improve the survival of the affected subject.

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