

Bicuspid aortic valve caused subaortic stenosis with bulging of valve calcification through subaortic area in a young patient

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February 16, 2023

Abstract

Bicuspid aortic valve is the most common congenital cause for the development of aortic valve calcification and stenosis. Calcification cause valvular stenosis or valvular insufficiency due to coaptation failure. We report a unique case of calcification of bicuspid valve was extending to left ventricular outflow tract and attached to interventricular septum which caused subvalvular stenosis.

Title page-Case Image

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Running title: Bicuspid Aorta and Subaortic Stenosis

Keywords: bicuspid aorta, calcification, surgery, subaortic stenosis

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Abstract

Bicuspid aortic valve is the most common congenital cause for the development of aortic valve calcification and stenosis. Calcification cause valvular stenosis or valvular insufficiency due to coaptation failure. We report a unique case of calcification of bicuspid valve was extending to left ventricular outflow tract and attached to interventricular septum which caused subvalvular stenosis.

Case Image

A 28-year-old man admitted our outpatient cardiology clinic with exertional dyspnea. He has been diagnosed with bicuspid aortic valve for five years ago, however, he did not go routine follow-up. Cardiac examination revealed regular pulse (90 beats/min) with normal blood pressure and a 3/6 systolic crescendo-decrescendo murmur loudest at the right upper sternal border. Electrocardiography revealed normal sinus rhythm with left ventricular hypertrophy. Transthoracic echocardiography showed bicuspid aorta with aortic aneurysm and severe aortic regurgitation. Interestingly, severe calcification of the valve was extending to subvalvular area and causing severe left ventricular outflow tract obstruction (subaortic stenosis)(Figure 1, Video 1). Transesophageal echocardiography and computed tomography revealed that calcification was starting from coronary cusp and extending to the interventricular septum and caused severe septal hypertrophy (Figure 2, Video 2). The patient underwent aortic valve replacement with mechanical prosthesis and thoracic aorta replacement with a Dacron graft after heart team discussion. The patient was well after surgery and discharged without any problem (Figure 3, Video 3).

BAV is the most common congenital cause for the development of aortic valve calcification ¹. Calcification of BAV has different genetic tendency in contrast to tricuspid valves². BAV mostly undergo strain, and wall shear stress on their edges of the leaflets during opening ³, however, in this case calcification started from annularly and extending to LVOT and basal septum. Besides the valve calcifications and stenosis, subaortic stenosis caused patient's symptoms.

Funding : None

Acknowledgement: Informed consent was obtained from the patient to use of images for academic purposes.

Conflict of interest : None

References

Figure Legends

Figure 1. (A)Parasternal long axis view of transthoracic echocardiography (TTE) showing calcification of aortic valve extending to the interventricular septum (arrows). (B) Continuous wave doppler of TTE showing severe gradient in left ventricular outflow tract (C) Apical view of TTE showing calcification. RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; Ao, aorta.

Figure 2. (A, B) Transesophageal echocardiography (TEE) revealing calcification (arrows) attached to the interventricular septum and causing subaortic stenosis (C,D) Computed tomography showing eccentric calcification of aortic valve. RA, right atrium; LA, left atrium; LV, left ventricle; Ao, aorta.

Figure 3. (A, B) Intraoperative view of aortic valve and partially resected calcification of interventricular septum. (C, D) Postoperative TTE showing mechanical aortic valve and diminished LVOT gradient. Video 2. TTE after operation. ; LA, left atrium; LV, left ventricle; Ao, aorta.

Video Legends

Video 1. Transthoracic echocardiography before surgery showing bicuspid aorta and huge calcification attached to septum

Video 2. Transesophageal echocardiography showing subvalvular stenosis and calcification.

Video 3. Transthoracic echocardiography after aortic valve replacement and correction of subvalvular stenosis with resection of calcification.



