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Pacman Heart as a Congenital Cardiac Defect Associated with Flail Mitral Valve and a Partial form of Shone's Complex; A Case Report

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ABSTRACT

Partial muscular inter-ventricular septal defect (VSD) or Pacman heart is a rare congenital or occasionally acquired anomaly. Concurrent Pacman heart and Shone's complex are extremely rare and have never been reported until now. We described a 37-year-old male patient with congenital Pacman heart, flail mitral valve (FMV), and a history of multiple congenital anomalies, including subvalvular aortic stenosis, bicuspid aortic valve (AV), and coarctation of the aorta compatible with the incomplete form of Shone's complex. Cardiac surgery was not necessary to perform for this anomaly due to the absence of a left-to-right shunt through this partial defect.

Keywords: Flail mitral leaflet, Congenital heart defects, Bicuspid aortic valve

Introduction

Shone's complex is a rare congenital anomaly characterized by sequential obstructive lesions at multiple levels of the left heart structures and aorta (1). Partial ventricular septal defect (VSD) also known as Pacman heart is a rare cardiac abnormality that may be congenital or occasionally acquired (2). This abnormality is often clinically silent and is diagnosed incidentally when echocardiography is carried out to evaluate simultaneous structural heart diseases. The combination of Pacman anomaly and Shone's complex is extremely rare. We described a case of congenital Pacman heart associated with flail mitral valve (FMV) and a partial form of Shone's complex comprising subvalvular aortic stenosis (AS), bicuspid aortic valve (AV), and coarctation of the aorta. Two-dimensional (2D) transthoracic echocardiography (TTE) is the imaging modality of choice for the diagnosis of Pacman. The incremental data are provided by three-dimensional (3D) and transesophageal echocardiography (TEE) regarding simultaneous structural heart diseases (3).

Case Report

A 37-year-old man complaining of progressive exertional dyspnea (New York Heart Association class 3) for six months and fever for one week was referred to our medical center. His medical history included subaortic web resection and aortic coarctation repair 22 and 28 years ago, respectively.

The patient had a febrile presentation (temperature: 38.5^{0}C), normal blood pressure (right and left arm blood pressure: 120/70 mmHg), and a regular pulse rate of 76

beats/min. Cardiac auscultation revealed a high-pitched holosystolic murmur best heard at the apex and a high-pitched early diastolic decrescendo murmur best heard at the 3rd left intercostal.

TTE showed severe highly eccentric mitral regurgitation (MR) with the rupture of chordae tendineae of the anterior MV leaflet and flail leaflet (Figure 1A-C). The left ventricle (LV) was enlarged with normal LV ejection fraction (EF). The AV was bicuspid with moderate aortic regurgitation (AR) and no residual subvalvular stenosis (Figure 1D). A triangular-shaped defect was seen in the muscular portion of the left ventricular side of the interventricular septum without interventricular communication. The color Doppler study showed no left-to-right shunt through the defect. The so-called "Pacman defect" resembled a mouth (opening in diastole and closing in systole). The largest size of the defect was at early diastole (Figure 2A-B, Movie S1). 3D-TTE was also beneficial for visualizing the defect and the FMV. No vegetation was observed on TTE and TEE. The patient's two separate blood cultures were both positive for *Streptococcus viridans*, which was the most likely cause of FMV. The patient was started with antibiotics for four weeks and underwent mitral and aortic valve replacement using an open approach two months after the antibiotic therapy. Medical follow-up was uneventful.

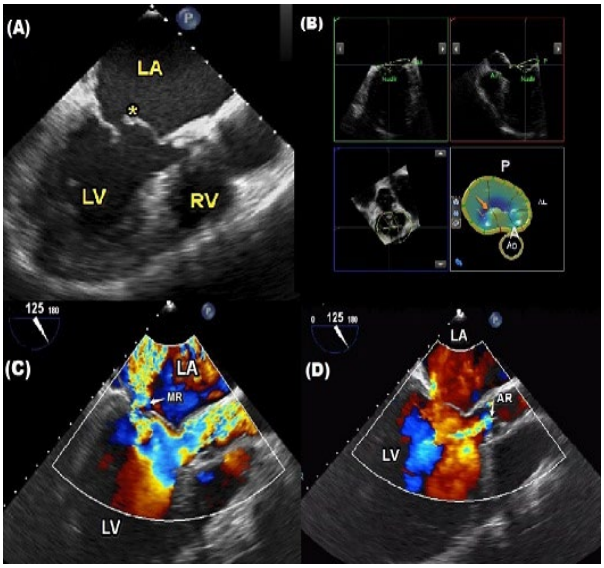

Figure 1. Flail mitral valve (FMV) and severe MR; A) The midesophageal two-chamber view of the mitral valve at 60 degrees revealed flail A2 scallop (yellow asterisk). B) Three-dimensional transesophageal echocardiography (TEE) using the multiplanar reconstruction (MPR) method confirmed flail A2 scallop (red arrow). C) Color Doppler TEE at 125 degrees showed severe MR (white arrow). D) Color Doppler TEE at 125 degrees showed moderate AR (white arrow). LA = left atrium; LV = left ventricle; RV = right ventricle; AV = aortic valve; MR = mitral regurgitation; AR = aortic regurgitation

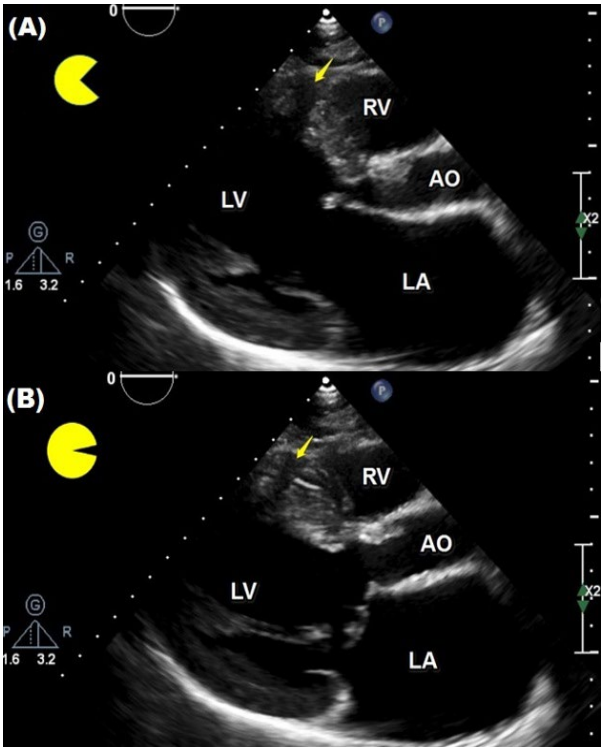

Figure 2. Partial VSD: A) Two-dimensional transthoracic echocardiography (TTE-Parasternal Long Axis View) revealed partial ventricular septal defect (VSD) with a maximal opening like a mouth during diastole (yellow arrow). B) Closed partial VSD during systole (yellow arrow).

Discussion

Partial VSD is included in the differential diagnosis of complete VSD, left ventricular non-compaction, and focal hypertrophic cardiomyopathy. Accordingly, accurate diagnosis using cardiac imaging modalities is rather important since it prevents unnecessary surgeries and treatments. Because partial VSD is rare, reporting it, describing its association with Shone's complex, and sharing echocardiographic images will assist cardiologists in making a proper diagnosis and treatment plan.

The previous case reports described the Pacman anomaly associated with mitral valve prolapse, bicuspid aortic valve and Parachute mitral valve. The combination of Pacman heart and Shone's complex is extremely rare and has not been reported until now.

The Pacman anomaly appears as a partial defect of the muscular portion of the interventricular septum, which changes size during systole and diastole **(4)**. It is often clinically silent and is diagnosed incidentally using TTE. The complications of the Pacman deformity, such as myocardial rupture and conduction disturbance, are very rare **(5)**, therefore, cardiac surgery is not recommended for this kind of partial defect. Cardiac surgery which was carried out for this patient was due to the severe MR and moderate AR but, the surgeon did not repair the partial VSD.

Conclusion

Due to the absence of a left-to-right shunt of this partial defect, cardiac surgery is not recommended.

Acknowledgments

None.

Conflict of Interest

The authors declare that there is no conflict of interest.

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