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Partial Atrioventricular Septal Defect in an Elderly Patient: A Case Report

Marwan El Qady[1*], Mina Boutgourine[1], Ahmed Salah Tandia[1], Saloua El Karimi[1], Mustapha El Hattaoui[1]

[1]Department of Cardiology, Mohammed VI University Hospital, Marrakesh, Morocco

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***Corresponding author:** Marwan El Qady
Department of Cardiology, Mohammed VI University Hospital, Marrakesh, Morocco

Abstract **Case Report**

Long-term survival of patients with partial atrioventricular septal defect (PAVSD) without operative therapy is very rare. We report the case of an 80-year-old woman who presented with dyspnea, palpitations, and deteriorating general condition and was diagnosed by routine echocardiography.

Keywords: dyspnea, fatigue, partial atrioventricular septal defect (PAVSD).

INTRODUCTION

Partial atrioventricular septal defect (PAVSD) is a form of endocardial cushion defect. It is a congenital heart defect rarely seen in adults. Survival of patients with this defect into the sixth or seventh decade of life without operative treatment is extremely rare. We report an 80-year-old woman with a partial AV septal defect who presented with progressive dyspnea, fatigue and palpitations.

CLINICAL CASE

We report an 80-year-old woman with a partial AV septal defect who has had dyspnea for 30 years (NYHA class II) that has become progressive over the past 3 years (NYHA class III) associated with palpitations usually of short duration lasting a few minutes and generalized fatigue. Her medications were carvedilol, sintrom and furosemide. Clinical examination found a hemodynamically and respiratory stable patient with blood pressure at 135/95 mm Hg and heart rate at 95 beats/min. Cardiac auscultation revealed a holo-systolic murmur rated III/VI of maximal intensity at the apex, a systolic murmur at the tricuspid focus and a B2 burst at the pulmonary focus. Pulmonary auscultation was unremarkable and there was no evidence of edema and jugular venous distension. Chest X-ray revealed cardiomegaly. The ECG showed atrial fibrillation at 100 beats per minute. Transthoracic echocardiography (TTE) showed dilated atria, moderate septal hypertrophy, moderate mitral regurgitation, ostium primum septal defect measuring 14 mm with left-to-right shunt, the left ventricle was non-dilated, hypertrophic, with good systolic function, LVEF 55%, the right ventricle was dilated with moderate systolic dysfunction with signs of pulmonary hypertension (65 mm Hg) and severe tricuspid insufficiency. The IVC was undilated and compliant. A diagnosis of partial AV septal defect was made. Surgical repair of the defect was offered to the patient, but she refused further diagnostic and therapeutic interventions. As a result, she was placed on medical therapy with furosemide, spironolactone, anticoagulant therapy, and beta-blockers and was advised to stick with her treatment.

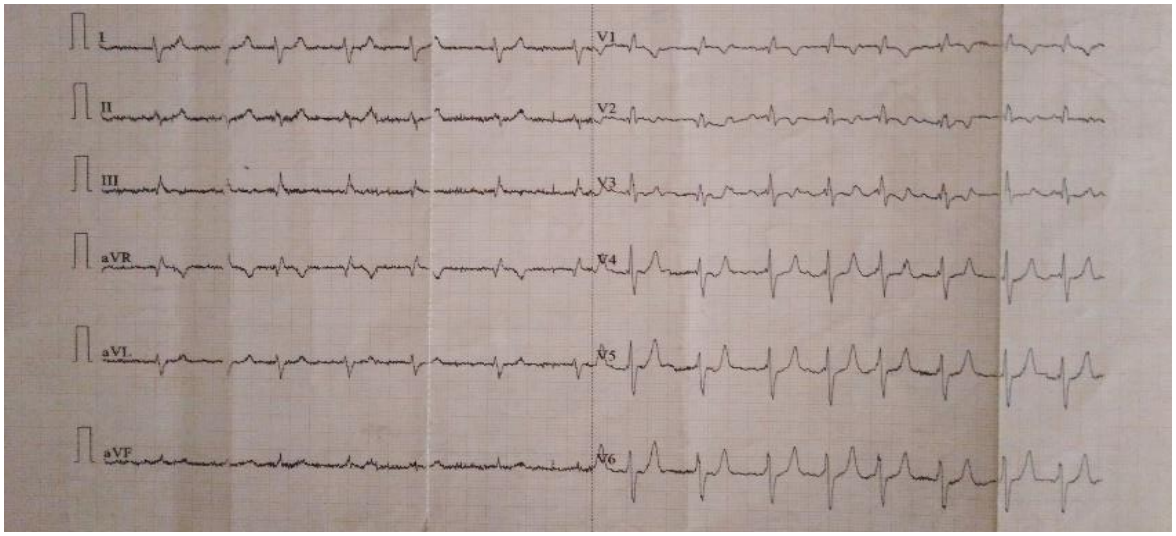

Figure 1: EKG showing atrial fibrillation at 100 beats per minute

Figure 2: TTE that objectifiespartial AV septal defect

DISCUSSION

Partial AV septal defect represent part of the spectrum of AV septal defects, including an atrial septum ostium primum defect but with two separate AV rings, no significant interventricular septal defect, and usually a noticeable anterior cleft of the mitral valve [1].

Partial atrioventricular septal defect in elderly patients is extremely rare. Several large series [2, 3] of patients with a primum atrial septal defect have included single patients in the seventh decade of life. There have been only three reports [1, 4, 5] of patients with this defect surviving into the eighth decade of life.

Conditions for long-term survival include [6]:

1. Mild valve dysfunction;
2. Maintenance of sinus rhythm and absence of arrhythmias;
3. Mild cardiac dysfunction, including late onset of heart failure;
4. No or other simple cardiac abnormalities.

The presentation of partial AV septal defect is more variable. Surgical series have shown that these patients are usually symptomatic from early to mid-life [7, 8]. Patients with primum atrial septal defects who remain asymptomatic and survive to the seventh and eight decades without surgery, should be considered an exception. Somerville, in an early report [4], recognized that patients with partial AV septal defects present symptoms with increasing frequency after the third decade of life.

All major surgical series [7, 8] have clearly demonstrated that nearly three-quarters of these patients are symptomatic at presentation, with impaired exercise capacity and exertional dyspnea being by far the most common manifestations.

Partial AV septal defect is widely recognized as a prognostic factor in the progression of arrhythmias, such as atrial fibrillation, complete AV block, sinus disease, and ventricular tachycardia, which are the most common causes of deterioration and frequently occur with age [9].

Our patient was symptomatic, she had progressive dyspnea with episodes of short duration palpitations since the age of 54 years but the patient never consulted until the age of 80 years following the worsening of the symptomatology.

Two-dimensional echocardiography clearly defines the characteristic anatomy and is the preferred diagnostic procedure. Cardiac catheterization or additional angiography may be necessary if clinical signs suggest the presence of pulmonary hypertension, mitral insufficiency or associated ischemic heart disease.

The prognosis of these patients depends on the severity of the partial AV channel defects and the associated adverse conditions such as arrhythmia, severity of mitral insufficiency or pulmonary hypertension.

Our patient had a moderate degree of mitral regurgitation and clinical signs and echocardiographic findings of pulmonary hypertension.

The prognosis of patients treated medically with ostium defect primum is poor [7, 4]. Low operative risk and excellent long-term outcomes support surgical repair of partial AV septal defects as the primary mode of treatment for both pediatric and adult patients [7, 8]. The preferred age for elective repair of partial AV septal defects is 3 to 5 years.

Hynes *et al*., [11] believe that surgery is indicated in symptomatic elderly patients with partial AV septal defects and the results of their small series as well as those reported by St. John Sutton *et al*., [11], found that most elderly patients with an atrial septal defect have disabling symptoms that can be significantly decreased by operation and that surgical intervention is associated with low morbidity and mortality.

CONCLUSION

Our case adds further evidence that survival is possible in patients with a partial AV septal defect. 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.

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