Giant coronary sinus secondary to partial anomalous pulmonary venous connection with combined mitral valve disease

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Partial anomalous pulmonary vein connection (PAPVC) is a rare congenital abnormal cardiac defect involving the pulmonary veins draining into the right atrium (RA) directly or indirectly by venous connection. Nowadays, these patients are diagnosed when still in childhood and operated on with excellent results [1]. The drainage directly to the coronary sinus accounts for 3% of patients with PAPVC, and only a few such cases with coexisting mitral valve disease are described in the literature [2, 3]. Partial anomalous pulmonary vein connection most commonly presents with an atrial septal defect (ASD), reportedly in 80–90% of cases. Of these, 85% are reportedly sinus venosus type, while 10–15% are secundum type [4, 5].

A 55-year-old woman with mitral valve disease of rheumatic etiology, atrial fibrillation, and previous ischemic stroke was admitted to the department of cardiology due to shortness of breath, decreased exercise tolerance, and fatigue. The symptoms of cardiac failure had been exacerbating for 6 months. That was the first hospitalization of the patient due to exacerbation of the symptoms of heart failure. Previously, the patient had been treated in the outpatient clinic only with diuretics, β-blockers and oral anticoagulation therapy (OAT). Her heart failure therapy was intensified with β-blocker, ACE inhibitor and diuretics, but she gradually deteriorated to NYHA class III. Due to atrial fibrillation (AF) the medical therapy included OAT which was ineffective (INR = 1.65). Upon physical examination, elevated jugular venous pressure (JVP), loud S1 tone and split S2 tone with a slight murmur at the left upper sternal border, swollen ankles and shortness of breath were present. In ECG AF with right heart bundle branch block was present. Transthoracic echocardiogram (TTE) revealed enlargement of the right ventricle (37 mm) and right atrium (50 mm), severe tricuspid valve regurgitation, combined degenerative mitral valve disease with moderate stenosis (planimetric area 1.0 cm^2) and regurgitation (Figure 1 C). Pulmonary hypertension was diagnosed with systolic pulmonary artery pressure of 55 mm Hg and a pulmonary to systemic flow ratio of 1 : 2.1. Additionally, enlargement of the coronary sinus (38 × 15 mm in parasternal long-axis view) (Figures 1 A, B) was noted. On transesophageal echocardiography (TEE) a large thrombus protruding from the left atrial appendage (LAA) was found, as well as patent foramen ovale (PFO) with minute left-to-right shunting and inflow of the left pulmonary veins; however, typical venous return of both right pulmonary veins was absent (Figure 1 D). Cor-

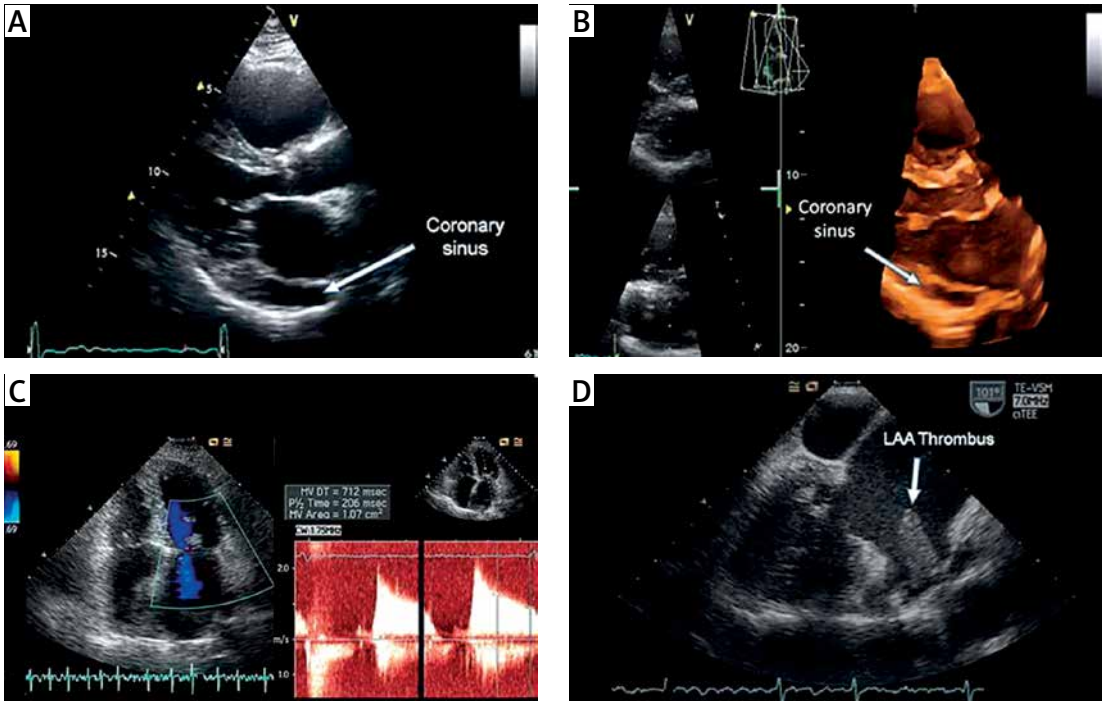

Figure 1. A – Transthoracic echocardiography, parasternal long-axis view – dilated coronary sinus. **B** – Transthoracic three-dimensional echocardiography, parasternal long-axis view – dilated coronary sinus. **C** – Transthoracic echocardiography with pulsed wave Doppler and color Doppler flow mapping indicating stenosis of the mitral valve leaflets and moderate regurgitation. **D** – Transesophageal echocardiography: thrombus protruding from the left atrium appendage

onary angiography showed no significant lesions, although in the pulmonary angiogram anomalous drainage from the left pulmonary veins was noted (Figure 2). Computed tomography (64-row multislice) revealed anomalous left upper and lower pulmonary vein drainage into the coronary sinus and further to the right atrium (Figure 3). Subsequent cardiac surgery was performed and involved mitral valve replacement with a mechanical prosthesis (Medtronic 27), tricuspid valve annuloplasty and removal of the thrombus from the appendage. However, pulmonary venous return was not corrected. The decision not to perform repair of the PAPVC was made by a surgeon due to the mild clinical symptoms of the disease. Follow-up TTE indicated proper function of the artificial mitral valve, moderate tricuspid valve regurgitation and persistent PFO with an enlarged coronary sinus.

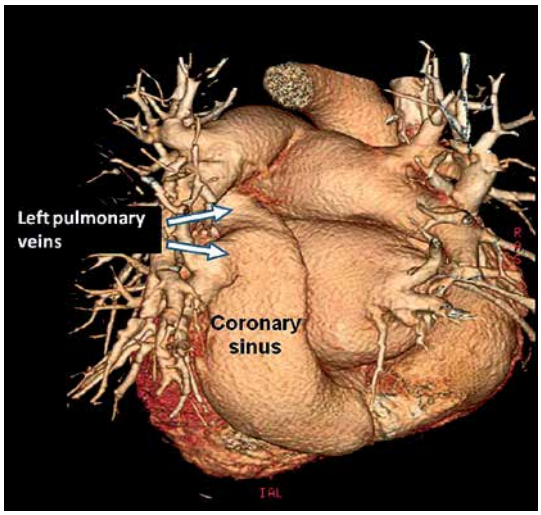

Figure 2. Volume rendered CT image of the heart showing anomalous left upper and lower pulmonary vein connections with coronary sinus drainage into the right atrium

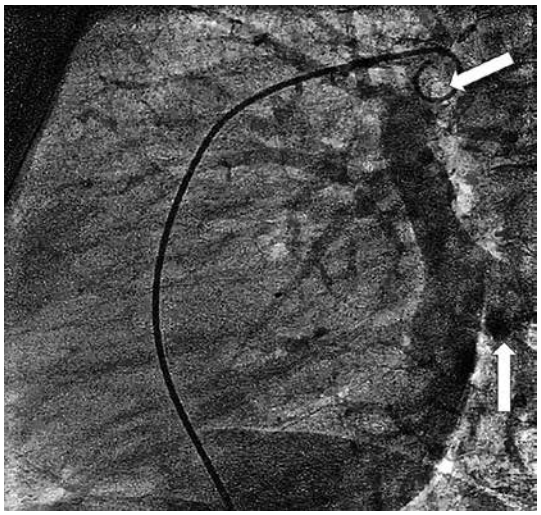

Figure 3. Cardiac catheterization: anterio-posterior view – arrows indicating anomalous left upper and lower pulmonary vein drainage into coronary sinus

After surgery the patient improved from the third to the second functional NYHA class.

Our patient represents a rare combination of structural heart disease – PAPVC and combined mitral valve disease. The treatment strategy of congenital heart defects diagnosed in adults is in the majority of cases a challenging clinical dilemma. Partial anomalous pulmonary venous connection accounts for about 0.5% of congenital heart disease, usually with coexisting sinus venosus ASD, but is very uncommon as an isolated lesion. More than 90% of all PAPVCs originate from the right lung, 7% originate from the left lung, and 3% of patients are found to have bilateral PAPVCs originating from both lungs connecting to either the superior vena cava (SVC), the inferior vena cava (IVC), the right atrium or the innominate vein [6]. Symptoms are mainly dependent on the severity of associated shunt, and complications. The associated shunt can cause a high output state leading to volume overload and heart failure. Echocardiography and especially TEE is the initial method of choice for the noninvasive detection of PAPVC [2, 5]. It should be remembered that compression of the left atrium by the enlarged coronary sinus may occur, mimicking mitral stenosis [7]. Left to right shunt may result in significant volume overload, which can lead to progressive dilatation of both tricuspid and mitral valve annuli resulting in severe mitral and tricuspid regurgitation. Surgical repair of PAPVC is necessary when patients exhibit symptoms or demonstrate right heart failure or pulmonary hypertension [4]. The traditional surgical indication for repair of a left-to-right shunt at the atrial level is a pulmonary-to-systemic blood flow ratio > 1.5 [6]. El Bardissi *et al.* suggested that surgical repair should take place early in patients with RV dilation or mild to moderate tricuspid regurgitation, or at early stages of pulmonary vascular disease, to prevent pulmonary hypertension [6].

Due to severe tricuspid regurgitation and mitral stenosis, our patient underwent mitral valve replacement with tricuspid valve annuloplasty, according to current recommendations [8]. The decision to abandon venous correction was taken at the discretion of the cardiac surgeon due to episodic hemodynamic instability of the patient during surgery which results in our view in a suboptimal outcome. However, despite unoperated PAPVC the patient has improved and remains stable in second NYHA class without symptoms of pulmonary hypertension 3 years after surgery, and he refuses the proposed reoperation.

Adult patients with rare congenital heart defects often require an individualized clinical approach. Coexistence of a rare congenital anomaly, acquired mitral valve disease and small ASD illustrates the need for scrutiny when diagnosing adults with pulmonary hypertension.

Conflict of interest

The authors declare no conflict of interest.

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