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Case Report

A Rare Case of Congenital Heart Disease with Bifid Cardiac Apex: A Unique Finding in Left Ventricle

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Abstract

Bifid cardiac apex is a rare anomaly of human hearts. We report of the case of a 34-year-old man with a previous history of ventricular septal defect (VSD) and subvalvular pulmonary stenosis. He had undergone pulmonary commissurotomy and VSD closure 22 years before he was referred to our center for evaluation of progressive dyspnea. Transthoracic echocardiography revealed atrial septal defect (ASD), multiple VSDs, severe pulmonary regurgitation, and a bifid cardiac apex. The patient was referred for re-do surgery for ASD and VSD closure along with pulmonary valve replacement, but he refused the surgery.

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Introduction

Bifid apex is rarely seen in otherwise normal human hearts or in association with congenital heart defects.[1] This malformation is seen in the normal hearts of sea mammals such as the whale, dugong, and manatee due to the possible need to delink the two ventricles for adaptation to the diving habits of these marine mammals and associated acute alterations in the pulmonary circulation. In human hearts, bifid apex can occur due to down-grade phylogenesis of the concerned genes.[1]

Case Report

The patient was a 34-year-old man, who had a history of previous cardiac corrective surgery due to ventricular septal defect (VSD) and subvalvular pulmonary stenosis about 22 years before he was referred to our center for evaluation of new-onset progressive dyspnea on exertion of 3 months' duration.

At presentation, he suffered from dyspnea on exertion (New York Heart Association [NYHA] function class II-III) and easy fatigability. On physical examination, a thrill in the left sternal border was palpable. Both heart sounds (S_1 and S_2) were diminished in intensity. There was diastolic murmur in the left second intercostals area, consistent with pulmonary regurgitation, and 4/6 holosystolic murmur best heard in the left sternal border, consistent with VSD. Electrocardiography showed atrial fibrillation rhythm with rapid ventricular response (heart rate about 110-120 beat/min), normal axis, right bundle branch block (RBBB) pattern, secondary ST-T

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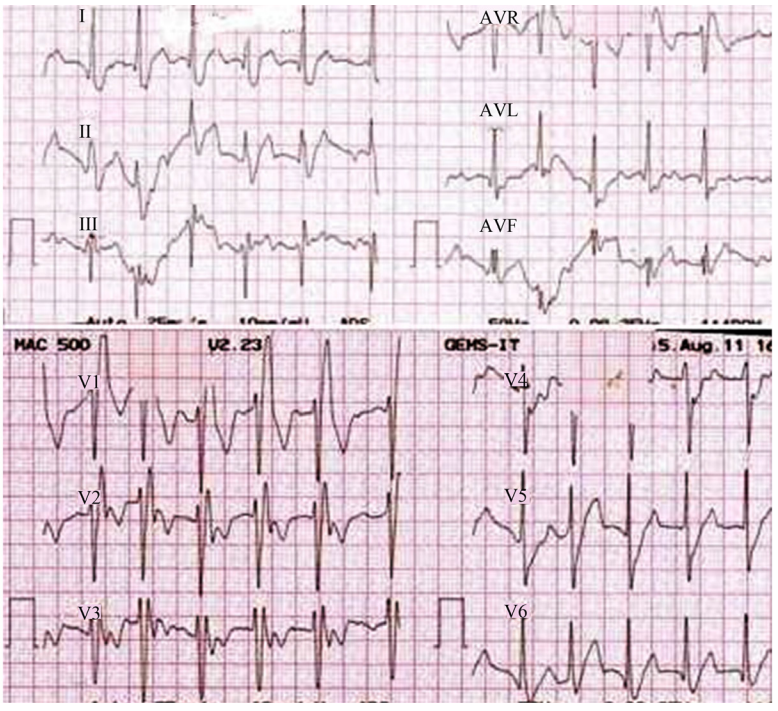

Figure 1. The twelve-lead electrocardiogram shows atrial fibrillation rhythm, normal axis, right bundle branch block pattern, secondary ST-T changes, and right ventricular hypertrophy

changes, and right ventricular hypertrophy (RVH) criteria (Figure 1).

Echocardiography revealed a normal left ventricle (LV) size with a mildly reduced LV ejection fraction (LVEF = 50%), as well as a large muscular ridge (3 × 1.5 cm) in the apical portion, highly suspicious of an abnormally located papillary muscle resulting in a double-orifice (bifid) apex (Figures 2 and 3). There was only a chordal attachment to the posteromedial papillary muscle, and no any chordal attachment to the anterior mitral leaflet was found.

There was severe right ventricular (RV) enlargement with moderate to severe systolic dysfunction and a large muscle (moderator) band in mid RV with moderate systolic obstruction by color flow Doppler. Other echocardiographic findings were mild mitral regurgitation, moderate tricuspid regurgitation, and moderate pulmonary artery hypertension (estimated systolic pulmonary artery pressure about 50-55 mmHg). The pulmonary valve leaflet was seen to flail, and there was severe pulmonary insufficiency without residual pulmonary stenosis.

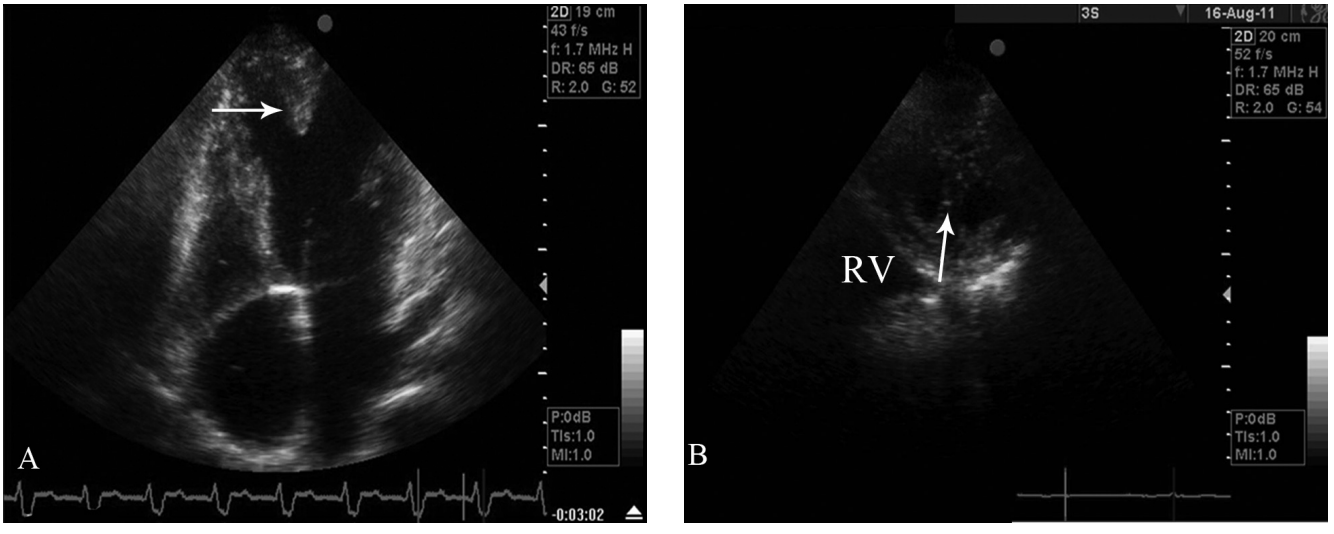

Figure 2. Apical four-chamber (A) and apical short-axis (B) echocardiographic views, revealing an abnormally located papillary muscle in the left ventricular apical portion (arrows)
RV, Right ventricle

A large residual VSD in both proximal and distal sides of the surgical patch was found; it resulted in LV-to-RV and LV-to-right atrium (RA) shunts (Gerbode defect). There were also an additional mid-muscular septum VSD (5.5 mm) and a large serpiginous apical VSD (15mm), which connected to the RV at the septal insertion of the moderator band with a left-to-right shunt (peak pressure gradient of 40 mm Hg). Another finding was a moderate-sized (10 mm) secundum type atrial septal defect (ASD) with a left-to-right shunt.

The patient underwent cardiac catheterization and O_2 saturation study. Oximetry showed a significant O_2 step-up at two levels in the low superior vena cava to the high RA, in favor of an atrial shunt through the ASD and in mid RA-to-RV, in favor of a ventricular shunt via the VSD with a significant left to right shunt in sum (pulmonary-to-systemic flow ratio [Qp/Qs] = 2.8), moderately increased pulmonary artery pressure (40/15 mmHg), and ventricularized pulmonary artery pressure tracing, in favor of severe pulmonary insufficiency.

Left ventriculography demonstrated multiple (VSDs) in mid septal and around the previous VSD patch, which immediately visualized the RV. Left ventriculography in right anterior oblique view showed a bifid apex (Figure 3) with mild mitral regurgitation.

The patient was referred for re-do surgery for ASD and VSD closure along with pulmonary valve replacement.

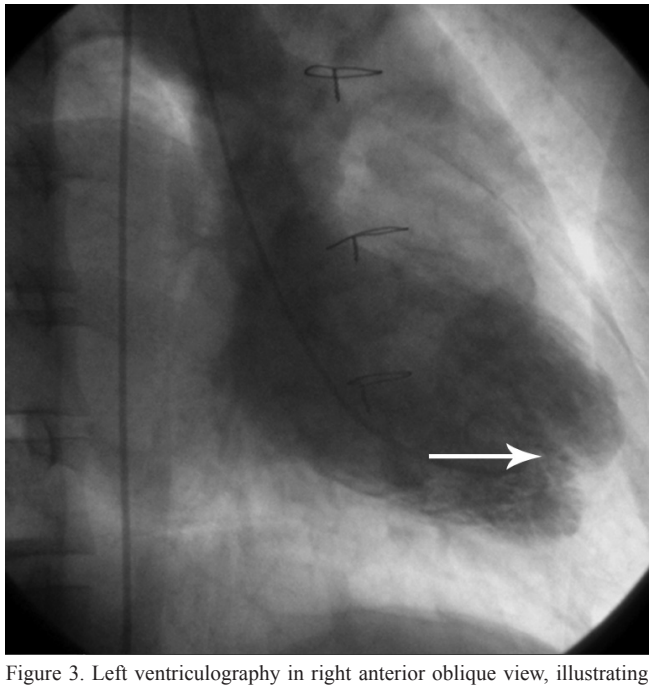

Figure 3. Left ventriculography in right anterior oblique view, illustrating an abnormally located papillary muscle in the left ventricular apex (arrow), resulting in a bifid apex

Discussion

Bifid apex is seen rarely in normal human hearts or in association with congenital heart defects.[2] The RV and LV develop independently on either side of the primitive plate before they combine[3] and, subsequently, muscle fibers produce a bridge across the ventricles. If the union of the two ventricles is disturbed at the apex (as was the case in our patient), a bifid apex in formed.

This rare unique morphologic anomaly has been described previously in 3 cases, associated with congenital heart disease.[1, 2, 4] The first case was a 3-year-old boy undergoing surgery for ASD.[2] The second case was an 11-year- old girl with bifid apex, persistent left superior vena cava, muscularized coronary sinus, bare atrioventricular cleft, bilateral hepatocardiac channels, and bull's horns in the RA appendage.[1] And the third case was a 17-year-old patient diagnosed with a large VSD, a small ASD, and a double-chambered RV.[4]

Conclusion

Bifid apex is rarely seen in normal human hearts in association with congenital heart disease. Echocardiography has an important role in the diagnosis of this unique anomaly of the cardiac apex and other associated congenital heart malformations.

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