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

44 y.o. male with positive family history of acute coronary syndromes and coronary artery disease risk factors including hypertension and obesity was referred for cardiac evaluation due to newly discovered, recurrent left bundle branch block (LBBB). First diagnosis was made in November 2012 during routine cardiac evaluation. Since few months the patient complains of fatique and low exercise tolerance. Until that time he had no problems with the cardiovascular system. First echocardiography showed hypoechogenic mass in the left ventricle of 3 over 2.5 cm. He was treated with anticoagulation therapy before admission and no thrombus was noticed in control echocardiography after one month. Patient was admitted for further evaluation. Echocardiography revealed moderate mitral regurgitation (II/III degree), hypokinesis of the apex and intraventricular septum, LVH (6.5/4.9cm), IVS 11mm, LA 4.5cm, RV 2.8cm and LVEF 48%. LBBB was present in the EKG. After dobutamine administration significant improvement in intraventricular septum contractility and mild improvement in apical contractility was noticed, IM II/III degree, EF = 45% but EKG converted to sinus rythm with 45-50/beats per min. and ST segment depression in V2-V6. Blood pressure at this time of examination was 190/110 mmHg. No chest pain occured during this examination. Coronarography revealed normal right and left anterior descending coronary arteries without atherosclerosis but significantly reduced distal perfusion at the level of TIMI grade II. Circumflex annomally was visualized with severe enlargement of the artery (diameter ca. 9mm) and shunt towards the coronary sinus and finally the right atrium During the invasive procedure right heart catheterisation was performed. In 3D Angio-CT reconstruction images the circumflex artery was enlarged throughout the whole lenght with proximal diameter of 8 x 9 mm and distal diameter of 9 x 10 mm, two marginal branches originating from it. It connects to the coronary sinus (arterial-venous fistula ca. 25 mm before entering the right atrium

DISCUSSION

Coronary A-V fistulas present as rare congenital anomalies in adult patients and are found in 0,2-2.1% of angiograms. Clinical presentation varies from asymptomatic to life-threating events including syncope, cardiogenic shock or sudden cardiac death. Therapeutic approach was mostly surgical (46% in children vs. 38% in adults), conservative or in few cases percutaneous (18% in children and 5% of adult patients). Usage of antibiotics and antiplatelet and/or anticoagulant therapy is recommended in most patients.

EXPERT'S OPINION

The patient is young with anomalous circumflex artery shunt to the right atrium, that causes LV ischaemia during dobutamine stress test, LV dilatation, hypokinesia of inertventricular septum and LV apex and diminished LV systolic function. Surgery – closure of the shunt is the choice of treatment. Mitral valve reconstructive surgery could be performed simultanously, but more detail echocardiographic evaluation of the mechanism of MR must be performed before surgery. In my opinion there are no indications for MitralClip - when surgical treatment is planned in this case. Anticoagulation should be administered, as thrombus was diagnosed in the cavity of LV.

CONCLUSION

Surgical treatment – closure of the shunt and reconstruction of MV should be proposed for the patient together with anticoagulation therapy.

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