

Case report

Partial atrioventricular septal defect in the elderly: A case report

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Atrial septal defect is one of the most common congenital heart lesions. Isolated atrial septal defects are mostly asymptomatic, and remain therefore undiagnosed until adulthood in a significant number of patients. Partial atrioventricular septal defect, on the other hand, has a life expectancy of 3 to 37.5 years, but occasionally patients may present in their adulthood. We report a case of an 83-year-old female with partial atrioventricular septal defect who had syncope, near-syncope and palpitations for the last three years and was diagnosed by routine echocardiography. Because atrial septal defect has long-standing deleterious effects of volume overload on the right cardiac chambers, the consequent pulmonary hypertension and right atrial enlargement increase the vulnerability to atrial arrhythmias and stroke. The late onset of pulmonary hypertension was presumed to be the reason for the very late onset of the symptoms of this patient.

Key words: Elderly patient, partial atrioventricular septal defect.

INTRODUCTION

The atrial septal defect (ASD) is one of the most common congenital heart anomalies in adults (Bonow RO et al., (2012). However, patients with partial atrioventricular septal defects (PAVSD), previously known as ostium primum ASD, usually present at an early age, and only few of those can reach adulthood without timely diagnosis and surgical correction. (Tandon R et al., 1974; Barnett MG et al., (1988)). Herein, we report the oldest case of native PAVSD that was diagnosed at an age of 83.

CASE REPORT

An 83-year-old female was admitted to our hospital with syncope and near-syncope attacks and palpitations for the last three years. The radiographic coronary angiogram performed in another hospital two months ago did not reveal any coronary artery disease. Physical examination revealed an irregular pulse at 82 beats/min, blood pressure of 110/70 mmHg, cardiac examination revealed III/IV pan-systolic murmur at the left sternal border. There were no signs of heart failure. The chest radiograph demonstrated increased pulmonary

vascularity, as well as enlarged central pulmonary arteries and cardiomegaly, consistent with left-to-right shunting.

Electrocardiogram revealed right ventricular hypertrophy, atrial fibrillation with approximately 82 beats/minute, an electrical axis of approximately +120°, right ventricular hypertrophy and right bundle branch block (inverted and plane T wave in V1 and V2, respectively) (figure 1). An electrical axis of approx +120° is uncommon for an AVSD, possibly one of the reasons for the lesion being undiscovered for so long. Transthoracic echocardiogram showed dilated right and left atria, and a 2,8 cm defect at the lower interatrial septum (figure 2), moderate right atrioventricular valve (AV) insufficiency with a maximum gradient of 44 mmHg (figure 3), which indicates an elevated systolic pulmonary arterial pressure (PAP), and a moderate left AV insufficiency. The ejection fraction was 55 %. Transesophageal echocardiography revealed a cleft mitral valve (Figure 4) and a 2.5 cm defect at the lower atrial septum (Figure 5). Cardiac catheterization revealed: a Qp/Qs ratio of 3.0, right and left atrial pressures of 18 mmHg both, a systolic pulmonary artery pressure (PAP) of 40 mmHg. The patient refused the surgery for the replacement of the mitral valve with a

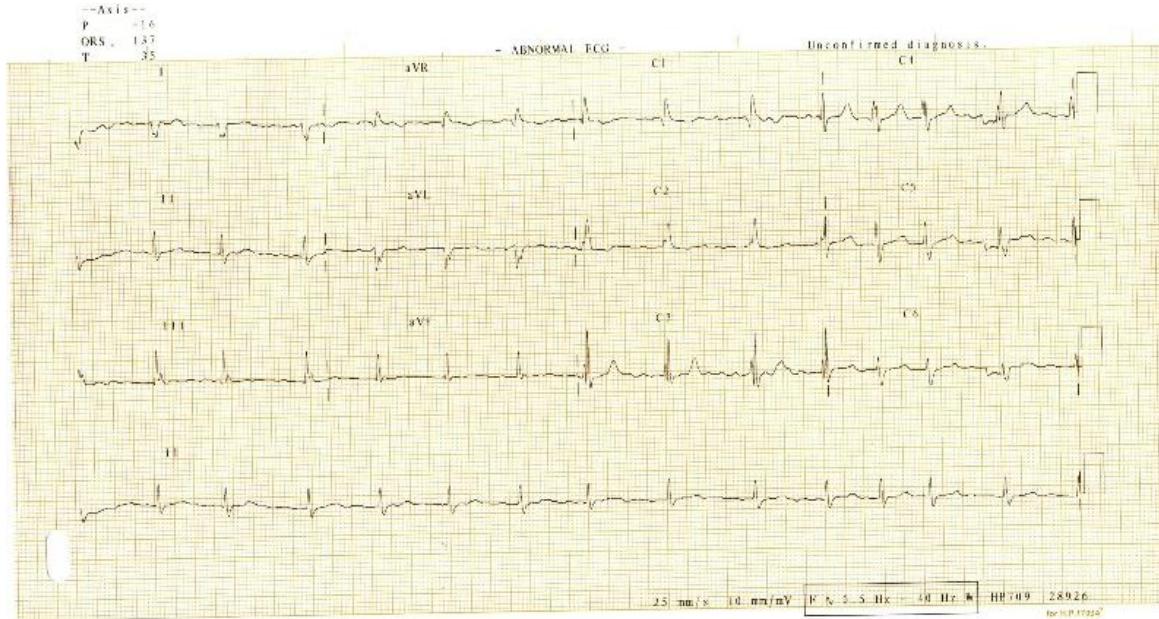


Figure 1. The Electrocardiogram indicating atrial fibrillation with approximately 82 beats / minute, an electrical axis of approximately $+120^\circ$, right ventricular hypertrophy and right bundle branch block.

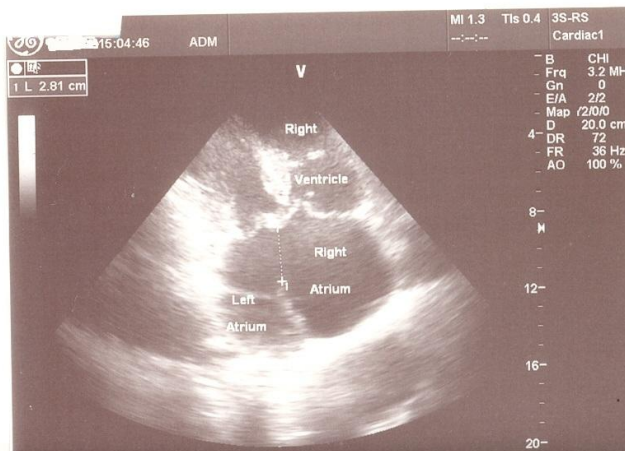


Figure 2. Transthoracic echocardiogram showing a dilated right and left atrium with normal ventricles and a 2,8 cm defect at the lower interatrial septum.

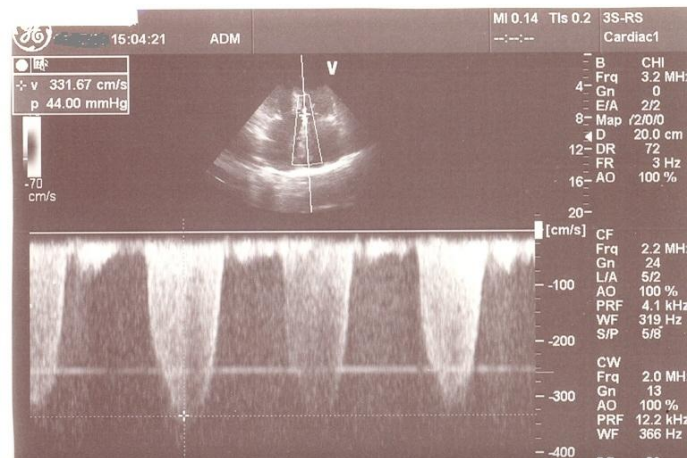


Figure 3. Tricuspid insufficiency with a maximum gradient of 44 mmHg.

biological prosthetic one and the repair of the primum ASD. Consequently, her medical treatment was designed accordingly, consisting of diuretics when needed and oral anticoagulant therapy for atrial fibrillation, and she was advised to stick to her treatment.

DISCUSSION

Partial atrioventricular septal defect is characterized by an ostium primum atrial septal defect (ASD) with an intact ventricular septum (Minich LL et al. (2010), and usually a

cleft in the anterior mitral valve leaflet (Cetta F et al. (2008). Rarely, it can be associated with other anomalies such as sinus venosus type of ASD (Ardıç I et al. (2012). Patients presenting with PAVSD in adulthood are rare (Barnett MG et al. (1988). Life expectancy in patients with PAVSD ranges from 3 to 37.5 years (Gauer IC et al. (2003). Arrhythmias are the most common cause of symptomatology in unoperated ASD patients, occurring in 20 % of patients, especially after 30 years of age (Kharouf R et al. (2008).

Medline research revealed a reported case of a 75-year-

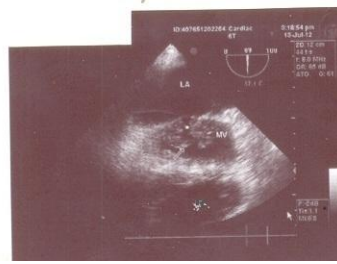


Figure 4. Transesophageal echocardiography revealing the cleft mitral valve (arrow) and the muscular structure dividing the valve from the LV.



Figure 5. Transesophageal echocardiography showing a 2.5 cm defect at the lower atrial septum (arrow) with normal ventricles.

old man who presented with dyspnea and deteriorating general condition, and passed away shortly after presentation. The autopsy revealed a PAVSD and an extremely dilated heart (Gauer IC et al., (2003)). And, recently, a 69-year-old female with partial atrioventricular canal and pulmonary hypertension was reported, successfully treated with complete surgical repair of the defect (Woźniak S et al., (2011)). The case presented in this paper has been almost asymptomatic for over 83 years except for the episodes of syncope and palpitations during the last three years. To the best of our knowledge, the present patient is the oldest PAVSD case reported in the literature.

Echocardiography provides full diagnostic information for all forms of atrioventricular defects. Transthoracic echocardiography and color Doppler interrogation of the atrial septum, combined in some cases with bubble contrast should confirm the presence of ASDs in the majority of elderly patients presenting with an unusual clinical entity and a dilated right heart chamber (Meisner H et al., (1998)). Surgery is mandatory for primum ASDs due to progressive left AV regurgitation, development of arrhythmias, and likelihood of pulmonary vascular disease (Murashita T et al., (2004)).

Although the efficacy of surgical treatment in adults with PAVSD has not been well documented, abnormalities of

the left AV valve and subvalvular apparatus have been shown to be risk factors for residual regurgitation and an increased incidence of early and late re-operation after repair of incomplete atrioventricular septal defects (Murashita T et al., (2004)). Thus, even mild regurgitation should be repaired. Our patient had a moderate degree of left AV regurgitation with cleft appearance. We planned to replace the mitral valve with biological prosthetic one, in addition to repairing the ASD. Because of the unpredictability of mitral valve function, lifelong follow-up should be stressed in these patients (Barnett MG et al. (1988)). Somerville J. reviewed the factors responsible for spontaneous deterioration and death of patients with ostium primum defects without surgical correction (Somerville J (1965)). The commonest associated factor in patients who died in sinus rhythm was severe mitral regurgitation. Additionally, the development of established arrhythmias is the commonest cause of deterioration. However, pulmonary hypertension uncommonly complicates an ostium primum defect, but occasionally causes death with pulmonary artery thrombosis and right heart failure (Somerville J (1965)). The late onset of pulmonary hypertension was presumed to be the reason for the very late onset of the symptoms of this patient. She refused to undergo any surgical correction, and was eventually discharged with a proper

medical therapy. She was advised to stick to the monthly INR test follow-up visits and yearly re-assessment visits.

CONCLUSION

Although ASDs can be easily diagnosed, sometimes cardiologists and pediatricians may overlook the diagnosis. Our case reminded us that geriatric physicians and adult cardiologists should be aware of this reality, and keep in mind congenital heart defects in the elderly patients with unusual symptomatology, even those that can be quite rare in advanced age such as PAVSD.

Consent

Written informed consent was obtained from the next of kin of the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

SG designed the study, carried out subject recruitment, performed echocardiography and CAG, analysed the data, and wrote the manuscript. DC and HM assisted in the recruitment and manuscript revision. All authors read and approved the final manuscript.

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