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Patient with constrictive pericarditis (RCD code: III)

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Abstract

52-year-old Caucasian man was admitted with clinical symptoms of chronic heart failure. Constrictive pericarditis was suspected almost 9 years before admission, but patient refused to under go diagnostic evaluation. Peripheral oedema, ascites, hepatomegaly, increased jugular veins pressure and pleural effusion were present on admission. Laboratory test showed increased level of brain natriuretic peptide with normal levels of liver enzymes and total bilirubin. Chest X-ray revealed calcifications of pericardium. In echocardiography biatrial enlargement was present with septal bounce, dilated inferior vena cava, respiratory variation of mitral and tricuspid inflow. Computed tomography showed massive calcifications of pericardium and hydrothorax of both pleural cavities. Cardiac magnetic resonance confirmed enlargement of both artia with dyssynchrony of intraventricular septum and pericardium thickening. Cardiac catheterization confirmed diagnosis of constrictive pericarditis. Patient was qualified for pericardiectomy. He died during procedure due to bleeding complications. According to ESC Guidelines, pericardiectomy is the treatment of choice in patients with significant symptoms with mortality rate 6–12%. JRCD 2015; 2 (5): 161–164

Key words: heart failure, pericardial thickening, pericardial calcifications

Case presentation

52-year-old Caucasian man was admitted to department of cardiology with symptoms of chronic heart failure. On admission peripheral oedema, ascites and hepatomegaly were present. Pressure in jugular veins was increased, breath sounds were silenced over the base of the right lung. Patient was in New York Heart Association (NYHA) functional class II. His heart rate was regular, 80 beats per minute (bpm), blood pressure was 110/80 mm Hg. Symptoms had been present since 2004, in 2005 right pleural cavity drainage was performed and obtained fluid was diagnosed toward multiple factors (tuberculosis, neoplasm, bacteria, cytology – all tests were negative). In computed tomography scan (CT scan) in 2012 calcifications of pericardium were detected.

In electrocardiography (ECG) sinus rhythm 80 bpm was present with P mitrale, low voltage in limb-leads and negative T wave in leads II, III, aVF, V1-V6. In biochemical analysis the level of N-terminal prohormone of brain natriuretic peptide (NT-pro BNP) was elevated – 735 pg/ml, other tests showed no significant changes: aminotransferases, gamma-glutamyl transpeptidase, alkaline phosphatase and total bilirubin were at normal range, no changes in morphology, tests toward hepatitis B, hepatitis C and HIV were negative. Additional tests showed decreased level of albumin with high level of gamma, alfa 1 and alfa 2 globulins. Analysis of the urine showed no signs of Bence-Jones protein presence. Chest X-ray revealed bilateral pleural effusion, moreover densities in mid and basal fields of right lung and calcifications of pericardium were present (Figure 1). In echocardiography biatrial enlargement was visible with normal-sized ventricles. Left ventricle (LV) ejection fraction was normal. Pericardial calcifications and septal bounce were visible. Symptoms of constriction were present: inferior vena cava was dilated (28 mm) with respiratory variation <50%, mitral flow was pseudonormal (E/A 1,3, E/E'<8), respiratory variation of tricuspid and mitral flow >25%. Moreover, mitral early diastolic velocity (E') of medial wall was higher than lateral wall – medial E' 19 cm/s, lateral E' 15 cm/s.

Ultrasound of the abdomen confirmed ascites and hepatomegaly. In bodypletysmography mild reduction of ventilation – restrictive

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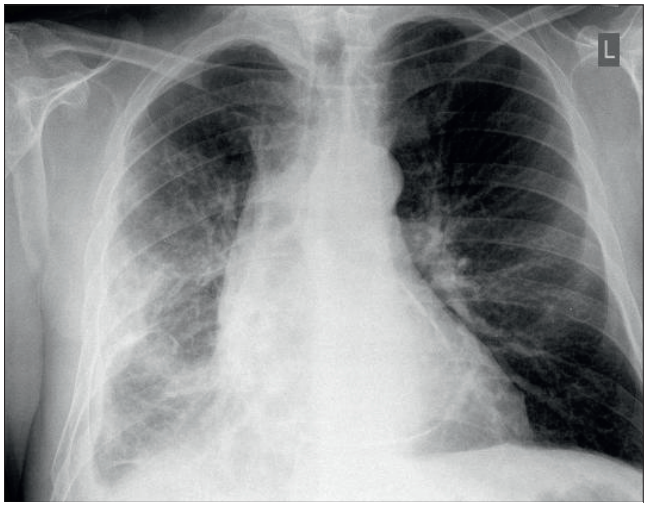

Figure 1. Chest X-Ray. Calcifications of pericardium

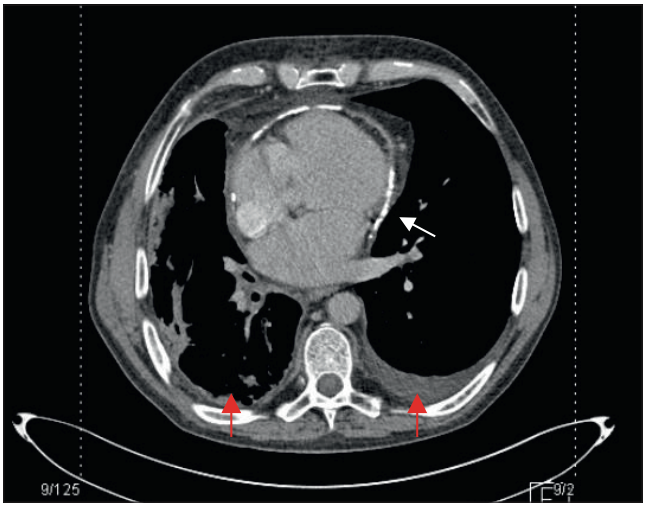

Figure 2. Computed tomography of the heart. Massive calcifications of pericardium(white arrows); hydrothorax of both pleural cavities (red arrows)

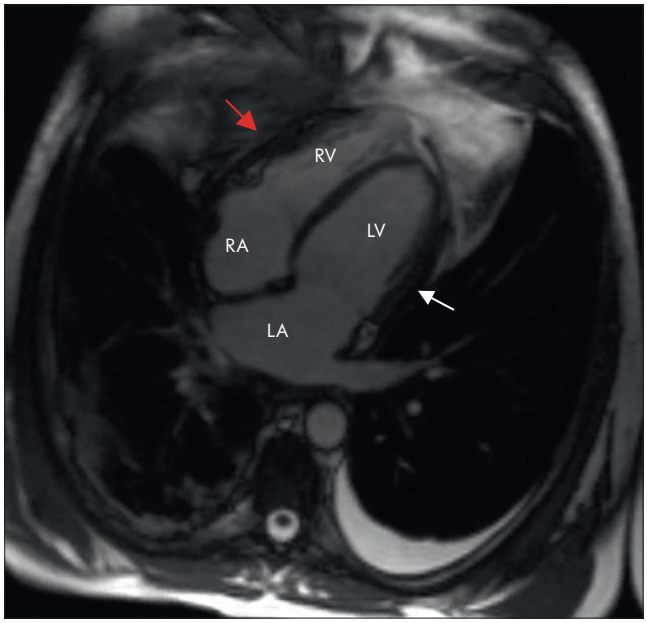

Figure 3. Cardiac magnetic resonance. Biatrial enlargement. Pericardium thickness is 5–6 mm over the right ventricle (red arrow), 8–9 mm over the left ventricle (white arrow). LV – left ventricle, RV – right ventricle, LA – left atrium, RA – right atrium

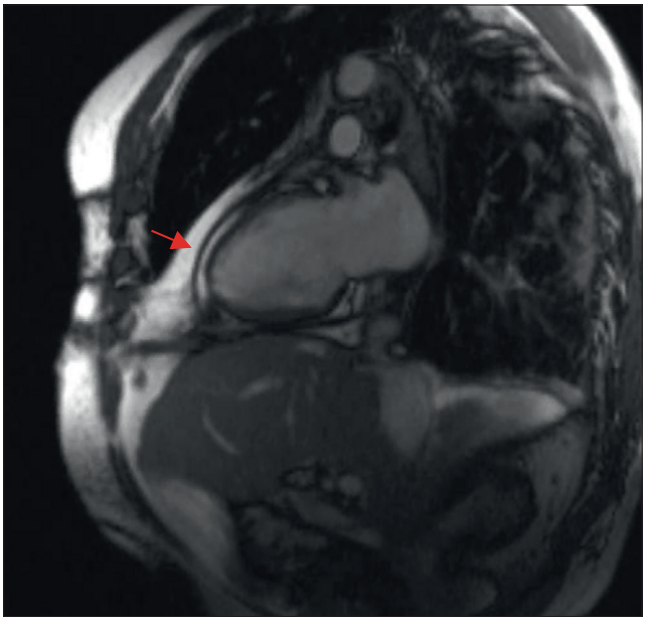

Figure 4. Cardiac magnetic resonance. Massive calcifications of pericardium (arrow)

type, was present. CT scan showed massive calcifications of pericardium, hydrothorax of both pleural cavities and irregular densities in mid and basal fields of the right lung (Figure 2). No signs of pulmonary embolism were present. Cardiac magnetic resonance (CMR) confirmed biatrial enlargement (left atrium area 33,5 cm^2, right atrium area 30,5 cm^2), normal size of both ventricles (LV ejection fraction 57%, end diastolic volume 133 ml, weight 93 g; RV ejection fraction 45%, end diastolic volume 139 ml) with dyssynchrony of intraventricular septum. Pericardium thickness was 5–6 mm over the RV, 8–9 mm over the LV, thickness of the walls of LV was: intraventricular septum 0,7 cm, posterior wall: 0,8 cm and RV wall thickness was 0,4 cm (Figure 3, 4).

Right heart catheterization showed increased end diastolic pressure in both ventricles, significantly increased pressure in venous system. Pressure in the pulmonary circulation was slightly elevated with normal pulmonary resistance. Cardiac index was 2,67 l/min/m^2 (Table 1). Evidence of constriction pericarditis were present – "square root" sign in the pressure tracking of the right ventricle (Figure 5). Angiography showed no stenosis of coronary vessels.

Patient was consulted with numerous specialists due to concomitant diseases. Eventually, he was qualified for optimal medical therapy. Surgical intervention was reserved only for lifesaving indications.

Table 1. Hemodynamic data

Pressure [mm Hg]	
Right atrium	27/24/22
Pulmonary artery	40/25/29
Right ventricle	43/12/22
Pulmonary capillary wedge pressure	33/30/27
Left ventricle	117/8/30
Aorta	102/65/79
Saturation [%]	
Inferior vena cava	78,5
Superior vena cava	75,3
Right atrium	76,1
Right ventricle	75,3
Pulmonary artery	75,5
Aorta	99,4
Cardiac Output [l/min]	5,8
Cardiac Index [l/min/m^2]	2,67
Qp/Qs	1:1
Vascular-Pulmonary Resistance [ARU]	28,2
Total-Pulmonary Resistance [ARU]	410
Vascular-Systemic Resistance [ARU]	785

Review of literature

Constrictive pericarditis is a rare pericardial disease in which the pericardial sac is losing its elasticity, cardiac filling becomes impaired by the stiffness of pericardial walls [1].

In developed countries most causes of constrictive pericarditis occurred after viral or idiopathic pericarditis (42–49%), also post-surgical and post-radiation pericarditis are common causes. Post-infectious constrictive pericarditis is rare nowadays (3–6%, including post-tuberculosis infection). Bacterial pericarditis are more prone to develop into constrictive pericarditis, this risk is intermediate in immune-related and neoplastic pericardial diseases and lowest in viral and idiopathic pericarditis [2].

Symptoms of constrictive pericarditis are related to impaired ventricular filling caused by the stiffness of pericardium. The symptoms are: fatigue, dyspnea, peripheral oedema, pleural effusion, venous congestion and ascites with hepatomegaly. Calcifications and thickening of pericardium are usually present in advanced stages, but up to 20% of the patients who suffer from constrictive pericarditis have normal thickness of pericardium. The diagnosis is based on clinical symptoms and imaging tests: transthoracic echocardiography and X-ray of the chest, than CMR and CT scan as second-level diagnostic techniques. Cardiac catheterization is performed when there are

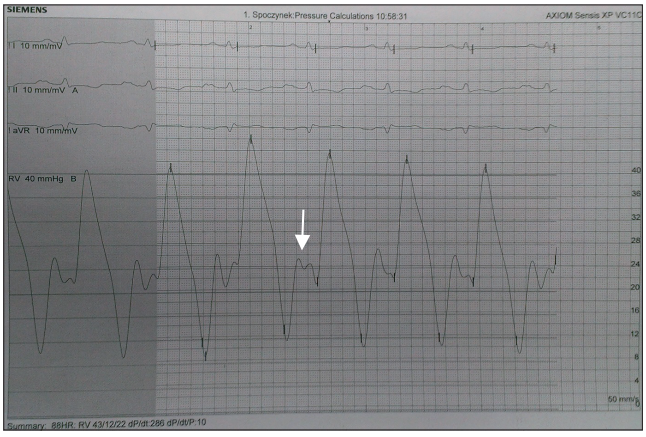

Figure 5. Right heart catheterization. Right ventricle pressure tracing. "Square root" sign (arrow)

difficulties with establishing diagnosis and in order to evaluate coronary arteries status before pericardiectomy [1,2].

In ECG there are no specific findings, atrial fibrillation and low voltage seemed to be present in part of the patients. Calcifications are typical sign in chest X-ray (in the shape of ring around the heart) however they are not present in majority of the patients [1].

Echocardiography shows thickening of the pericardium, intraventricular septum bounce, premature opening of the pulmonary valve, dilatation of inferior vena cava with no respiratory variation, biatrial enlargement, abnormal passive filling of the ventricles (high E velocity), reversed lateral/medial E' ratio of mitral annulus, variation of mitral and tricuspid flow during inspiration (mitral inflow falls >25% and tricuspid inflow increases >40%), respiratory variation in pulmonary venous flow [1,2].

CT scan shows calcifications and thickness of pericardium (>4 mm) however those symptoms aren't always present. Also dilatation of inferior vena cava, deformed ventricular contour and angulation of intraventricular septum could be visible. CT scan is also very feasible tool in pre-operative evaluation of the vascular structures [2].

CMR visualizes increased pericardial thickness, dilatation of inferior vena cava, septal bounce, moreover it can help to identify patients with inflammation background (CMR with late gadolinium enhancement (LGE) of the pericardium) and predict response to anti-inflammatory agents [1].

Cardiac catheterization helps to establish the status of coronary artery before the surgical procedure. Catheterization of the right heart shows increased pressure in the right atrium, Kussmaul's sign, increased end-diastolic RV pressure (usually to the level of 1/3 of RV systolic pressure), "square root" sign in diastolic RV and LV pressure tracings. LV and RV diastolic plateau pressure tracings are equal. There is a discordance between RV and LV pressure tracings – during inspiration the RV pressure is highest when LV pressure is the lowest [2].

Differential diagnosis focuses of exclusion of restrictive cardiomyopathy, cardiac tamponade and chronic liver diseases [1-3].

Pharmacology treatment should be considered in order to control the symptoms or in advanced cases, when the surgery is contraindicated. Diuretics should be administered to diminish venous pressure, oedema and ascites. Also if directed treatment is possible (p.e.

tuberculosis aethiology) pharmacology is advised. Patients with short history of constriction (p.e. temporary constriction during the course of pericarditis) may benefit from anti-inflammatory therapy [1,2].

In cases of chronic constrictive pericarditis the mainstay treatment is pericardiectomy (level of evidence IC). It is usually advised in patients in NYHA functional class III and IV, in mild and very severe cases it should be considered with much caution. Surgical mortality ranges between 6 to 12%. Completeness of the procedure is of great importance, as much of pericardium as possible should be removed. Also decortication of right atrium, superior vena cava, inferior vena cava and right ventricle adjacent to the diaphragm is necessary. Nevertheless in cases of severe calcinosis and adhesions radical pericardiectomy may be impossible and few islands of calcified pericardium should be left. Patients who undergo pericardiectomy should be referred to centers experienced in such procedures [1,2].

"End-stage" constrictive pericarditis is manifested by cachexia, atrial fibrillation, low cardiac output at rest (cardiac index ≤1.2 L/m^2 per min), hypoalbuminemia, hepatic failure caused by chronic congestion or cirrhosis. Patients with such severe disease are at greater surgical risk. Moreover pericardiectomy is less effective in this group. Worse overall survival is connected with prior radiation of mediastinum, impaired renal function, elevated pulmonary artery systolic pressure, abnormal left ventricular systolic function, lower serum sodium level and older age. Also patients with chronic liver disease (Child-Plug B or C [4]) have worse survival than group with milder dysfunction. Calcification of pericardium is not predictor of poorer survival rate [1,2].

Patient management and follow-up

Patient was initially treated with Bisoprolol 2,5 mg daily, Furosemide 40 mg 2-2-1, Spironolactone 50 mg daily, Atorvastatin 20 mg daily, Potassium supplementation and Pantoprazole 20mg daily. After several months there was no significant improvement in symptoms presentation.

Patient's case was discussed at the meeting of the Centre of Rare Cardiovascular Diseases and he was qualified for pericardiectomy. He was operated on January, 22, 2014. During procedure massive bleeding from right ventricle outflow tract occurred, despite blood transfusion and resuscitation procedures patient died in cardiogenic and hemorrhagic shock.

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