Introduction

Ebstein’s anomaly is a rare congenital heart disease of the tricuspid valve which is characterized by the adherence of septal and posterior leaflets to the underlying myocardium, apical displacement of the functional annulus, dilatation of the atrialized portion of the right ventricle, redundancy, fenestrations, and tethering of the anterior leaflet, dilatation of the true tricuspid annulus (1, 2). Mitral valve anomalies are more common than expected in this congenital disease compared with the general population (3). Accessory mitral valve causing left ventricular outflow tract (LVOT) obstruction has also been reported in a patient with Ebstein’s anomaly (4). We present a case of Ebstein’s anomaly with LVOT obstruction due to native mitral valve which we think is first in the literature.

Case Report

A 42-year-old woman applied to our hospital with a exercise-induced syncope attack. We detected atrial fibrillation with a rapid ventricular rate. We learned from the history that Ebstein’s anomaly had been diagnosed and the atrial septal defect had been surgically closed 15 years ago. Any operative treatment for tricuspid valve had not been done. She had been experiencing palpitation, exercise dyspnea, dizziness with exercise for 10 years. Atrial fibrillation with a rapid ventricular rate had been detected one year ago and she had been using verapamil 240 mg per day for this reason. We stopped verapamil and administered amiodarone instead. Soon afterwards normal sinus rhythm was obtained.

On the physical examination, a grade 2/6 mid-systolic murmur over the aortic focus, a grade 3/6 pan-systolic murmur over the left sternal border and a grade 2/6 pan-systolic murmur over the apex of the heart were auscultated. The initial 12-lead electrocardiogram (ECG) showed atrial fibrillation, a short PR interval and the delta waves (Wolff-Parkinson-White syndrome). After the administration of the amiodarone therapy, the ECG showed normal sinus rhythm, a short PR interval and the delta waves (Wolff-Parkinson-White syndrome) for cardiac hydatid disease: an Anatolian Experience. Anadolu Kardiyol Derg 2003; 3: 238-44.


References

Ebstein’s anomaly. Mitral valve morphology, systolic anterior motion of the anterior mitral leaflet and mitral valve regurgitation may be seen, and any gradient of LVOT may be measured noninvasively with the use of echocardiography in such patients.

Video 1. Two-dimensional and color Doppler echocardiograms showing the mitral anterior leaflet causing left ventricular outflow tract obstruction

References

1. Edwards WD. Embryology and pathologic features of Ebstein’s anomaly. Prog Pediatr Cardiol 1993; 2: 5-15. [CrossRef]
4. Isobe M, Tanaka M, Sekiguchi M. Subaortic stenosis due to accessory tissue of the mitral valve associated with Ebstein’s anomaly in an adult. Int J Cardiol 1996; 57: 298-300. [CrossRef]

Discussion

Ebstein’s anomaly occurs in approximately 1 per 200,000 live births and accounting for <1% of all congenital heart diseases (5). The main finding of Ebstein’s anomaly is the downward displacement of the septal and posterior leaflets of the tricuspid valve in relation to the mitral anterior leaflet more than 8 mm/m² (6).

Ebstein’s anomaly is a disorder which is not confined to the right heart and left-sided valve abnormalities are also more common than the general population (3). Mitral valve prolapse (3), dysplasia of the mitral valve (3), anomalies of cords and papillary muscles (7) have been described in patients with Ebstein’s anomaly. Subaortic stenosis due to the presence of a accessory mitral valve has also been reported in an adult case of Ebstein’s anomaly (4).

This is the first case in the literature which reveal LVOT obstruction due to native mitral valve in an adult patient of Ebstein’s anomaly who had a surgical closure of atrial septal defect. The patient’s condition improved with the medical treatment and normal sinus rhythm was obtained. The signs of heart failure or cyanosis, and any recurrent syncope attack were not observed during the one year control of our case.

If exercise-induced syncope as a symptom and mid-systolic murmur heard over the aortic area as a physical examination finding exist in a patient with Ebstein’s anomaly, mitral valve tissue leading to LVOT obstruction should be kept in mind. Two-dimension and color Doppler echocardiogram may clearly demonstrate such finding and for this reason we did not use an additional imaging technique in our case. In patients with Ebstein’s anomaly who developed recurrent syncope attacks as a result of LVOT obstruction or heart failure due to severe mitral regurgitation caused by redundancy of the mitral leaflets and chordae, a reconstructive surgery for mitral valve leaflets and chordae may be considered.

Conclusion

Ebstein’s anomaly should not be considered as a disease limited to the right heart. Mitral valve anomalies are seen more frequently than normal population in this disorder. Echocardiography is a useful diagnostic tool in the evaluation of the mitral valve abnormalities related to

Dental volumetric tomography in the radiological detection of carotid artery calcification

Karotid arter kalsifikasyonunun radyolojik belirlenmesinde dental volumetrik tomografi

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Introduction

Atherosclerosis, a progressive inflammatory disorder, may lead to coronary heart disease (e.g., angina, myocardial infarction, and sudden death), cerebrovascular disease (e.g., stroke and transient ischemic attack) or peripheral vascular disease (e.g., claudication and critical limb ischemia). Atherosclerosis and its outcomes represent important