Infected endocarditis of partial atrioventricular septal defect – A case report

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Abstract

Introduction. Partial atrioventricular septal defect (AVSD) is a form of congenital heart disease (CHD) rarely detected in adults. Infective endocarditis represents a severe complication that carries a substantial risk. Case report. We here reported a case of a 43-year-old female with previously diagnosed adult CHD (partial AVSD and bicuspid aortic valve) presented to the hospital with fever and malaise 14 days prior to admission. On the lung computed tomography scan inflammatory consolidations were found and dual antibiotic therapy (ceftazidime and clarithromycin) was administered without significant regression of pulmonary inflammatory consolidations. The antibiotic treatment was continued with amoxicillin/clavulanic acid combined with levofloxacin and metronidazole. Transthoracic and transesophageal echocardiography revealed a large vegetation (dimension, 3.6 x 1.8 cm) attached to the septal side of the tricuspid valve with high embolic potential. Endocarditis team reached a decision for immediate surgical intervention. The operative findings revealed the partial AVSD, common atrioventricular valve with cleft of the anterior mitral leaflet in the A2 segment and detached and cleft septal leaflet of the tricuspid valve. Vegetation (size 4 x 3 cm) was attached to the septal side of the tricuspid annulus, basal segment of the anterior mitral leaflet and edge of the atrial septal defect freely floating between right atrium, right ventricle and left atrium. Excision of the vegetation and AVSD plastics were done, as well as the reconstruction of the mitral and tricuspid annuli and leaflets. The treatment was continued with antibiotics and completed in 18 days with full recovery. Conclusion. Early and precious diagnosis and optimal management that combines both conventional and surgical approaches are crucial for reducing the risk of complications and mortality in patients with infective endocarditis in grown-up congenital heart disease.

Key words: endocarditis, bacterial; atrioventricular septal defect; tricuspid valve; diagnosis; cardiovascular surgical procedures; anti-bacterial agents; treatment outcome.

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Introduction

Infective endocarditis (IE) in adults with congenital heart disease (CHD) has been increasing along with the increasing number of patients with CHD who were reaching adulthood 1–3 as grown-up congenital heart disease (unrepaired, repaired, palliated) 4. An underlying CHD is found in 11% to 13% of all IE 5. Despite latest advances in diagnosis and treatment (conventional or surgical management), mortality and the rate of complications are high 6 and late course is unpredictable 5.

The term atrioventricular septal defect (AVSD) covers a spectrum of heart anomalies with a common atrioventricular (AV) junction. With an incidence of 4–5.3 per 10,000 live births, AVSD comprises 7% of all CHD. Controversies exist on nomenclature and subdivision of the varying morphology of AVSDs, and several different descriptions are currently used. There are three types of AVSD: 1) Complete AVSD includes an ostium primum atrial septal defect and inlet ventricular septal defect (VSD). Clefts in the mitral and tricuspid valve leaflets result in one common, large AV valve connecting the atrial and ventricular chambers; 2) Partial AVSD includes an ostium primum AVSD with separate mitral and tricuspid valve orifices and clefts in the mitral and/or tricuspid valve leaflets. Partial AV canal occurs in 1-2% of all congenital heart defects; 3) The term intermediate AVSD (also called transitional) is variably defined and is an infrequent form of AVSD. Two-orifice AV valve with a single valve annulus is usually present and often restrictive VSD just below AV valves 7.

Case report

A 43-year-old woman was admitted to a secondary health care hospital with a history of fever up to 39°C. Two weeks prior to admission the patient complained of malaise, poor appetite, cough and pain at the right side of chest. Antibiotic therapy with ciprofloxacin was started 10 days prior to admission, but she remained febrile. The patient was diagnosed grown-up CHD in the age of 38. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TOE) showed AVSD as a combination of ostium primum AVSD (diameter of atrial septal defect was 14 mm), with left to right shunt (LR shunt), cleft of the anterior mitral valve leaflet (AML) with mild mitral regurgitation as well as cleft of the tricuspid septal leaflet. Also, the bicuspid aortic valve was present without stenosis and regurgitation. The right chamber was normal in size with thickened wall (0.7 cm) and preserved systolic function. The left ventricle was normal in size and function. Contrast echocardiography with agitated saline contrast showed negative contrast effect. Diagnostic cardiac catheterization demonstrated a moderately significant left-to-right shunt Qp:Qs of 1.2 and confirmed the diagnosis of partial AVSD, without significantly elevated pulmonary artery pressure (PA 36/16/24 mmHg). The patient refused surgical correction that was proposed. She had history of arterial hypertension and type 2 diabetes.

Physical examination during admission to the regional hospital revealed systolic heart murmur audible on mitral area. Laboratory tests revealed leukocytosis (white blood cells count 12.0 × 10^9/L) and raised inflammatory markers (C-reactive protein 41 mg/L). Chest X-ray and thoracic computed tomography (CT) scan showed inflammatory consolidations in left middle and lower parts and right lower parts of the lung with accompanied mediastinal lymphadenomegaly. The patient was treated with dual antibiotic therapy (cefazidine and clarithromycin) for 14 days without significant regression of pulmonary inflammatory consolidations. Repeated blood cultures remained sterile. TTE revealed tricuspid valve endocarditis. The antibiotic treatment was continued with amoxicillin/clavulanic acid combined with levofloxacin and metronidazol for 8 days. Then, the patient was referred to a tertiary health center, for TOE and further diagnostic testing and treatment. The antibiotic treatment was continued with amoxicillin/clavulanic acid and levofloxacin. TOE showed large vegetation (3.6 x 1.8 cm) attached to the septal leaflet of the tricuspid valve floating between the right atrium and right ventricle through the tricuspid valve with mild tricuspid regurgitation (Figure 1).

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**Fig. 1 – Transesophageal echocardiography of four chambers shows atrial septal defect (ASD), type primum. Mitral and tricuspid valve orifices are in the same level. Vegetation is connected to the septal leaflet of the tricuspid valve floating between right atrium (RA) and right ventricle (RV) through the tricuspid valve.**

LA – left atrium; LV – left ventricle.
The embolic potential of vegetation was high. Thoracic CT scan with pulmonary angiography revealed bilateral segmental and subsegmental septic pulmonary emboli. Endocarditis team reached a decision for immediate surgical intervention. The operative findings revealed the AVSD type primum (dimension 4 x 4 cm), cleft of the anterior mitral leaflet in the A2 segment and cleft septal leaflet of the tricuspid valve. There was direct communication between the left ventricle and the right atrium through AML and ostium primum. Vegetation size was 4 x 3 cm and it was attached to the septal leaflet of the tricuspid valve, basal segment of the AML and edge of the primum AVSD freely floating between the right atrium, right ventricle and left atrium. Excision of vegetation, AVSD plastics with Xeno Sure Biologic patch, as well as the reconstruction of the anterior mitral and septal tricuspid cusps with mitral and tricuspid annuloplasty were done (Figure 2). The intervention was performed without any complications. TTE was repeated following the procedure, and there was no remaining AVSD, mitral and tricuspid regurgitation and no signs of infective endocarditis. After the surgery, antibiotic therapy was continued with amoxicillin/clavulanic acid in combination with levofloxacin and metronidazole for 14 days. The patient was discharged from the hospital in good general condition with no signs of inflammation with recommendations for oral therapy with moxifloxacin and clavulanic acid. After 18 days, full recovery was achieved. The patient was doing well at 3 months follow-up.

**Discussion**

We present herein a rare case of CHD in adulthood followed by IE. To our knowledge no such case of IE is yet described in literature. In large cohort study on 84,308 adult patients admitted to hospitals with CHD Rodriguez et al. found that only 0.4% had AVSD. Partial AVSD is often discovered in early childhood, but sometimes it may go undetected with no symptoms up to adulthood as in the case of our patient whose symptoms presented for the first time at the age of 38. Somerville has reported that patients with partial AVSDs face symptoms with increasing frequency after the third decade of life. Gatzoulis et al. have reported that three quarters of patients with grown-up congenital heart disease who underwent primary repair of partial AVSD are asymptomatic at mean age 37.7 years like our patient.

AVSD is typically present in the neonatal period being an important cause of cardiac morbidity and mortality in this age group. Mortality is higher in children with a complex AVSD and in those with 2 or more major noncardiac malformations. Children with partial and transitional AVSD are mostly asymptomatic so recommendation for surgical repair is might be delayed to preschool or older ages. Surgery for partial AVSDs with large left-to-right atrial shunts in adults can be performed with low mortality and morbidity. On the other hand, in operated or medically treated patients, the long-standing course is unpredictable. The risk of developing IE in adult CHD is more than 10 times increased than in normal population. Infective endocarditis might be late complication that carries substantial mortality risk. Recently, Tutarel et al. studied IE in adults with CHD and found that during follow-up of 6.7 years, 19.4% died. High mortality rate highlights the need for consciousness of IE and adult CHD. The majority of studies about IE in adult CHD are retrospective and limited in number so valid conclusions cannot always be made. There is an evident need for future long-term follow-up studies on this disease. Awareness and understanding of the disease and its complications are essential in order to determine an early treatment and prevent complications.

**Conclusion**

This is extremely rare case of partial AVSD, uncommonly seen in adulthood, complicated with infective endocarditis. Early diagnosis and optimal management that combines both conventional and surgical approaches are crucial for reducing the high embolic risk, risk of complications and mortality. For obtaining the best results, complicated forms of infective endocarditis with underlying congenital heart disease should be evaluated and treated at early phase in tertiary surgical centers by an experienced endocarditis team.

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