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CASE REPORT

Parachute mitral valve accompanied by bicuspid aortic valve on three-dimensional transesophageal echocardiography

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Abstract We report the findings of three-dimensional (3D) transesophageal echocardiography (TEE) in a patient with a parachute mitral valve (MV) accompanied by aortic valve (AV) malformation. The results indicated an enhanced echo in MV anterior leaves, incrassate, and shortened subvalvular chordae tendineae, and posteromedial papillary muscle that had echo reinforcement, calcification, retroposition, and a significant decrease compared with anterolateral papillary muscle. In addition, the anterolateral papillary muscle was huge, with the bilateral papillary muscles fused partly, and the posterior subvalvular chordae tendineae incrassate, shortened, and attached parachute-like to the anterolateral papillary muscle. The MV appeared dome-shaped for the open limit in diastole with an MV area of 1.6 cm. Moreover, the left ventricle increased in size and the bicuspid AV was malformed. Continuous wave Doppler angiograph showed that the flow rate increased to 398 cm/seconds at the AV orifice area. A 3D form of the MV structure was observed from the left ventricle using 3D-TEE inspection. The anterolateral papillary muscle was fused with its posteromedial homologue. The chordae tendineae was attached to the anterolateral papillary with the parachute-like structure, indicating dome movement.

Introduction

Congenital mitral valve (MV) malformation is a rare disease, with an incidence of 0.21–0.42% of all congenital heart diseases [1–3]. It is often accompanied by other congenital heart diseases and consists mainly of double orifice MV, simple anterior leaves cleft of MV, and parachute MV [4]. In this paper, the checked results of transthoracic echocardiography

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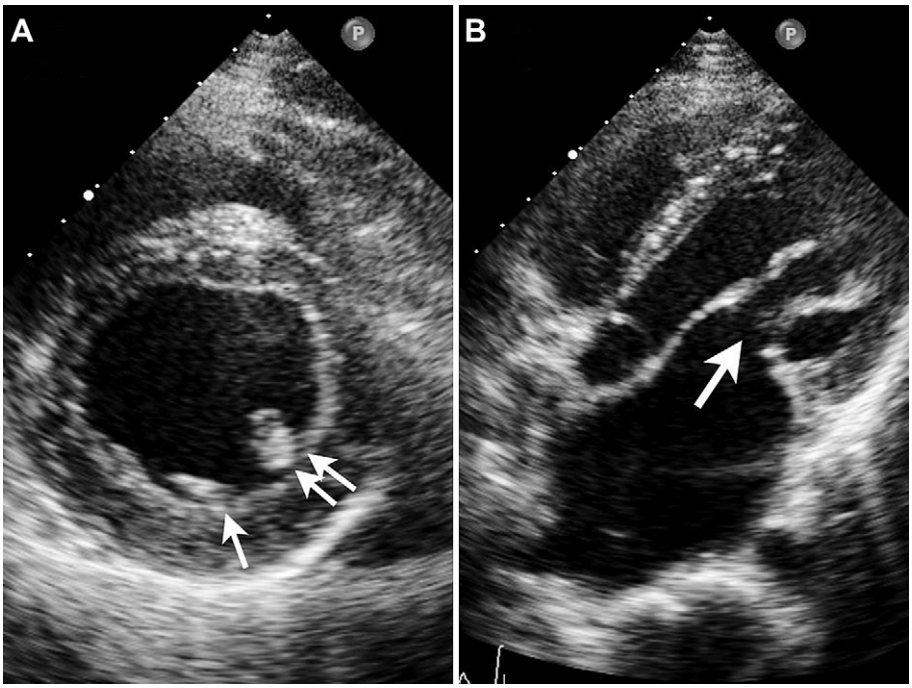

Figure 1. (A) A huge anterolateral papillary muscle was observed (double arrows), while the posteromedial papillary was smaller and closer to the anterolateral papillary muscle; (B) the chordae tendineae shortened and was attached parachute-like to the anterolateral papillary muscle (single arrow).

(TTE), transesophageal echocardiography (TEE) probe, and three-dimensional (3D) TEE from one case of parachute MV accompanied by bicuspid aortic valve (AV) malformations were shown and compared with the surgical results.

Case report

The patient was a man aged 23 years who presented with palpitation, weakness, and dyspnea after overwork, with increasingly aggravated symptoms. A holosystolic murmur-like blowing could be heard. Blood pressure was 123/74 mmHg. Electrocardiogram revealed left ventricular hypertrophy and mitral P wave. During radiography examination, the left ventricle and left atrium were obviously increased in size.

After hospitalization, TTE and 3D-TEE were performed on the patient. The results of TTE showed enhanced echo in the MV anterior leaves, subvalvular chordae tendineae became incrassate and shortened, and posteromedial papillary muscle had echo reinforcement, calcification, retroposition, and significant decrease compared with anterolateral papillary muscle. In addition, the anterolateral papillary muscle was large, the bilateral papillary muscles were partly fused, posterior subvalvular chordae tendineae was incrassate, shortened, and attached parachute-like to the anterolateral papillary muscle, and the MV appeared dome-shaped for an open limit in diastole with an MV area of 1.6 cm^2 (Fig. 1). The left ventricle had increased size and the bicuspid AV was malformed. Color Doppler flow imaging examination indicated that a mass of backflow signals (MV regurgitation grade 5) from the mitral orifice emerged along the mitral orifice between left ventricle and left atrium in systole. Continuous wave Doppler angiograph showed that the flow rate increased to 398 cm/seconds at the AV orifice area. The rate of antegrade blood flow of the MV was normal.

A 3D form of the MV structure was observed from the left ventricle through 3D-TEE inspection (Fig. 2). Anterolateral papillary muscle was fused with its posteromedial homologue. The chordae tendineae was sent forth from the anterolateral papillary with the parachute-like structure, indicating dome movement.

The diagnosis of congenital cardiac malformations and asymmetrical parachute MV malformation with AV bicuspid deformity was confirmed using ultrasonic cardiogram

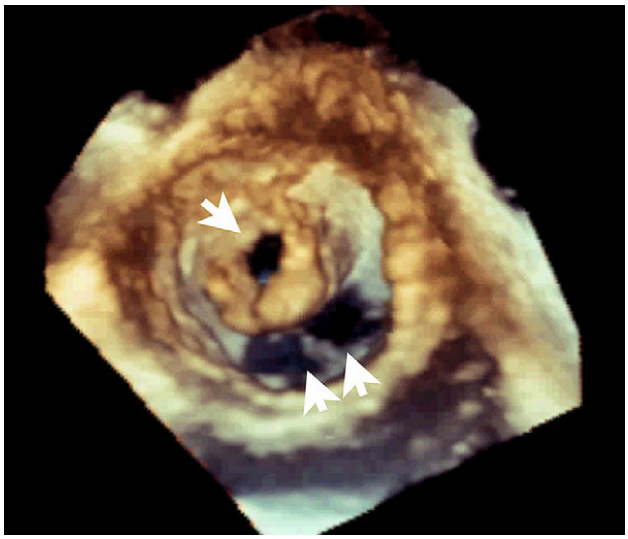

Figure 2. Transesophageal real-time three-dimensional echocardiography showing restricted mitral valve opening (single arrow) and bicuspid aortic valve (double arrows).

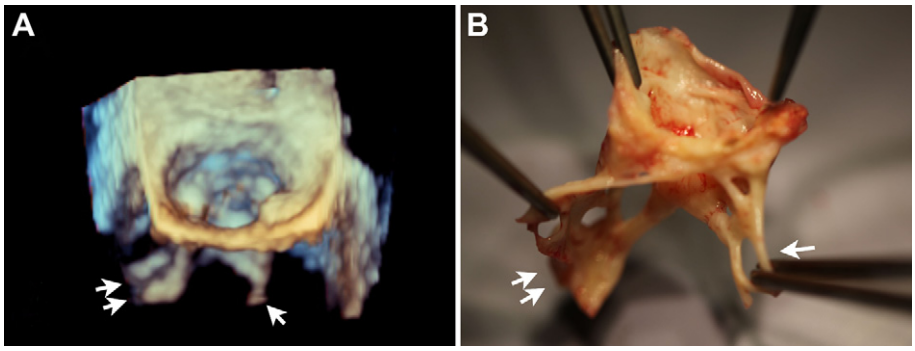

Figure 3. (A) Transesophageal real-time three-dimensional echocardiography; and (B) postoperative specimen showing that the valve edges of mitral valve anterior leaves were curly and stunted, whereas the chordae tendineae of the anterior leaves were shortened and fused (double arrows). The posteromedial papillary muscle was dysplastic.

inspection. Double-valve replacement was performed with AV and MV under conditions of general anesthesia and deep hypothermic cardiopulmonary bypass on September 8, 2010. Operative findings indicated that the valve edges of MV anterior-leaves were calcific, curly, and stunted, whereas the chordae tendineae of the anterior leaves were shortened. The posteromedial papillary muscle was dysplastic, had obvious retropositioning, and was partly fused with the anterolateral papillary muscle. The AV also had bicuspid deformity. Preoperative ultrasonic diagnosis was completely identical to the operative findings (Fig. 3).

Discussion

Normal MV papillary muscles are located at the mid-segment of the left ventricular wall and maintain an opposed position at a certain distance [5]. The MV bicuspid device consists of a valve ring, a valve leaflet, and the chordae tendineae together with papillary muscle [6]. A parachute MV is collectively defined by one of the following situations: Both mitral leaflets adhere to a group of thick papillary muscle, or they are affixed to two groups of thick papillary muscle due to partial fusion of a unilaterally hypogenetic papillary muscle to a contralateral papillary muscle [7]. The operated patient showed stunted and smallish MV anterior leaves, partly fused bilateral papillary muscle, and chordae tendineae that was almost entirely attached to the dominant papillary muscle anterolaterally, suggesting that this case belonged to the latter description and should thus be called "parachute-like asymmetric mitral valve" [8]. Parachute MV malformation is very rare and is often accompanied by other congenital heart diseases in which its chordae tendineae is pudgy and the activity of the valve leaflet is limited, leading to a decrease in effective orifice area and forming a funnel-shaped left ventricular inflow tract [9]. Therefore, the bloodstream reaches ventriculus sinister through the gap between the chordae tendineae, forming mitral incompetence or stenosis of mitral orifice accompanied by mitral incompetence. Despite having clear clinical symptoms, this disease can be easily misdiagnosed as rheumatic heart disease because of its combination with other congenital cardiac malformations in adult patients. However, it could be distinguished from rheumatic heart disease by carefully observing the subvalvular apparatus, particularly the papillary muscle and chordae tendineae using ultrasonic cardiography [10]. This case showed that an enlarged left ventricle and an incrassate left ventricular wall, secondary to the incorporative AV bicuspid deformity. Compared with two-dimensional echocardiography, the 3D-TEE provides excellent visualization of native pathologic MV apparatus components, as confirmed by surgical findings. Application of 3D-TEE offers 3D visual information of MV and AV orifice, which provided richer messages of MV, AV, and their surrounding structures to the clinician before operation. In addition, it supplies a reliable and elaborate basis for preoperative selection of surgical method. Therefore, a more direct, quicker, and more accurate diagnosis could be made of left heart valve malformation using both TTE and 3D-TEE.

References

[1] Collins-Nakai RL, Rosenthal A, Castaneda AR, Bernhard WF, Nadas AS. Congenital mitral stenosis. a review of 20 years'experience. Circulation 1977;56:1039—47.
[2] Carpentier A, Branchini B, Cour JC, Asfaou E, Villani M, Deloche A, et al. Congenital malformations of the mitral valve in children. Pathology and surgical treatment. J Thorac Cardiovasc Surg 1976;72:854—66.
[3] Gersony WM. Ventricular septal defect and left sided obstructive lesions in infants. Curr Opin Pediatr 1994;6:596—9.
[4] Ginghină C, Vlădaia A, Ghiorghiu I, Serban M, Popescu BA, Jurcuţ R. Echocardiography in congenital mitral valve regurgitation—the liaison between cardiologist and surgeon. J Med Life 2009;2:407—13.
[5] Hakim FA, Kendall CB, Alharthi M, Mancina JC, Tajik JA, Mookadam F. Parachute mitral valve in adults-a systematic overview. Echocardiography 2010;27:581—6.
[6] Marino BS, Kruge LE, Cho CJ, Tomlinson RS, Shera D, Weinberg PM, et al. Parachute mitral valve: morphologic descriptors, associated lesions, and outcomes after biventricular repair. J Thorac Cardiovasc Surg 2009;137:385—393.e4.
[7] Ucar O, Vural M, Cicekcioglu H, Pasaoglu L, Aydogdu S, Koparal S. Case report: Multidetector CT presentation of a parachute-like asymmetric mitral valve. Br J Radiol 2008;81:e266—8.
[8] Oosthoek PW, Wenink AC, Macedo AJ, Gittenberger-de Groot AC. The parachute-like asymmetric mitral valve and its two papillary muscles. J Thorac Cardiovasc Surg 1997;114:9—15.
[9] Erdogan O, Aktoz M. Parachute mitral valve abnormality and bicuspid aortic valve in an asymptomatic adult patient. Can J Cardiol 2008;24:e57.
[10] Purvis JA, Smyth S, Barr SH. Multi-modality imaging of an adult parachute mitral valve. J Am Soc Echocardiogr 2011;24:351 e1—3.