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 75-year-old man with valvular pulmonary stenosis and concomitant clinically significant atherosclerosis and cardiovascular risk factors such as smoking, hypertension and hyperlipidemia. He was first hospitalized in 2009 due to chest pain Echocardiography revealed right ventricular hypertrophy, right atrium enlargement, dilatation of the pulmonary arteries, increased gradient in right ventricle outflow tract to 75/39 mmHg. MRI confirmed pulmonary valve stenosis with opening area of 0.5 cm². The coronary angiography showed significant stenosis of the right coronary artery. Right heart catheterization showed significantly increased systolic pressure in right ventricle- 107 mmHg, the pressure in pulmonary artery was of 40/9/25 mmHg. After coronary angioplasty of the right coronary the symptoms disappeared.

In August, 2013 the patient was admitted to hospital due to decreased exercise capacity and relapse of chest pain on exertion. Echocardiography showed right ventricle hypertrophy (free wall thickness of 7 mm), valvular pulmonary stenosis with the gradient of 74/47 mmHg, right atrium enlargement (23 cm²); tricuspid annular plane systolic excursion (TAPSE) was slightly decreased (14mm). In cardiopulmonary exercise test maximal oxygen consumption was of 27ml/kg/min. Test was abrupted due to severe fatigue at 74% of predicted heart rate, 7.4 METs, no significant ST-T changes. Coronary angiography showed 85% stenosis in RPD, and stent was implanted.

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DISCUSSION

The authors of this case asked whether optimal pharmacological therapy should be supplemented by pulmonary valvotomy in this patient. Valvular pulmonary stenosis is the most common type of right ventricular outflow tract obstruction (80-90%). It is usually isolated lesion and accounts for 7% - 12% of congenital heart diseases. It can be associated with abnormalities of the structure of pulmonary artery wall which can lead to pulmonary artery dilatation. Usually pulmonary trunk and left pulmonary artery are affected.

Patients with mild or moderate stenosis are usually asymptomatic. Mild stenosis is usually stable over time. In patients with moderately stenosed pulmonary valve the gradient can rise in time due to calcification of the leaflets and reactive subpulmonary hypertrophy of the right ventricular muscle. Patients with severe stenosis usually complain if dyspnea and exercise intolerance. Typical sings in physical examination are harsh systolic murmur over stenotic segment and wide splitting of the second heart sound. Echocardiography shows the level of stenosis, anatomy of the pulmonary valve, right ventricular hypertrophy and other concomitant abnormalities. It is also used to assess right ventricular systolic pressure. Based on the echocardiographic measurement of maximal systolic gradient through the valve the stenosis is classified as: mild: <3 m/s (36 mmHg), moderate: 3-4 m/s (36-64 mmHg), severe: >4 m/s (>64 mmHg). These measurements are frequently inaccurate, therefore usually right ventricular systolic pressure is used to assess the severity of the disease.

The treatment of choice in severe (gradient >64 mmHg in patients with normal right ventricular function) stenosis of pulmonary valve is baloon valvoplasty irrespectively of symptoms. If this method is not effective and right ventricular systolic pressure is >80 mmHg surgical replacement of pulmonary valve is required. In patients with gradient through the pulmonary valve <64 mmHg interventional treatment should be considered when there is right ventricular systolic dysfunction, the patient is symptomatic or significant arrhytmias occur.

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EXPERT'S OPINION

This patient does not have symptoms of pulmonary artery stenosis but has recurrent symptoms of coronary artery disease which deserves typical treatment. The contractility of the right ventricle is depressed and the stenosis is severe which makes the patient a candidate for interventional procedure.

CONCLUSION

The patient requires interventional treatment of the severe pulmonary valve stenosis. Based on the anatomy of the valve the type of interventional treatment should be chosen: baloon angioplasty (generally prefered) or surgical pulmonary valve replacement (dysplastic valve). Probably this intervention should be preceded by stenting of the right carotid artery.

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