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Coronary sinus atrial septal defect in adult (RCD code: IV-2B.1)

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Abstract

We present a case report of a 45-year old man, who was admitted to medical center due to dyspnea and cough. Right bundle branch block was detected on electrocardiography. Transthoracic echocardiography demonstrated coronary sinus dilatation, enlargement of right-side chambers, moderate to severe pulmonary hypertension and atrial septal defect with left-to-right shunt. Coronary sinus atrial septal defect was diagnosed by transesophageal echocardiography and computed tomography. Surgical repair of the defect was performed. Atrial flutter occurred in early postoperative period and was successfully treated by electrostimulation. Coronary sinus atrial septal defect in adults is a rare congenital heart disease. Transesophageal echocardiography and computed tomography are imaging modalities of choice in such cases. Surgical treatment is needed to prevent further complications. JRCD 2017; 3 (3): 86–88

Key words: rare disease, atrial septal defect, coronary sinus, echocardiography, computed tomography

Background

Patients with arterial septal defects (ASDs) frequently remain asymptomatic until adulthood [1]. However, the majority of symptoms develop beyond the fourth decade including reduced functional capacity, exertional shortness of breath, palpitations (supraventricular tachyarrhythmias) and less frequently pulmonary infections, right heart failure.

Case presentation

A 45-year old man was admitted in September 2014 to St. Paraskeva Medical Center due to dyspnea and cough. The symptoms gradually progressed to functional class II by New York Heart Association (NYHA). Physical examination revealed body mass index of 33 kg/m^2, regular pulse of 80 bpm, blood pressure of 135/100 mmHg on both upper limbs, accentuated second heart sound with systolic murmur over pulmonary artery point, vesicular breathing sounds, and no peripheral edema. He had a history of chronic bronchitis. He denied alcohol consumption, smoking, family history of cardio-vascular diseases. Electrocardiogram (ECG) revealed sinus rhythm of 80 bpm, right bundle branch block. Chest X-ray showed pulmonary venous congestion. Transthoracic echocardiography (TTE) demonstrated coronary sinus (CS) dilatation, enlargement of right ventricle (4,1 cm) and right atrium (4,7 cm minor dimension), moderate tricuspid regurgitation with right ventricular systolic pressure (RVSP) of 55 mmHg (high probability of pulmonary hypertension) and ASD with left-to-right shunt [2]. CS type ASD was suspected. Transesophageal echocardiography (TEE) showed large CS defect with left-to-right shunt, aneurysmal dilatation of CS probably associated to a persistent left superior vena cava (Figure 1). Bubble contrast study via left antecubital vein for diagnosis of persistent superior vena cava was not performed.

Coronary computed tomography angiography showed normal coronary arteries without calcifications (Figure 2 A).The large unroofed CS type ASD with CS aneurism was confirmed (Figure 2 B, C). The study also detected a common truncus of the left pulmonary veins draining into the left atrium (Figure 2 D).

Surgical treatment was recommended for CS defect with clinically significant left-to-right shunt associated with cardiomegaly and symptoms. Surgical repair was performed in December 2014. Postoperative TTE revealed decrease in right ventricular size (3,0 cm), right atrial size (4,3 cm minor dimension) and RVSP to 35 mmHg. Dyspnea and cough were reduced. The patient was admitted again in February 2015 due to palpitation. ECG revealed atrial flutter with 2:1 conduction and ventricular rate of 128 bpm. Transesopha-

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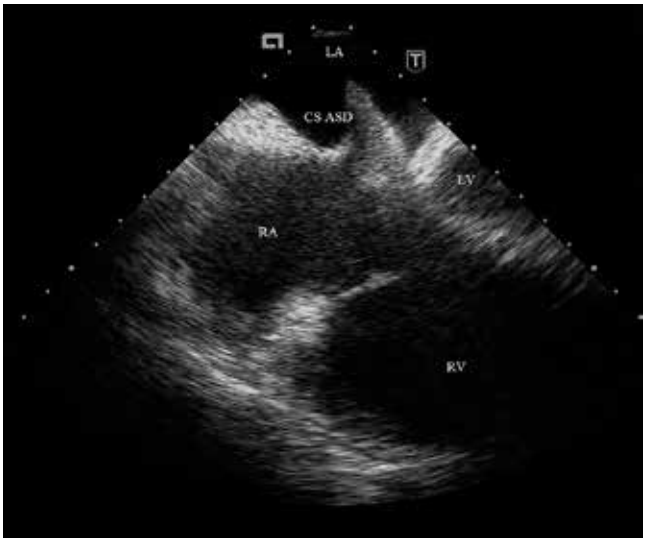
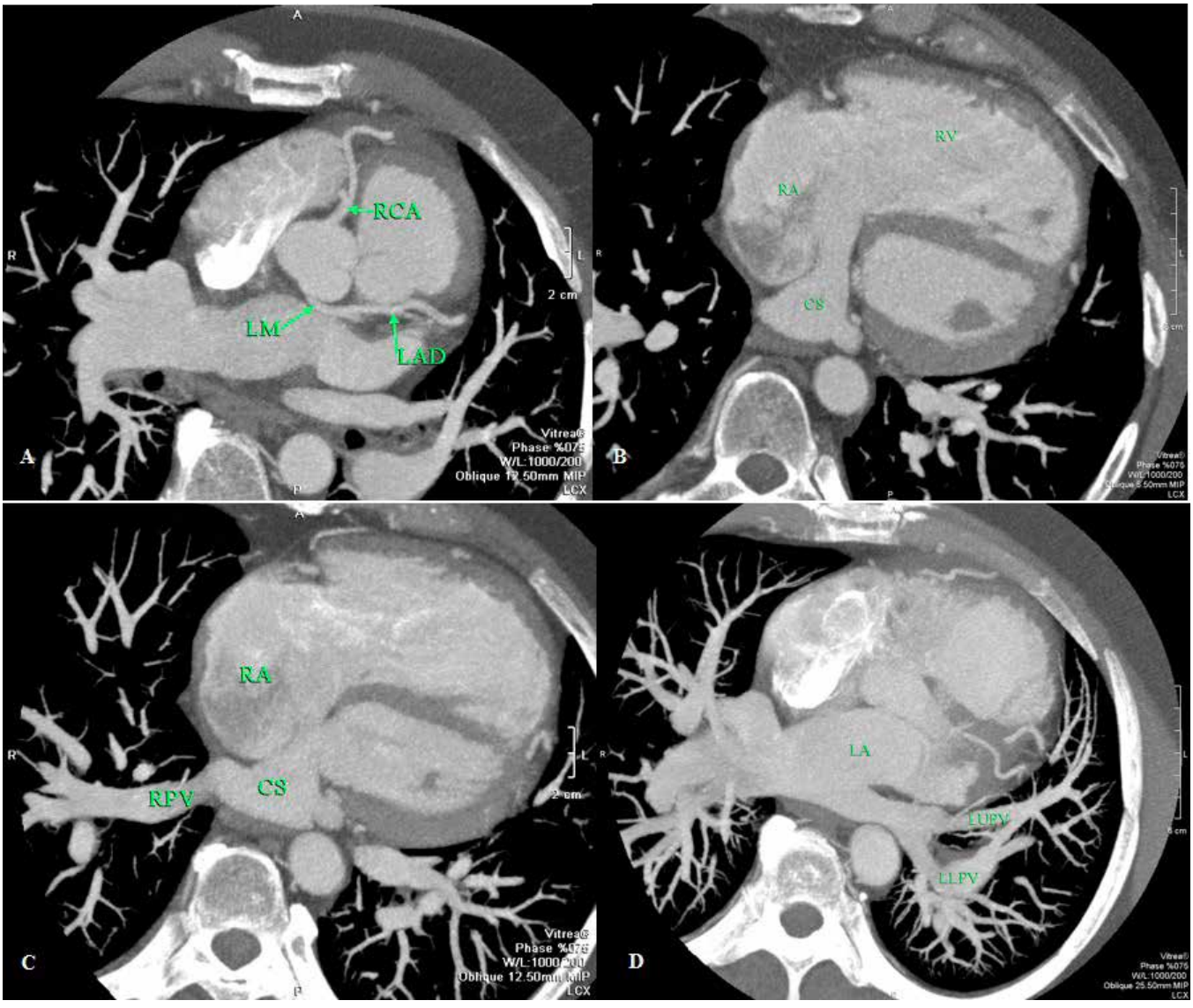


Figure 1. Transesophageal echocardiography. LA – left atrium, RA – right atrium, LV – left ventricle, RV – right ventricle, CS ASD – coronary sinus atrial septal defect

Figure 2. Cardiac computed tomography angiography. A. Normal coronary arteries without calcification. LM – left main coronary artery, LAD – left anterior descendent artery, RCA – right coronary artery. B. Dilated coronary sinus with aneurism. CS – coronary sinus, RA – right atrium, RV – right ventricle. C. Dilated coronary sinus connection with left atrium and right pulmonary vein inflow. RPV – right pulmonary vein. D. Common truncus of the left pulmonary veins which drain into left atrium. LA – left atrium, LUPV – left upper pulmonary vein, LLPV – left lower pulmonary vein



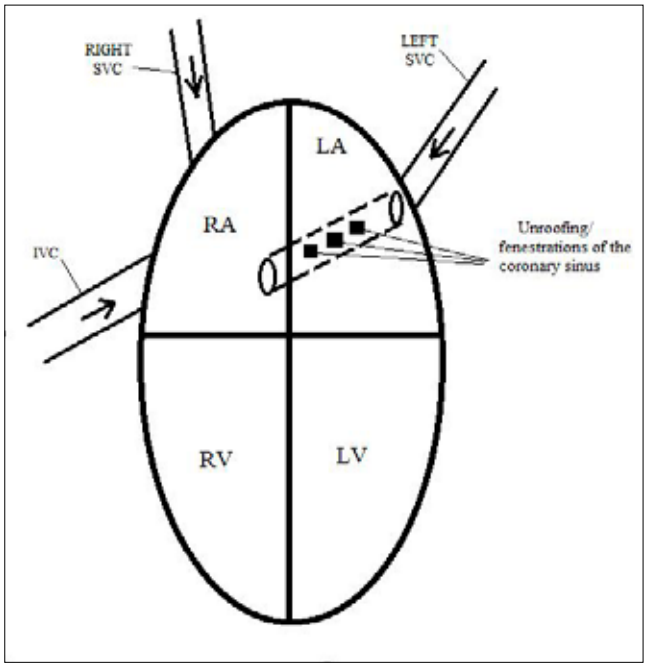


Figure 3. Schematic drawing of an unroofed coronary sinus. SVC – superior vena cava, IVC – inferior vena cava, LA – left atrium, RA – right atrium, RV – right ventricle, LV – left ventricle

geal electrostimulation was performed, sinus rhythm was restored. At present the patient receives treatment for systemic arterial hypertension (combination valsartan 160 mg + hydrochlorothiazide 12,5 mg) and aspirin 100 mg daily. He isn't complaining of cough and dyspnea.

Discussion

Isolated CS defect is exceedingly rare and occurs less than 1% of all ASDs [1]. Defect is characterized by a deficiency in the tissue separating the CS from the left atrium.This results in partial or complete unroofing of the CS leading to a predominantly left-to-right shunt through the CS (left atrium to CS to right atrium) (Figure 3).

The orifice of the ostium is frequently large because of the increased flow. From the right atrium side, the defect is located at the level of the CS ostium and may also include some deficiency in atrial tissue around the ostium. From the left atrium side, the size can be variable depending on the degree of unroofing of the CS [3]. Unroofed CS is associated with other congenital heart diseases, of which persistent left superior vena cava is one [6].

Although TTE has developed as one of the main assessment tools, the diagnosis of unroofed CS remains tricky and unsatisfactory, especially in patients with large body habitus who might have poor echocardiographic windows. TEE commonly provides better visualization of cardiac structures and is superior in demonstrating shunts. However, it is an invasive procedure that requires premedication and/or sedation in some patients [6]. Non-invasive computed tomography is an excellent high-resolution imaging to accurately diagnose the anatomy in rare ASDs [4].

Surgical repair has low mortality (<1% in patients without significant co-morbidity) and good long-term outcome [1]. Atrial tachyarrhythmias occurring early after intervention are mostly transient [5]. In patients of advanced age with ASDs, individual surgical risk due to co-morbidities must be carefully weighed against the potential benefits of ASD closure [1].

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