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CASE REPORT

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Double orifice mitral valve in a patient with bicuspid aortic valve and coarctation of the aorta: A rare presentation

Fariba Bayat[1] | **Mohammad Hasan Namazi**[1] | **Mohammad Khani**[1] | **Shadi Shekarkhar**[1] | **Aref Fatehi**[1] | **Mohammadreza Tabary**[2] | **Isa Khaheshi**[1]

[1]Cardiovascular Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

[2]School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

Correspondence
Shadi Shekarkhar, Cardiovascular Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
Email: Shadi.shekarkhar@yahoo.com

Abstract

Double orifice mitral valve is a rare condition and may be accompanied by a bicuspid aortic valve (sometimes normal functioning) and coarctation of the aorta. Echocardiography is valuable in detecting the accompanied anomalies. Management depends on the severity of mitral valve function and the severity of associated anomalies.

KEYWORDS

3-dimensional echocardiography, bicuspid aortic valve, coarctation of the aorta, double orifice mitral valve

1 | INTRODUCTION

Double orifice mitral valve (DOMV) is an uncommon congenital anomaly with an incidence of 0.05% in the general population[1,2] and was first described by Greenfield in 1876.[3] DOMV is often accompanied by other congenital heart defects, especially atrioventricular septal defects. The isolated occurrence of DOMV is not common in the general population.[3] The most common anatomic presentation is an accessory bridge of fibrous tissue that expands between the wall and aortic leaflets and divides the valve openings into two segments.[4] This kind of arrangement in the left valve is seen in atrioventricular septal defect. Duplicating the entire valvular structure is the rarer variant, mostly involves the otherwise-normal mitral valve. Thus, left atrium and ventricle are connected by two separated valves. This anomaly is discovered accidentally when evaluating for other complex anomalies such as tricuspid atresia.[2] This report describes the combination of a double orifice mitral valve with the bicuspid aortic valve, severe coarctation identified by 2D and 3D transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) and also its management.

2 | CASE REPORT

A 25-year-old man was taken to the cardiology clinic due to uncontrolled hypertension and undetectable distal pulses in order to rule out coarctation. He has had a history of hypertension for 2 years. His medication was Carvedilol 6.25 mg BID and Losartan 25 mg BID with poor control of hypertension. No further work-up was performed to evaluate other underlying conditions.

In echo laboratory, he underwent 2D and 3D TTE and TEE; during study by Philips IE33, these data were revealed: The size of 4 cardiac chambers was normal with proper right ventricle (RV) and left ventricle (LV) ejection fraction, the aortic valve was bicuspid (fusion of right to left coronary cusp) with no aortic regurgitation (AR) or aortic stenosis (AS) (aortic valve mean gradient = 2.7 mm Hg, peak gradient = 4 mm Hg), normal mitral valve with no mitral stenosis

(MS), trivial mitral regurgitation (MR) (mitral valve mean gradient = 2 mm Hg, peak gradient = 3.9 mm Hg); he suffered a severe coarctation (narrowest part of aorta = 3 mm) with peak systolic gradient 62 mm Hg and diastolic gradient 11 mm Hg (Figure 1A and B).

The interesting finding was his double orifice mitral valve, which was symmetric (the anterolateral orifice and posteromedial orifice had approximately the same size) without stenosis or regurgitation (Figure 2).

For more and precise evaluation, TEE was performed and confirmed previous data.

He also underwent CT angiography, and the presence of coarctation and aberrant left subclavian artery was seen. The size of the aorta at the diaphragm level was 16.2 mm.

Due to the normal functioning mitral valve and normal functioning bicuspid aortic valve, the normal size ascending aorta, and a significant aortic coarctation, the patient was scheduled for balloon dilatation and stenting.

The right femoral artery, right radial artery, and left femoral vein were punctured. One pigtail catheter was used in aortic arch through the right radial artery, and the gradient was measured between the pigtail and femoral artery, which was 70 mm Hg. The coarctation stenosis was crossed by hydrophilic wire 0.035 through the multipurpose catheter and after that, the wire changed to Amplatz Super Stiff™, which was deployed at the aortic root. One pacemaker wire was positioned in RV through the femoral vein as a backup. The bare, self-expandable stent "Sinus-xl (OptiMed)" size

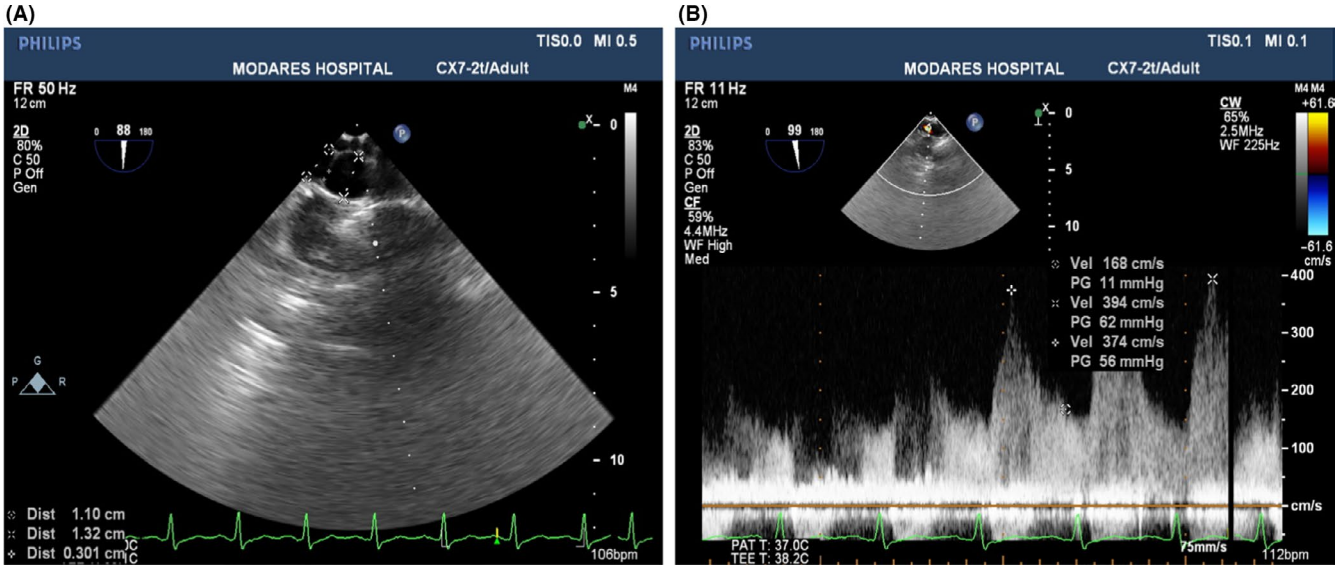

FIGURE 1 A, Upper esophageal (UE) descending aorta long-axis view at the angle of 88 degrees. It provides an opportunity for detailed visualization of coarctation point (3 mm) and the size of aorta proximal (11 mm) and distal to it (13 mm); B, Upper esophageal (UE) descending aortic long axis at the angle of 99 degrees. It shows coarctation with systolic gradient = 62 mm Hg and diastolic gradient = 11 mm Hg (holodiastolic tail)

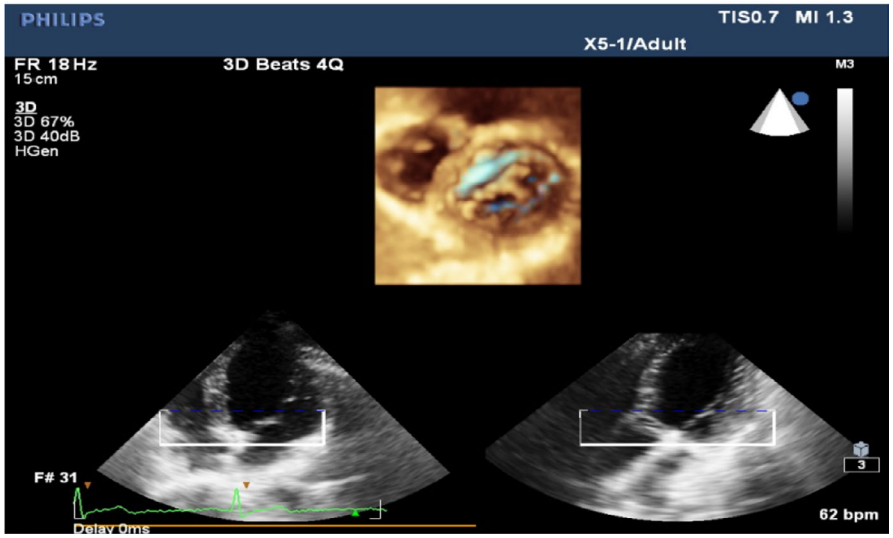

FIGURE 2 3D echocardiography, which shows a double orifice mitral valve

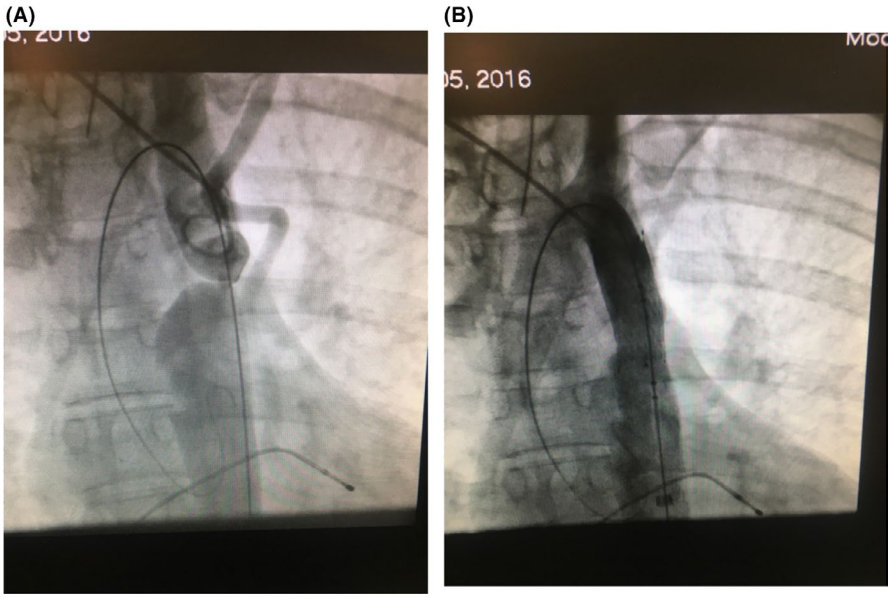

FIGURE 3 A, AP view of the coarctation of aorta before coarctoplasty. A large collateral artery is seen. Transradial pigtail catheter is seen adjacent to the stenosis. Pacemaker lead is seen in RV; B, AP view of the coarctation after coarctoplasty. Dilation of stenosis with bare-metal stent and postdilation with balloon

20 × 40 was deployed through the 10 French femoral sheath. Then, the stent was postdilated by balloon BIB®16 × 40 with proper final results and the gradient decreased to 5 mm Hg (Figure 3A and B).

Postprocedural echocardiography showed that the gradient has decreased significantly (peak systolic gradient decreased to 4.5 mm Hg).

The patient was discharged in good condition. In follow-up visits in months 1 and 6, he had no complaints and had good functional activity. Follow-up echocardiography result after 6 months showed an ejection fraction of 60%, no AS, no AR, no MS, trivial MR, and peak systolic gradient of 3.6 mm Hg in thoracic descending aorta with no residual stenosis.

3 | DISCUSSION

Double orifice mitral valve was first described by Greenfield in 1876. Banerjee et al reported the incidence of DOMVs as 0.05% in 13 400 new cases. It is diagnosed mainly during childhood in routine cardiac evaluation.[5] Moreover, an isolated DOMV is not common, and DOMV is often diagnosed as an appendix of other associated anomalies, such as coarctation like the experience in our case.[2,3]

The mitral valve can function normally in about 50% of patients with DOMV. In the other 50%, it may cause clinically significant mitral stenosis or mitral regurgitation. The patients with an isolated DOMV usually have no functional abnormalities or clinical findings of mitral regurgitation or stenosis.[6,7] In our case, despite the presence of other anomalies, the function of mitral valve was normal. In terms of size, the mitral orifices are not equal in most of the patients. Atrioventricular septal defect, aortic coarctation, interrupted aortic arch, patent ductus arteriosus, primmum atrial septal defect, tetralogy of Fallot, and Ebstein anomaly are among the most commonly associated lesions.[8,9] 2D transthoracic echocardiography is the main diagnostic tool; however, transesophageal echocardiography may provide more functional details.[10] In our case, DOMV was associated with bicuspid, but normal functioning aortic valve, normal size ascending aorta, and severe aortic coarctation.

Management is completely related to anatomic abnormalities in the mitral valve apparatus, other accompanying anomalies, and clinical symptoms. Surgery is an option in case of severe stenosis or incompetence. Other associated cardiac anomalies may also necessitate surgery.[11] Due to severe increased gradient, we did stent coarctoplasty and recommended follow-up echocardiography to detect subsequent abnormal hemodynamics or complications, as the function of the mitral valve was normal. This case report endorses the role of echocardiography in detecting congenital anomalies in patients with nonspecific symptoms.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

AUTHOR CONTRIBUTIONS

Each author has contributed sufficiently and met the criteria for authorship. Here, we state all the authorships. *Fariba Bayat:* is the main author, contributed to data acquisition and manuscript preparation. *Mohammad Hasan Namazi:* contributed to data acquisition and performing the laboratory tests. *Mohammad Khani:* contributed to manuscript writing. *Shadi Shekarkhar:* is the corresponding author and designed and supervised all the aspects. *Aref Fatehi:* contributed to data acquisition in terms of performing cardiac evaluations. *Mohammadreza Tabary* and *Isa Khaheshi:* contributed to

manuscript writing, editing and final revision of the manuscript and the submission process.

ORCID

Mohammadreza Tabary https://orcid.org/0000-0002-3746-7371
Isa Khaheshi https://orcid.org/0000-0001-5926-4797

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