Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

This is a case of a 52 years old Caucasian man, who was referred to hospital with symptoms of heart failure (NYHA class II). He had a history of recurrent hospitalizations due to heart failure exacerbations. On physical examination his blood pressure was 110/80 mmHg, heart rate was 80 bpm. There were mute breath sounds over the base of the right lung, heart sounds were normal. He had oedema of both lower limbs, ascites, hepatomegaly and increased jugular vein pressure. He was treated with bisoprolol, furosemide, spironolactone, atorvastatin, pantoprazole, potassium and phospholipids supplementation.

On ECG sinus rhythm 80 bpm was observed with P mitrale, low voltage in limb leads and negative T wave in II, III, aVF, V1-V6. Laboratory tests revealed creatinine level of 92 umol/L, normal levels of electrolytes, INR 1,06, elevated LDL cholesterol, and NT-proBNP of 735 pg/mL. Serum albumin level – 26,5 g/L, serum protein level – 58,9 g/L. In serum proteinogram: high gamma globulin level – 17,9 g/L, high alfa1 and alfa2 globulin levels – 4,2% and 14,4% were detected respectively. In urine analysis Bence-Jones protein was negative. Liver function: total bilirubin – 12,4 umol/L, ALP 60 U/L, GGTP 49 U/L, AFP 1,83 ng/mL.

Chest X-ray showed right-side hydrothorax, densities in mid and basal fields of right lung and calcification of pericardium.

In echocardiography enlargement of both atria with normal size of ventricles was observed. Systolic function of left ventricle – ejection fraction (LVEF) was 55% with abnormal interventricular septum motion. Inferior vena cava (IVC) was widened (28mm) with restricted respiratory variation <50%. Other features of constriction: pseudonormal mitral flow, E/E'<8, respiratory variation of tricuspid and mitral flow >75%. Calcification of pericardium was visible during examination.

Ultrasonography of the abdomen showed ascites, hepatomegaly and dilated IVC.

In computed tomography (CT) irregular densities in mid and basal fields of right lung was detected with hydrothorax of both pleural cavities, massive calcification of pericardium.

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Angio-CT excluded pulmonary embolism.

Magnetic resonance imaging (MRI) showed: pericardium width of 5-6 mm over right ventricle (RV), 8-9mm over left ventricle (LV), normal size of both ventricles, aneurysmal dilatation of apical part of RV overlapping the apex of LV. LV EF 57%, EDV (end diastolic volume) 133 ml, mass 93 g, RV EF 45%, RV EDV 139 ml. Dyssynchrony of intraventricular septum's motion was observed as well as both atria enlargement. LV walls thickness was: 0,7 cm – intraventricular septum, and posterior wall - 0,8 cm, RV wall thickness was 0,4 cm.

In cardiac catheterization elevated end-diastolic pressure in both ventricles was observed, as well as elevated venous pressure. Slightly elevated pulmonary pressure with normal flow resistance was seen. In coronary catheterization no stenoses of coronary vessels was found.

Patient was consulted by specialists to estimate the risk of the procedure. Gastrological consultation: surgery only due to lifesaving indications. Pulmonological consultation: lifesaving surgery, open lung biopsy during cardiac surgery may be considered. Thoracic surgeon's consultation: the patient requires invasive pulmonological diagnostics or open biopsy. However, he does not give consent for invasive pulmonological diagnostics. He declares that he will continue pulmonary diagnostics after cardiologic evaluation is over.

DISCUSSION

Pericardium is a fibrous sac that surrounds the heart. It is inelastic, limits acute heart dilatation but can adapt to long-standing stress – like in left ventricle remodeling or accumulating of pericardial effusion without pericardial constriction.

Pericardial constriction is caused by thickened, usually calcified pericardium that impairs the filling of heart chambers and limits their volume.

Constrictive pericarditis is usually caused by long-lasting inflammation of pericardium, which leads to thickening, fibrosis and calcification. The most frequent causes are radiation of mediastinum, idiopathic pericarditis after cardiac surgery and tuberculosis.

Symptoms of constrictive pericarditis result from elevated systemic venous pressure and diminished cardiac output. Typical signs are: jugular venous distension, hepatic congestion, ascites, peripheral edema, poor exercise tolerance, even cachexia.

Echocardiography, computed tomography or cardiac magnetic resonance are basic modalities to establish the diagnosis. In differential diagnosing (restrictive cardiomyopathy), echocardiographic features and cardiac catheterization allows to distinguish constrictive pericarditis from cardiomyopathy.

The treatment of constrictive pericarditis depends on its onset and duration. In some patients with acute onset, pharmacology alone could resolve the constriction [1]. In chronic constrictive pericarditis surgery seems the best option. In ESC Guidelines on the Diagnosis and Management of Pericardial Diseases "pericardiectomy is recommended as the only treatment for permanent constriction. If severe calcified adhesions between peri- and epicardium or a general affection of the epicardium ("outer porcelain heart") are present surgery carries a high risk of either incomplete success or severe myocardial damage. An alternative approach in such cases may be a "laser shaving" using an Excimer laser. Areas of strong calcification or dense scaring may be left as islands to avoid major bleeding. Pericardiectomy for constrictive pericarditis has a mortality rate of 6%–12% [2]. Complete

normalization of cardiac hemodynamics is reported in about 60% of the patients.

EXPERT'S OPINION

Pericardiectomy is accepted standard of treatment in patients with chronic constrictive pericarditis who have persistent symptoms despite optimal medical therapy. This patient has been treated with diuretics for a longer time, however the symptoms persist and the diseases progresses. It has to be remembered, that while the majority of patients have a significant improvement in symptoms following pericardiectomy, there is a significant perioperative morbidity and mortality. Independent adverse predictors of long-term outcome included older age and worse NYHA class, as well as renal dysfunction, pulmonary hypertension, left ventricular dysfunction, and hyponatremia [1,3]. He has significant peripheral edema, and sings of enteropathy. On the other hand, he has no signs of cachexia, his left ventricular function is preserved, he has only mildly elevated pulmonary pressure and the renal function is not impaired. He seems therefore worth considering for surgical pericardiectomy.

Preoperative diuretics should be used sparingly with the goal of reducing elevated venous pressure, ascites, and edema. This approach can help to optimize the patient's hemodynamics prior to surgery and may improve their functional status.

Since the patient presents with additional pulmonological conditions, combined evaluation (cardiologists, cardiac surgeon, pulmonologists, thoracic surgeon and anesthetist) should be performed in order to reach final conclusion with regard to further surgical treatment of this patient.

CONCLUSION

The patient should be considered for surgical pericardiectomy. A cooperative evalatution, including cardiac surgeon, pulmonologists, thoracic surgeon and anesthetist should be performed in order to reach final conclusion with regard to further surgical treatment of this patient. The patient has to be informed with all pros and cons of every therapeutic option before he gives his consent.

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