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An Unusual Presentation of Partial Atrioventricular Canal Defect in an Elderly Woman

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Abstract **Case Report**

A partial atrioventricular canal defect (PAVCD) is a form of endocardial cushion defect. It's a rare congenital cardiac malformation, few reported patients have survived into the seventh decade of life. A 71-year-old female with a history of diabetes and hypertension was admitted for heart failure (HF) symptoms. Transthoracic echocardiography revealed a left to right shunt in the basal portion of the interatrial septum, 19mm ostium primum atrial septal defect (ASD), a mitral valve cleft, and severe mitral valve regurgitation. PAVCD consists of an ostium primum ASD and the dividing of the common atrioventricular (AV) valve into 2 separate AV orifices, resulting in a so-called cleft in the mitral valve. The ventricular septum is intact. Echocardiography is the key tool for diagnosis and anatomic classification. Surgical repair is recommended in adults with primum ASDs causing impaired functional capacity, right atrial or right ventricular enlargement, and left-to-right shunt without cyanosis, as long as systolic pulmonary artery pressures are lower than 50% of systemic pressures. Reconstructive surgery includes, in addition to the closure of the interatrial septum, a suturing of the mitral cleft. The estimated mean age at death of the patients with ostium primum ASD is about 40 years. The objective of this clinical case is to highlight the good clinical tolerance of atrial shunts and the anatomical particularities in the PAVCD.

Keywords: Partial Atrio-Ventricular Canal Defect, adult congenital heart disease.

INTRODUCTION

A Partial Atrio-Ventricular Canal Defect (PAVCD) is a form of endocardial cushion defect, infrequently encountered in adults. It consists of an ostium primum atrial septal defect (ASD) with two separate atrioventricular (AV) valve rings and a cleft mitral valve leaflet [1].

It's a rare congenital cardiac malformation. Echocardiography is the key tool for the diagnosis and anatomic classification of this malformation.

CASE REPORT

A 71-year-old female patient, with a history of diabetes, and hypertension was admitted to our emergency room for heart failure symptoms made of class IV New York Heart Association dyspnea, associated with lower limb edema.

Physical examination revealed a hemodynamically stable patient with a mitral regurgitation murmur, ASD murmur, and mild signs of pulmonary congestion.

An electrocardiogram (EKG) showed atrial fibrillation with a ventricular rate response of 102 beats/min, right bundle branch block, and diffuse repolarization abnormalities.

Chest X-ray showed posterior bilateral pleural effusion, diffuse thickening of the interstitial peribronchial vascular tissue, and enlargement of the cardiac shadow.

Transthoracic echocardiography (TTE) showed a normal size of the left ventricle (LV), non-hypertrophic with normal ejection fraction. Color-Doppler evaluation revealed a left to right shunt in the basal portion of the interatrial septum, 19mm ostium primum ASD, mitral valve cleft without the involvement of the interventricular septum, and severe mitral valve regurgitation. The right and left atrium were dilated. The right ventricle was mildly dilated with moderate to

severe tricuspid regurgitation. Systolic pulmonary artery pressure was 62 mmHg (Figure 1 and 2).

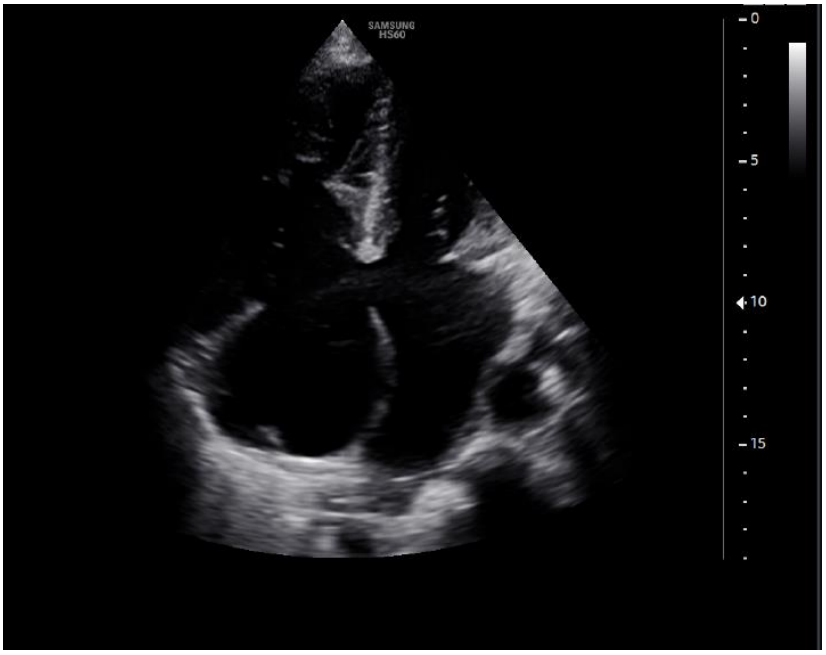

Figure 1: Echocardiography apical 4 chamber view showing the ostium primum ASD and PAVCD

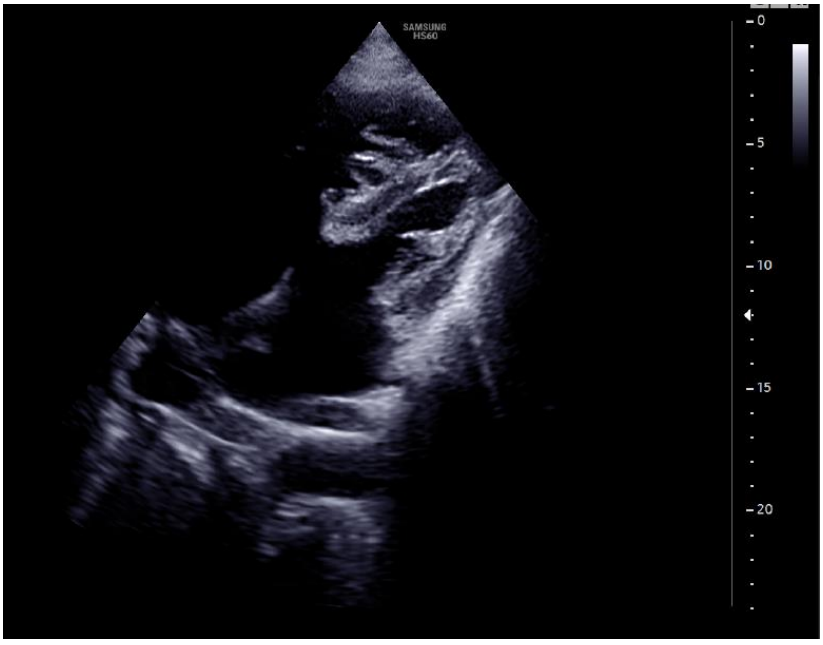

Figure 2: Echocardiography view showing the mitral valve cleft

The patient was managed medically with a salt and fluid restriction, and intravenous diuretics initially. An optimization of heart failure medical therapy was done. The patient denied any other exploration and wanted to be treated medically.

DISCUSSION

Atrioventricular septal defects (AVSD) are anatomic defects arising from faulty development of the embryonic endocardial cushions. This spectrum ranges from PAVCD made of a primum ASD and cleft mitral valve, to complete AVSD made of defects of both the primum atrial septum and interventricular septum with the presence of a common AV valve.

A PAVCD consists of an intact ventricular septum and an ostium primum ASD and the dividing of the common AV valve into 2 separate AV orifices, resulting in a so-called cleft in the mitral valve. It occurs frequently in non-down syndrome patients over 90% counter to complete AV septal defects.

ASDs are one of the most common congenital heart anomalies after mitral valve prolapse and bicuspid aortic valve and can be diagnosed until adulthood [2]. AV septal defect incidence is 3% to 5% of all congenital heart malformations, and PAVCD incidence is at 1-2% of it [3].

A primum ASD is an interatrial communication between the AV valves and the anterior-inferior margin of the fossa ovalis. The ASDs discovered in adults are most often ostium secundum or more rarely sinus venosus. A primum ASD in the AV canal is usually present in young children or even before birth and its discovery in adulthood has become very rare.

In the PAVCD, the AV valves are almost always abnormal, including a cleft in the anterior mitral leaflet. The direction and amount of flow through an ASD are determined by its size and by the relative atrial pressures; this relates to the compliances of the left and right ventricles. The estimated mean age at death of the patients with ostium primum septal atrial defect is around 40 years [4].

It is most often revealed during a routine clinical examination revealing a systolic murmur at the pulmonary focus or the mitral focus (cleft mitral insufficiency). The typical EKG features of PAVCD are the association of the right bundle branch block, left axis, counterclockwise rotation of the vector loop in the frontal plane, first-degree AV block, and atrial fibrillation [5]. Our patient has these typical electrocardiographic findings.

TTE is the gold standard for the diagnosis and anatomic classification of this malformation. It shows the ostium primum ASD, the left-right shunt, the underlying common AV valve, and the defect of the ventricular septal inflow. It also helps to determine the resounding on the right and left cardiac chambers and the pulmonary artery pressures.

Cardiovascular magnetic resonance is recommended when additional quantification of ventricular volumes, AV valve regurgitation, or intracardiac shunting is necessary. Cardiac catheterization helps to identify Eisenmenger syndrome and is required for non-invasive signs of elevated pulmonary artery pressures to determine pulmonary vascular resistance.

The guidelines recommend surgical repair in adults with PAVCD with significant right ventricle overload made of impaired functional capacity, right atrial or right ventricular enlargement, and left-to-right shunt to cause physiological sequelae without cyanosis at rest or during exercise, as long as systolic pulmonary artery pressures are lower than 50% of systemic pressures and pulmonary vascular resistance is less than one-third of systemic vascular resistance [6].

Valve surgery is recommended in symptomatic patients with moderate to severe valve regurgitation or in asymptomatic patients with severe regurgitation when the LV systolic diameter is up to 45mm and the LV ejection fraction is up to 60%.

Reconstructive surgery is usually performed around the age of 5. It includes, in addition to the closure of the interatrial septum, a suturing of the mitral cleft.

CONCLUSION

PAVCD is a congenital cardiac anomaly, few reported patients have survived into the sixth or seventh decade of life.

We report the case of a patient with a PAVCD at the age of 71 years which highlights the good clinical tolerance of atrial shunts and the anatomical particularities of the mitral valve in this entity.

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