

A rare case of quadrileaflet mitral valve and ostium primum atrial septal defect

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Abstract

A 60-year-old female was referred to our clinic for evaluation of her rapidly progressive dyspnea, she had no previous history of heart disease. A murmur was noted on her examination and transthoracic echocardiography was so difficult to be performed due to poor acoustic windows so she was referred to do a transesophageal echocardiography that showed an ostium primum atrial septal defect (ASD) with left to right shunt and a quadrileaflet mitral valve with severe regurgitation. Later on, she underwent surgery with Ostium Primum ASD closure by a patch and double cleft repair by suture after right heart catheterization.

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Abstract

A 60-year-old female was referred to our clinic for evaluation of her rapidly progressive dyspnea, she had no previous history of heart disease. A murmur was noted on her examination and transthoracic echocardiography was so difficult to be performed due to poor acoustic windows so she was referred to do a transesophageal echocardiography that showed an ostium primum atrial septal defect (ASD) with left to right shunt and a quadrileaflet mitral valve with severe regurgitation. Later on, she underwent surgery with Ostium Primum ASD closure by a patch and double cleft repair by suture after right heart catheterization.

Introduction:

Partial atrioventricular canal (PAVC) defect is a form of endocardial cushion defect that is infrequently seen in adults with very rare case reports published. Although the results of surgery in younger individuals is well known and studied, yet there is a very rare data about results of surgery in older patients.

Case Report:

A 60-year-old female patient was referred to our clinic for evaluation of her rapidly progressive dyspnea that started few days before her presentation. She had no previous history of cardiovascular disease, nor other risk factors. On clinical examination, a systolic murmur was heard over the apex. A transthoracic echocardiography was difficult due to poor acoustic windows but there was marked dilatation of the right cardiac chambers so the patient was scheduled for a transesophageal echocardiography (TEE) that revealed an atrial septal defect (ASD) type ostium primum with left-to-right shunt, right chambers dilatation and mitral regurgitation (Video 1, 2). 3-dimensional (3D) transesophageal echocardiography was performed with Philips X7-2t probe and allowed us a correct 3D measurement of the defect size (2.1 x 2.3 cm) using Qlab10 software for post-processing imaging (Figure 1), the evaluation of a quadrileaflet mitral anatomy (Video 3, Figure 2) with a cleft in the anterior mitral leaflet (Video 4, figure 3) and another cleft in the posterior mitral leaflet (Video 5, figure 4). The valve appeared to be divided into four scallops A1-A2 P1-P2 as we can see from surgical view of mitral valve (Video 6).

Later on, the patient underwent surgical closure of Ostium Primum ASD by a patch and double cleft repair by suture after right heart catheterization.

Six months after the procedure, the patient reported significant improvement in her functional status and her shortness of breath improved considerably. TTE performed 6 months after the surgery documented a significant reduction in the right chambers dimensions.

Discussion

Partial atrioventricular canal (PAVC) defect is a form of endocardial cushion defect infrequently encountered in adults; few reported patients have survived into the sixth or seventh decade of life.¹⁻³ PAVC defect was defined as an ostium primum atrial septal defect but with two separate atrioventricular valve rings, a cleft in the anterior mitral leaflet and no appreciable interventricular communication. Failure of fusion of the leaflets derived from the superior and inferior cushions can result in clefting of the aortic leaflet subsequent to otherwise normal development; it is the latter arrangement that results in the trifoliate left component of the common valve found in the setting of AVSDs.⁴ Trileaflet mitral valves have only been recently reported in the literature as an extremely rare distinct disease entity.^{5,6} There are 6 published reports of 7 patients with trileaflet mitral valves, all of which were diagnosed by 3D echocardiography.⁷

We report an even more rare case where the clefts affect both the anterior mitral leaflet and the posterior mitral leaflet in a patient with ostium primum atrial septal defect turning the valve into a quadrileaflet valve.

Conclusion

The present clinical case highlights the importance of 3D TEE as the only tool for an accurate knowledge of the mitral valve anatomy and helping the surgeon in choosing the best mitral repair technique. In this specific case, the evidence of mitral regurgitation at the level of the clefts changed our decision towards the suturing of the same valve material and avoiding the use of annuloplasty.

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Figure/Video Legends

Video 1 ME 0° four-chambers view showing an ostium primum ASD and right chambers dilatation. ME =midesophageal; ASD= Atrial septal defect

Video 2 ME 0° four-chambers view showing left-to-right shunt and mitral regurgitation. ME = midesophageal

Video 3 3D TEE surgeon “en face” view showing clefts in the anterior and posterior mitral leaflets dividing the valve into four scallops. TEE= Transesophageal Echocardiography

Video 4 3D TEE view from LAA perspective focused on ostium primum ASD and AML cleft. LAA =Left atrial appendage; ASD = atrial septal defect; AML = Anterior mitral leaflet.

Video 5 3D TEE cropped on the posterior mitral leaflet showing cleft between P1-P2.

Video 6 3D TEE “en face” view of quadrileaflet mitral valve.

Figure 1 MPR of defect showing an ostium primum ASD with diameters 2.1 x 2.3 cm. MPR= MultiPlanarReconstruction; ASD = Atrial septal defect

Figure 2 “En face” view of the mitral valve from the left atrium. AV= Aortic valve; TV =tricuspid valve; LAA= left atrial appendage; ALC= anterolateral commissure; PMC= posteromedial commissure

Figure 3 Focused 3D image on the OP ASD and AML cleft. AO= aorta; OP ASD = ostium primum atrial septal defect; AML = Anterior mitral leaflet

Figure 4 3D shows another detail, a bilobate P2 scallop with an indentation between medial and lateral lobe (asterisks). From this view we can appreciate the main difference between cleft and indentation: cleft is deeper than indentation and reaches the posterior annulus.



