Recurrent Aortic Coarctation with Aneurysmatic Thinning of the Subaortic Basal Ventricular Septum in a Young Man

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Cardiac multimodality imaging is being used more frequently, and especially in some patients, it helps to make a better diagnosis and is plays an important role in the choice of treatment. In our case, we would like to present a young male with recurrent aortic coarctation, bicuspid aorta, severe aortic regurgitation, and ventricular septal aneurysm. Written informed consent was obtained from the patient who participated in this study.

A 34-year-old male presented to outpatient department with complaints of ongoing exertional dyspnea with New York Heart Association class II functional status. His medical history was remarkable with an aortic coarctation accompanied by bicuspid aorta and hypertension that was treated with percutaneous stenting method 11 years ago. On evaluation, his physical examination revealed a normal S1 and S2 with an S3 and holosystolic murmur at the aortic area, while his lung fields were clear on auscultation. His heart rate was normal (70/min) and regular. His blood pressure (BP) in his right upper extremity was 140/55 mmHg, while BP in his right leg was 125/50 mmHg.

Transthoracic echocardiography demonstrated a dilated left ventricle with an ejection fraction (EF) of approximately 45%, bicuspid aorta with a severe, very eccentric aortic regurgitation. Given to his poor echogenicity, we decided to perform a cardiac MRI to assess the both left ventricular function, aortic regurgitation, and also recurrent aortic coarctation at the stent site, which is suspected by blood pressure difference between left and right upper extremities.

Cardiac magnetic resonance imaging (CMRI) demonstrated mild to moderate left ventricular global systolic dysfunction (LVEF 42%) with severely increased LV indexed volumes and secondary increased trabeculation in the left ventricle (Figure 1). A distinctive finding was aneurysmatic thinning of the subaortic ventricular basal septum. Aortic valve was a Sievert type 0 bicuspid valve (Figure 2) with a severe, eccentric aortic regurgitation (regurgitant fraction 46%). Aortic root and ascending aorta were mildly dilated. Tubular aorta assessment showed that mid-stent portion (Figure 3) (Figure 4) which is corresponding to the mid descending aorta was mild-moderately narrowed (14.3 mm). Coarctation minimum area in the mid stent segment was 150 mm^2, 63 mm^2/m^2 and coarctation diameter/descending aorta ratio was 0.75 (14.3 mm/19 mm) suggestive of mild-to-moderate stenosis in the stent. Peak velocity in the stent was 272 cm/s. Late images after gadolinium showed a midmyocardial late gadolinium enhancement in the mid-cavity inferior segment. Those findings were consistent with a bicuspid aorta with severe very eccentric aortic regurgitation resulting in basal ventricular septal aneurysm and left ventricular systolic dysfunction accompanied by a recurrent, mild to moderately stenosed stent in the aortic coarctation site.

As he had been a smoker for the last 17 years (1 pack/day) with a family history and dyslipidemia risk factors for coronary artery disease, we performed a coronary computerized tomography angiogram (CCTA) to check the coronary artery system. Coronary arteries were normal and basal ventricular septal aneurysm was more remarkable on CCTA images as the resolution was better than echocardiography and CMRI. Three-dimensional volume rendering images were quite helpful for the orientation of the surgeon.

Our heart team decided on aortic valve surgery with or without interventricular aneurysm repair and follow up of mild-moderate recurrent stent restenosis at the coarctation site. However, cardiovascular surgeon has wished and suggested not to repair the ventricular septal aneurysm, as this would prolong the duration of surgery which can jeopardize the left ventricle even more, which has a low cardiac reserve with an EF of 42% with no significant

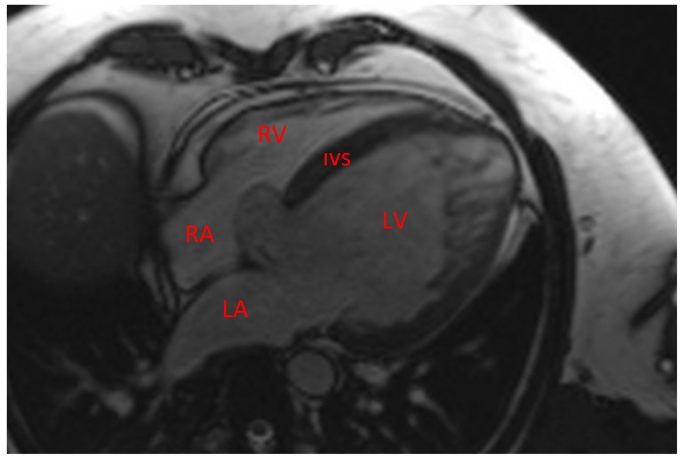

Figure 1. Cardiac magnetic resonance imaging, 4-chamber view. LV, left ventricle; RV, right ventricle; LA, left atrium; RA, right atrium; IVS, interventricular septum.

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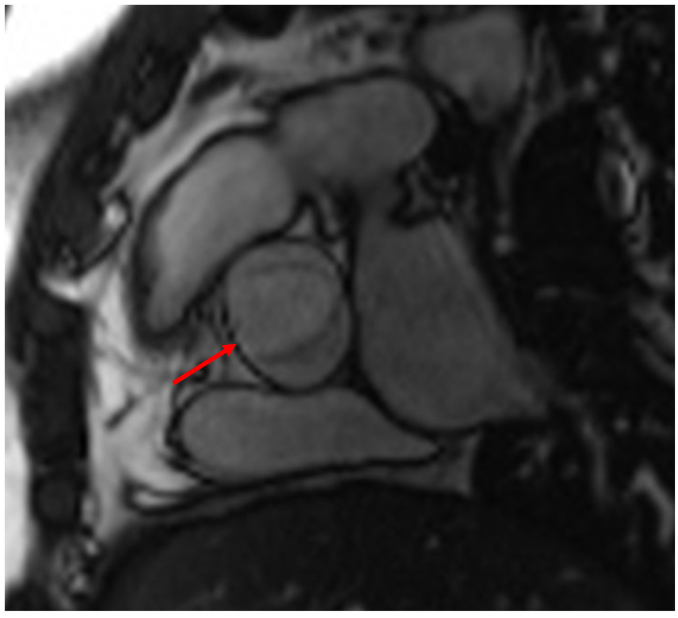

Figure 2. Cardiac magnetic resonance imaging, short axis view. Red arrow indicates aortic valve, which is clearly seen as bicuspid.

additional benefit top the patient in point to absence of clear indication.

Regrettably, the patient refused the surgery at the end and wanted to be followed up till he feels very symptomatic despite clear explanation of the heart team regarding the risks of waiting for a surgery at that point.

Aortic coarctation is first described by Morgagni in 1760 and causes localized or tubular narrowing in aorta and in some cases, aortic interruption.[1] In most cases, arterial hypertension accompanies due to decreased aortic compliance as the coarctation of aorta is characterized by a generalized arteriopathy and consists even after percutaneous or surgical treatment of aortic coarctation.[2]

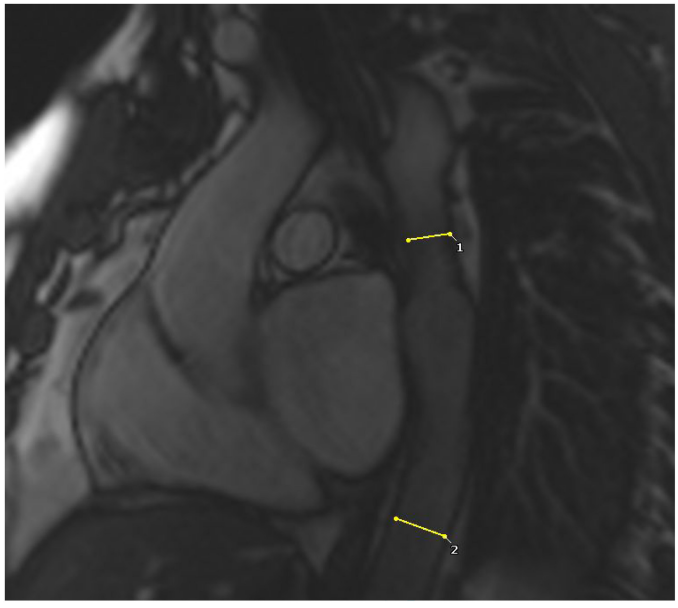

Figure 3. Magnetic resonance imaging, thoracic aorta. At level 1, diameter is 14.34 mm and at level 2 17.51 mm.

Figure 4. Computer tomography imaging of the previously stented area (red circle).

The decision of treatment—either surgical or percutaneous—is based on blood pressure, gradient, stenosis morphology. As European guidelines suggest, repair of coarctation—or recoarctation—is indicated in hypertensive patients with an increased non-invasive gradient between upper and lower limbs and confirmed with invasive measurement (peak-to-peak ≥ 20 mmHg) with preference for catheter-based treatment (stenting) when technically feasible with a class I indication. Even if the invasive peak-to-peak gradient is <20 mmHg, catheter-based treatment should still be considered in hypertensive patients with ≥50% narrowing relative to the aortic diameter at the diaphragm with class IIa indication. In normotensive patients, those class of recommendations decreases 1° (IIa for the first group and IIb for the second group).[3]

An important fact that also should not be forgotten is diagnosing hypertension in such patients based on right arm measurements, and right arm ambulatory blood pressure monitoring should be considered for the diagnosis of hypertension.

Bicuspid aortic valve occurs in 1%-2% of the population with a male predominance of approximately 3 : 1 and consistency with aortic coarctation is not seen rare. Further, in the patients with bicuspid aortic valve, accompanying aortic coarctation is an independent risk factor for aortic dissection.[4] Cardiac MRI is advantageous for imaging aortic coarctation as it can directly demonstrate an associated bicuspid aortic valve, any accompanying valve dysfunction, and secondary effects upon the left ventricle such as concentric hypertrophy or left ventricular dilatation.

In the rare occasion that a ventricular septal aneurysm requires surgical intervention, it is worth noting that surgical treatment can result in tricuspid regurgitation or even complete heart block because of the proximity of the septal leaflet of the tricuspid valve and the atrioventricular node to the membranous ventricular septum.

In the era of multimodality imaging, magnetic resonance imaging and cardiac computer tomography are being used more frequently day by day. Due to poor echogenic window in some cases, decision making only by transthoracic echocardiography would be inappropriate and additional imaging methods may be needed. In our case, we would like to make a point to importance of multimodality imaging.

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References

1. Sinha SN, Kardatzke ML, Cole RB, Muster AJ, Wessel HU, Paul MH. Coarctation of the aorta in infancy. *Circulation*. 1969;40(3):385-398. [CrossRef]
2. Choudhary P, Canniffe C, Jackson DJ, Tanous D, Walsh K, Celermajer DS. Late outcomes in adults with coarctation of the aorta. *Heart*. 2015;101(15):1190-1195. [CrossRef]
3. Baumgartner H, De Backer J, Babu-Narayan SV, et al. ESC Guidelines for the management of adult congenital heart disease: the Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC). Endorsed by: association for European paediatric and congenital cardiology (AEPC), International Society for Adult Congenital Heart Disease (ISACHD). *Eur Heart J*. 2021;42(6):563-645. [CrossRef]
4. Oliver JM, Alonso-Gonzalez R, Gonzalez AE, et al. Risk of aortic root or ascending aorta complications in patients with bicuspid aortic valve with and without coarctation of the aora. *Am J Cardiol*. 2009;104(7):1001-1006. [CrossRef]