

A Bicuspid Aortic Valve in a Patient with Infective Endocarditis

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Abstract

Bicuspid aortic valve is the most common congenital heart anomaly, present between 1 and 2 percent of the general population. We report a case of a 34-year-old man admitted with acute endocarditis involving the tricuspid valve, an incidental Bicuspid Aortic Valve, and a ventricular septal defect that was the most likely source of the endocarditis.

Keywords: Bicuspid Aortic Valve; Infective Endocarditis; Congenital Heart Disease; Aortic Valve; Valvular Heart Disease

History of Presentation

A 34-year-old male presents to the emergency department with a four-day history of diarrhea, generalized myalgias and subjective fever. He endorsed occasional intranasal cocaine use, most recently 3 days before the onset of symptoms. Upon arrival he was diaphoretic, his heart rate was 120 beats per minute and his temperature was 39.4 °C (102.9 °F). Physical exam revealed a 4/6 systolic murmur over the left third intercostal space and left parasternal line as well as purplish tender lesions on the left first toe and the right third digit. Initial laboratory tests were remarkable for leukopenia, acute kidney injury, and lactic acidosis.

Investigations

An electrocardiogram showed sinus tachycardia. A transthoracic echocardiogram (TTE) showed a BAV with an anterior and posterior cusp [Figure 1], vegetations on the anterior leaflet of the tricuspid valve with mobile components, and an infected membranous Ventricular septal defect (VSD) with a left to right flow (peak velocity was 4.6m/s). A Transesophageal echocardiogram (TEE) confirmed the above findings and better defined the vegetations (the largest was 16mm in size) [Figure 2&3]. Blood cultures were positive for Methicillin-Sensitive *Staphylococcus Aureus*.

Management

He was started on antibiotic therapy. Although blood cultures became negative after 8 days of antibiotics, fever persisted after 2 weeks, which was concerning for embolic phenomena. Computed tomography showed left frontal and left occipital lobe infarcts, bilateral pulmonary septic emboli, and splenic infarcts. Despite

the above findings the patient did not have any neurological symptoms and did not complain of cough, abdominal pain, or hemoptysis. A repeat TEE showed expansion of the infected tissue into the base of the Aortic Valve. Cardiothoracic surgery was consulted. He underwent a tricuspid annuloplasty and leaflet reconstruction with patch, ventricular septal defect patch closure, and mechanical aortic valve replacement using a 25mm On-X valve. There was evidence of myxoid degeneration, giant cells, and acute and chronic endocarditis in the aortic valve on tissue microscopy.

Post-operative course was complicated by multiple episodes of sinus pauses requiring a transvenous pacemaker insertion for 72 hours. A repeat TEE showed a reduced left ventricular ejection fraction of 40%. No vegetations were seen. He was discharged on intravenous antibiotics to complete 8 weeks of treatment, warfarin for valvular thromboembolism prevention, angiotensin-converting enzyme inhibitors and a beta-blocker for heart failure.

Follow-Up

The patient has been compliant with outpatient follow-up and medication adherence. 6 months after the surgery his functional status has returned to baseline.

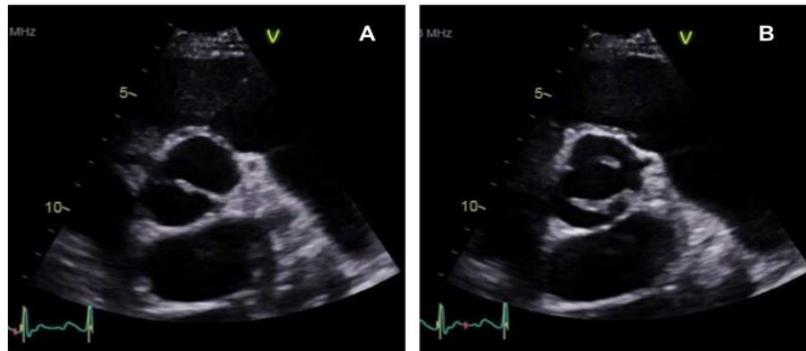


Figure 1: Transthoracic Echocardiogram: Parasternal Short Axis view of the Aortic Valve. [A] at end of diastole [B] at the end of systole. Notice it has 2 functional cusps instead of three. The valve commissures run from a 9 o' clock to a 3 o' clock position and are horizontal in orientation. The right and left coronary cusps are conjoined and there is no fibrous tissue (raphe) separating them.

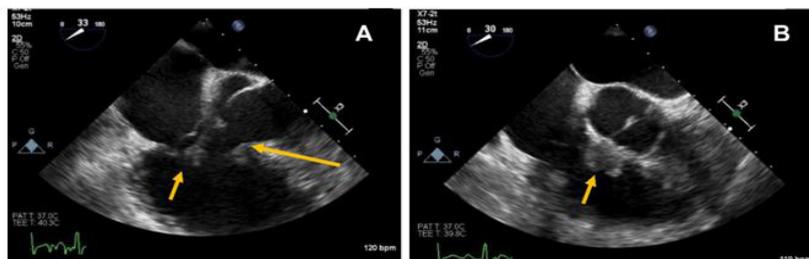


Figure 2: Transesophageal Echocardiography: Mid-esophageal short axis view. [A] The Tricuspid Valve is closed. Notice the vegetations (short arrow) and the peri membranous VSD(long arrow). [B] The Tricuspid Valve is open and the infected aneurysmal component of the VSD is visible (arrow).

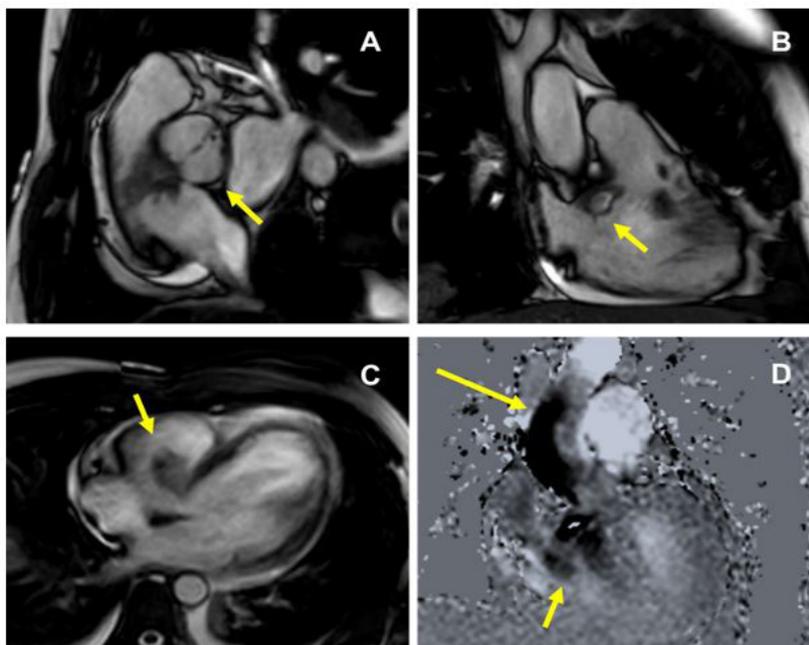


Figure 3: Cardiac Magnetic Resonance:[A] Sagittal view showing the BAV. [B] Coronal view showing the VSD (arrow). [C] Transverse view on diastole showing vegetations on the Tricuspid Valve (arrow). [D] Phase-contrast during systole showing blood flowing towards the Aorta (long arrow) and through the VSD into the Right Ventricle (short arrow)

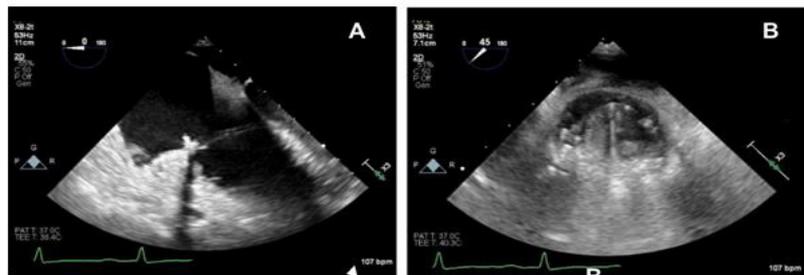


Figure 4: Post-operative Transesophageal Echocardiogram:[A] Tricuspid Valve. [B] Aortic Valve replaced by On-X valve with end-diastolic leaflet coaptation.

Discussion

Bicuspid aortic valve (BAV) is the most common congenital heart disease. It is present in between 1 and 2 percent of the general population, affecting 3 males for every female. It occurs as the result of the failure of the cusps to split after the Truncus Arteriosus separates into the Aorta and the Pulmonary arteries during development [1]. Consequently, the Aortic valve will have two functional cusps instead of three[2].

The most common fused cusps are the left and right coronary in approximately 80-86% of the cases, followed by the fusion of the right and left coronary cusps in 12-15%. A raphe is present in 75% of all patients with BAV[3].

The BAV can be classified according to the presence of raphe or not, the cusps that are conjoined or the spatial orientation of the commissures in the circumference of the aorta.

This patient had a Type 0 Anteroposterior BAV, which is seen in less than 5% of all patients with BAV. On this type of the fusion of the right and left coronary cusps is seamless and has no raphe, producing a valve with an anterior (the conjoined right and left) and posterior (non-coronary) cusps[4]. The commissures are oriented horizontally, running from a 9 o' clock to 3 o' clock position.

Patients with BAV are at high risk for developing complications later in life, as the result of turbulent flow caused by the defective aortic valve itself or by associated congenital heart defects, like in our case, where a ventricular septal defect was also present.

Furthermore, damage of the endothelium and connective tissue of the endocardium can be accentuated by high-risk behaviors such as substance abuse. The sympathomimetic effects of cocaine may increase the velocity of the jet by increasing heart rate and increased myocardial contractility [5].

The clinical outcomes of a patient with BAV can be variable from one individual to another. Some patients can reach adulthood without any symptoms and get a diagnosis incidentally based on a murmur during a physical exam or during a routine echocardiogram. Other patients can develop complications during infancy or early childhood.

Endocarditis of a morphologically normal tricuspid valve rarely presents in an immunocompetent patient without a history of intravenous drug use. This patient did not have an isolated right valvular endocarditis but rather a ventricular septal defect with a left to right flow that served as a nidus and a source for bacterial growth which then spread to the tricuspid valve and later to the aortic valve. This phenomenon has been described in several case reports [6&7] where a patient with congenital heart disease develops infective endocarditis in a morphologically normal valve.

Patients with infective endocarditis require surgery when there is persistent fever or bacteremia 10 days after appropriate therapy or when there are vegetations over 10mm in diameter such as in this case[8]. However, there are no trials assessing the effects of early surgery on morbidity and mortality.

References

1. Chan Kwan Leung, Veinot John P. Anatomic basis of echocardiographic diagnosis. 1st ed. London: Springer; 2011. doi:10.1007/978-1-84996-387-9
2. Rajamannan NM. Cardiac valvular medicine. 1st ed. London: Springer; 2013. doi:10.1007/978-1-4471-4132-7
3. Hoffman JIE. The natural and unnatural history of congenital heart disease. Wiley-Blackwell; 2009.
4. Hans H Sievers , Claudia Schmidtke. A classification system for the bicuspid aortic valve from 304 surgical specimens. J Thorac Cardiovasc Surg. 2007; 133(5): 1226-33. doi: 10.1016/j.jtcvs.2007.01.039
5. Sung Tae Kim, Taehwan Park. Acute and chronic effects of cocaine on cardiovascular health. Int J Mol Sci. 2019; 20(3): 584. doi: 10.3390/ijms20030584.
6. I Spyridopoulos, U Helber, C Mewis, W Voelker, B Steinhilber, H J Schulze, et al. Tricuspid valve endocarditis due to a jet lesion detected by echocardiography in a 27-year old man with congenital ventricular septal defect. J Cardiovasc Surg (Torino). 1996; 37(5): 517-520.
7. Woo Shin Kim, Seok Hyung Kang, Shin A Lee, Min Sun Ryu, Seong Hoon Park. A case of staphylococcal tricuspid valve endocarditis with para-aortic abscess in a patient with bicuspid aortic valve. Korean Circ J. 2011; 41(8): 482-485. doi: 10.4070/kcj.2011.41.8.482
8. DukHyunKang. Timing of surgery in infective endocarditis. Heart. 2015; 101(22): 1786-1791. doi: 10.1136/heartjnl-2015-307878