A Calcified Unicommissural Unicuspid Aortic Valve: is it the trigger to the calcified amorphous tumor?

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Abstract

Unicommissural unicuspid aortic valve (UAV) is a rare valvular malformation. It may lead to severe aortic stenosis with or without aortic regurgitation. UAVs have many characteristic changes such as premature dysmorphic valvular calcification and premature valvular dysfunction. Transesophageal echocardiography (TEE) was described as the gold standard for the diagnosis of UAVs. We reported the case of a 58 years old man who presented with syncope and dyspnea due to severe aortic stenosis secondary to a heavily calcified unicommissural UAV. After the surgery pathological specimen showed severe amorphous calcification just like a calcified amorphous tumor (CAT).

Introduction

Isolated aortic valve stenosis in adults, with or without aortic regurgitation, is almost always secondary to a congenital valvular malformation.[1] Unicuspid aortic valve (UAV) is a rare form of these valvular malformations. The prevalence of which is 0.02% in the adult population.[2] Unicommissural UAVs generally present in the 4^{th} to 6^{th} decade of life.[5] UAVs have many characteristic changes such as premature dysmorphic valvular calcification, aortic root dilatations.[5] In valves, dysmorphic calcification may occur aggressively and may lead to dense calcified masses like a calcified amorphous tumor. Cardiac amorphous tumors (CAT) are rare non-neoplastic intracardiac tumors that consist of nodular calcium deposits in a background composed of fibrin or amorphous fibrillary materials.[6] Size can vary from small punctate lesions to very large masses, moreover diffuse myocardial infiltrations. In this report, we reported a 59 years old man who was diagnosed with severe aortic stenosis concomitant severe aortic insufficiency secondary to unicommissural UAV. After aortic valve and ascending aorta replacement, the surgical specimen revealed unicommissural unicuspid aortic valve with severe calcification. Histological examination revealed nodular calcification in the amorphous material, including dense fibrin and blood capillaries, confirming the diagnosis of CAT.

Case Report

A 58-year-old man was admitted to the emergency department because of syncope and progressive dyspnea for several months. A grade 5/6 systolic murmur and a grade 4/6 diastolic murmur were auscultated. Electrocardiography showed normal sinus rhythm. Brain computed tomography and diffusion magnetic resonance imaging was revealed normal for cerebrovascular pathologies. Computed tomography (CT) showed heavily calcified CAT like mass (Figure 1.A) on the aortic valve and aneurysm of the ascending aorta (5.2 cm in diameter). Transthoracic echocardiography (TTE) revealed concentric left ventricular hypertrophy with normal ejection fraction. The aortic valve was thickened and heavily calcified, especially in the middle portion. There was a fissure like opening with CAT like calcification on the aortic valve suggesting unicommissural

UAV. The maximal velocity through the aortic valve was 4.3 m/sec, the mean pressure gradient between the aorta and the aortic valve was 43 mm Hg and the calculated aortic valve area was 0.7 cm^2. There was concomitant severe aortic regurgitation with ascending aorta dilatation. Transesophageal echocardiography (TEE) confirmed unicommissural unicuspid aortic valve (Figure 1.B-C) and CAT like heavy calcification in the middle portion with substantial ascending aorta dilatation (Figure 2.A-B). Due to symptomatic severe aortic stenosis and aortic regurgitation with concomitant ascending aorta dilatation, the patient was consulted with the cardiovascular surgeons. The aortic valve and ascending aorta replacement were successfully performed. Postoperatively, the patient was transferred to the cardiac intensive care unit then discharged with warfarin in a week. Histopathologic examination showed calcified nodules on an amorphous background of fibrin material on the aortic valve just like a calcified amorphous tumor (Figure 3.A). And degeneration with cystic changes in the ascending aortic media which were described as cystic medial degeneration (Figure 3.B).

Discussion

Unicommissural unicuspid aortic valve (UAV) is a rare valvular malformation which prevalence is %0.02 in the adult population.[2] It may clinically present with aortic stenosis, aortic regurgitation, or aortic root dilations.[5] Premature dysmorphic valvular calcification on the valves is one of the characteristic changes in UAVs. UAV patients generally show symptoms earlier in their lives, usually in 30-50 years.[5] Premature dysmorphic calcification and valvular dysfunction develop earlier and progress faster.[5] Our patient was diagnosed with unicommissural UAV, severe aortic stenosis concomitant severe aortic regurgitation by TEE which was described as the gold standard for diagnosis UAVs.[7] Ascending aortic dilatation with mild histological changes in media has been investigated in UAV patients in some studies. [2] Ascending aortic dilatation which was measured 5.2 cm in diameter. Heavily calcified CAT-like mass was revealed on the valve both CT and TTE. CAT is a rare non-neoplastic intracardiac tumor and mostly concomitant with pre-existing valve disease or end-stage renal disease. They can be small punctuate lesions to very large masses. In our patient, severe aortic stenosis with unicomissural UAV morphology might lead to heavy calcification like CAT especially in the middle portion of the aortic valve. Stenotic valve area might lead to earlier valvular dysfunction and premature dysmorphic calcification on the valve. In histopathological examination showed densely calcified nodules on an amorphous background of fibrin deposits that were also similar findings in CATs. Also degeneration with cystic changes in the ascending aortic media which were described as cystic medial degeneration.

Conclusion

Unicommissural UAV is a rare congenital disorder that leads to severe aortic stenosis with or without aortic regurgitation. It can be accompanied by aortic dilatation. Valvular dysfunction and dysmorphic calcification develop earlier and calcification on the valve can mimic cardiac amorphous tumors. In young patients with heart failure symptoms and systolic murmurs, it must be considered in the differential diagnosis. Deciding the appropriate time for surgery, it is important to diagnose and follow up earlier these patients.

Ethics Statement

The patient has provided written, informed consent to have the details of his case published. Institutional approval was not required for publication.

Conflict of interests

The authors report no conflicts of interest in this work.

Author Contributions

SU, CG, and EC contributed to the drafting of this manuscript. GG contributed to pathologically diagnosis and discussion

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Supplementary Material:

Movie Clip 1. Unicuspid unicommissural aortic valve motion in systole and diastole at TEE. LA: Left atrium, RA: Right atrium, RVOT: Right ventricular outflow tract.

Movie Clip 2. 3D unicuspid unicommissural aortic valve motion in systole and diastole at TEE. LA: Left atrium, RVOT: Right ventricular outflow tract.

Figure Legends:

Figure 1. A- Heavily calcified mass (1.14x1.63cm) on the aortic valve (yellow arrow) and ascending aorta dilatation (yellow star) at CT. B- Opened unicuspid unicommissural aortic valve (yellow arrow) in systole at TEE. C- Closed unicuspid unicommissural aortic valve (yellow arrow) in diastole at TEE. Ao: Aorta, Pa: Pulmonary artery, LA: Left atrium, RA: Right atrium, LV: Left ventricle. RVOT: Right ventricular outflow tract.

Figure 2. A- 3D opened unicuspid unicommissural aortic valve in systole at TEE and heavily calcified mass on the valve (yellow arrow) B- 3D closed unicuspid unicommissural aortic valve (yellow arrow) in diastole. LA: Left atrium, Pa: Pulmonary artery, RVOT: Right ventricular outflow tract.

Figure 3. A- The histopathologic examination showed calcified nodules (yellow arrows) on an amorphous background of fibrin material. B- The slide showed degeneration with cystic changes (yellow stars); loss of elastic and muscle fibers in the ascending aortic media. These histopathologic features were described as cystic medial degeneration.