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

A 71-year-old patient with history of permanent atrial fibrillation (on chronic acenocumarol therapy), arterial hypertension, type 2 diabetes mellitus was referred to Department of Cardiac and Vascular Diseases for cardiologic evaluation due to suspected pulmonary hypertension. Congenital heart disease had been suspected since infancy due to abnormal heart murmur. Since early childhood the patient experienced progressing impairment of physical capacity with dyspnea on exertion. Symptoms became more severe in the fourth decade of life. In August 2008 transthoracic echocardiography (TTE) revealed moderate mitral regurgitation (MR) with mild pulmonary and tricuspid regurgitations (PR, TR) and dilated main pulmonary artery (MPA), at that time left ventricle ejection fraction (LVEF) was normal.

Subsequent TTE examinations showed progression of PR and TR with elevation of right ventricle systolic pressure (RVSP) up to 80 mmHg. In September 2013 outcomes of pulmonological assessment suggested a possibility of pulmonary hypertension. Due to increased airway resistance in pletysmography ipratropium was introduced. On admission the patient complained of exertional dyspnea (NYHA class II/III), easy fatigue, recurrent peripheral edema and loss of appetite. Physical examination revealed systolic heart murmur (3/6 in Levine grading scale), symmetrical basal pulmonary crepitations and pitting edema of the lower extremities (grade 2). Laboratory tests showed non-therapeutic INR level (3,22 – acenocumarol therapy) and high NT-proBNP (4021 pg/ml). Chest X-ray disclosed pulmonary congestion, prominent pulmonary trunk and trace of pleural fluid. Abdominal ultrasound imaging showed mild ascites, hepatosplenomegaly, periportal fibrosis, dilated hepatic veins, and cholelithiasis Cardiopulmonary exercise test and 6-minute walk test confirmed significant impairment of exercise tolerance. Transthoracic echocardiography revealed enlargement of right heart chambers, probability of right ventricle systolic dysfunction (TAPSE 15 mm), moderate PR and MR, severe TR with considerably elevated RVSP (95 mmHg) and

retrograde systolic jet in the pulmonary artery. Angio-CT disclosed partially calcified persistent ductus arteriosus with left-to-right shunt. Right heart catheterization showed elevated pulmonary arterial pressure (68/24/39 mmHg) unresponsive to nirtoxide inhalation, blood pressure in the aorta was 184/44/95 mmHg.

Escalation of diuretic treatment resulted in regression of peripheral edema and pulmonary congestion, acenocumarol dosage was adjusted. The patient was evaluated by a cardiac surgeon and interventional cardiologist during a Heart Team consultation. She was found eligible for percutaneous device closure of PDA but did not consent to proposed treatment.

DISCUSSION

Patent ductus arteriosus (PDA) is a congenital heart disorder wherein fetal connection between pulmonary artery and the aorta fails to close after birth. As a result of increased blood oxygenation and drop of prostaglandin levels ductus arteriosus usually closes spontaneously within 48 hours after delivery. The incidence of PDA is approximately 1 in 2000 in full-term infants and consists 5 to 10% of all congenital heart diseases in children. PDA is found approximately twice more often in females. This heart defect is a rare finding in adults – due to abnormal heart murmur it is usually discovered in infancy. Mortality rate of adult patients with untreated PDA in is estimated to be 1.8% per year. Causing right ventricle overload and increased pulmonary blood low, PDA may eventually lead to Eisenmenger's syndrome.

Device closure is the method of choice in adult patients with persistent ductus arteriosus. Indications for PDA closure according to the current ESC guidelines are as follow: (1) PDA should be closed in patients with signs of LV volume overload; (2) PDA should be closed in patients with PAH but PAP <2/3 of systemic pressure or PVR <2/3 of SVR; (3) device closure is the method of choice where technically suitable.

EXPERT'S OPINION

Following the current ESC guidelines there are indications for PDA closure. In the case presented above there is no specific pharmacological treatment of pulmonary hypertension. Percutaneous device closure of PDA should be the preferable procedure.

CONCLUSION

It is recommended that the patient should undergoe percutaneous device closure of patent ductus arteriosus

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