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Case report

A complicated diagnosis of constrictive pericarditis in a patient with atrial fibrillation – The importance of temporary pacing inducing regular heart rhythm during invasive hemodynamic study

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ABSTRACT

Background: Constrictive pericarditis (CP) is a rare disease frequently with nonspecific initial clinical manifestations. Transthoracic echocardiography (TTE) is a key imaging method for the CP diagnosis. However, concomitant atrial fibrillation (AFib) may complicate the correct diagnosis, since some typical echocardiographic CP markers, especially those based on their respiratory changes may not be obvious or routinely evaluated.

Case report: We present a case of initially asymptomatic 61-year-old male with elevated cholestatic enzymes of unclear etiology detected repeatedly over a period of 3 years, with chronic AFib but without signs of heart failure. Prior to planned liver biopsy, a comprehensive TTE was performed showing some signs indicative of CP (including septal bounce and shift, annulus reversus and paradoxus and inferior fixed caval vein dilatation). Other TTE parameters based on respiratory variation, which are typically observed in CP with sinus rhythm, were unreliable in AFib. Cardiac computer tomography and magnetic resonance showed pericardial thickening and calcification supporting but not confirming the CP diagnosis. Only after right ventricular pacing during catheterization, the typical discordance of peak systolic right and left ventricular pressure during respiration confirmed CP diagnosis. After pericardiectomy, cholestatic enzymes decreased supporting the CP causal role.

Conclusions: CP should be considered in unexplained increase of cholestatic enzymes. In AFib, the TTE parameters based on respiratory variation may not be useful. However, a combination of the remaining TTE parameters can indicate CP and trigger further investigation. Temporary pacing to avoid beat-to-beat variability in AFib can ''unmask'' the ventricular pressures discordance, and thus be helpful in unclear CP cases.

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Introduction

Constrictive pericarditis (CP) is a relatively rare disease resulting from chronic inflammation and fibrosis of the pericardium [1]. The decrease in pericardial compliance leads to impaired diastolic filling with increased filling pressures, ventricular interdependence with enhanced variation left and right filling pressures and increased respiratory variation of transmitral and transtricuspid blood flows as well as patterns of hepatic and pulmonary vein flows. Non-invasive imaging methods (including echocardiography) are the key diagnostic tools for CP recognition and differentiation from restrictive cardiomyopathy. Some of the most important echocardiographic signs of CP are related to hemodynamic changes induced by enhanced respiratory variation of intracardiac pressures and flows [2]. While these signs are obvious in sinus rhythm, they can be difficult to appreciate in atrial fibrillation (AFib) when there is great beat-to-beat variation. Most commonly, the first reported symptoms are signs of heart failure (HF) with dyspnea and edema [3]; however, rarely the first symptom of CP may be a subclinical abdominal congestion with changes in liver enzyme levels.

We report a difficult and complicated CP diagnosis in a patient with permanent AFib in whom the first but unusual CP manifestation was a long lasting elevation of cholestatic enzymes preceding the abdominal discomfort with no other signs of HF. All the imaging methods including transthoracic echocardiography (TTE), cardiac computer tomography (CCT) and magnetic resonance (CMR) were supporting but not confirming the CP diagnosis. Thus, a temporary ventricular pacing during invasive hemodynamic examination had to be performed to overcome the beat-to-beat variation frequently seen in atrial fibrillation that eventually led to the correct diagnosis.

Case presentation

A 61-year-old male with asymptomatic elevation of cholestatic enzymes over past 3 years (2–10 times above normal range) was referred to our hospital for reevaluation. The patient was already thoroughly examined by a local gastroenterologist including repeated abdominal ultrasounds and blood tests. However, except for laboratory signs of a past asymptomatic hepatitis B without signs of reactivation or chronic hepatitis, no liver, pancreatic or biliary ducts pathology was found. Alcoholic, autoimmune and drug-induced etiologies were also excluded. Only a mild normocytic normochromic anemia was detected. The patient recently developed mild abdominal discomfort located in the right upper abdominal quadrant for last 3 months; however, the dyspepsia was occurring infrequently (twice a week) with no relationship to food intake or other activities. In view of new gastrointestinal symptoms and hepatopathy of unknown origin the gastroenterologist recommended liver biopsy as well as gastroscopy and colonoscopy which were scheduled to evaluate the cause of anemia. Cardiology reevaluation was required before the biopsy because of his past medical history of arterial hypertension, chronic AFib and a history of deep vein thrombosis. The patient signed informed consent and was further evaluated at our department.

Arterial hypertension was well controlled with beta-blocker and angiotensin receptor blocker for the last 5 years. The deep vein thrombosis was provoked by a leg injury 15 years ago. He also had a permanent AFib for 10 years and was regularly visiting his local cardiologist. Although recommended, he refused to use any anticoagulation therapy because of risk of injury during his training in historical martial arts and swordsmanship. His remaining medical history was unremarkable. He denied having any other cardiovascular symptoms including dyspnea, chest pain on exertion, palpitations, and edemas. According to past TTE reports performed elsewhere, his left ventricular (LV) ejection fraction (EF) was normal, right ventricular (RV) function was normal, both atria were enlarged, and no significant valvular disease was detected. Echocardiography results did not show any notable progression over last 5 years.

The patient was very well trained due to swordsmanship performed in iron armor weighing twenty kilograms. However, he admitted that he might have been recently more tired in comparison with his peers when practicing swordsmanship. Nevertheless, according to a standard NYHA classification he was still class I. Except for mild distension of jugular vein 4 cm above sternal angle, irregular heartbeats and a mildly sensitive palpation of right upper abdominal quadrant, his physical examination was unremarkable. His blood pressure was 120/80 mmHg, heart rate 70 beats/min, and peripheral oxygen saturation 98%. Except for mild non-specific repolarization abnormalities in inferior leads, ECG was unremarkable. Chest X-ray showed borderline size of the heart with small calcifications at the base of the heart, with no signs of congestion.

The echocardiography performed at our department confirmed the above-mentioned conventional echocardiographic results done previously elsewhere. In addition, we detected a decreased RV longitudinal systolic function. Moreover, a slight septal shift with respiration and also septal bounce (although not fully expressed) were noticed (Fig. 1A). The estimated pulmonary artery systolic pressure was borderline to mildly elevated 32–42 mmHg and the estimated high right filling pressure was high – 10–20 mmHg based on a distension of inferior caval vein with its minimal respiratory collapse (Fig. 1B). Contrarily, the estimated LV filing pressures seemed non-elevated when the ratio of early relaxation transmitral flow (E) to early myocardial relaxation evaluated

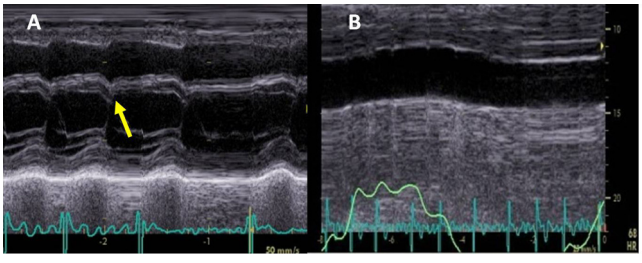

Fig. 1 – (A) An arrow indicates a brisk septal motion during diastole "septal bounce" characteristic for CP (M-mode of left ventricle from parasternal long axis view). (B) M-mode of dilated inferior caval vein with nearly no variation during respiration indicating high right ventricular filling pressures (subcostal view).

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