CLINICAL VIGNETTE

Bicuspid Aortic Valve Infective Endocarditis: Is There a Need to Widen the Indications for Antibiotic Prophylaxis

Ramtin Anousheh, MD, MPH and Ramona Mehrinfar-Zadeh, MD

Introduction

Bicuspid aortic valve (BAV), is the most common congenital cardiac valvular disease with prevalence ranging from 0.5% to 2%.[1] It increases the risk of infective endocarditis, thoracic aortic aneurysm and dissection, and sudden cardiac death.[2] Patients with bicuspid aortic valves do not routinely need antibiotics before dental and surgical procedures and currently the only indication for prophylaxisis is prior history of endocarditis.[3] We present a case of sub-acute infective endocarditis in an otherwise healthy female.

Case Presentation

A 51-year-old female initially presented to primary care clinic with dizziness and feeling unwell. She had prior diagnosis of acute sinusitis, documented on MR and had received several courses of antibiotics. Her symptoms did not improve over three months and she developed anemia, 30 lbs unintentional weight loss, intermittent palpitations and visual disturbances prompting multispecialty referrals including cardiology. At her outpatient cardiology evaluation a loud diastolic murmur was detected and a transthoracic echocardiogram (TTE) showed a large vegetation on a bicuspid aortic valve with moderate to severe aortic regurgitation with preserved left ventricular dimensions and function.

She was admitted to the hospital and diagnosed with culture-negative infective endocarditis and treated with six-weeks of intravenous vancomycin. Follow up TEE showed substantial decrease in the size of vegetation, ventricular dilatation and decreased systolic function. She subsequently underwent aortic valve replacement and repair of thoracic aortic aneurysm.

Discussion

Bicuspid aortic valve is primarily inherited in an autosomal-dominant pattern. Studies have found a 15% rate of familial clustering.[4] In a study of 142 patients with bicuspid aortic valve, 20% of first-degree relatives had some cardiac abnormality found on screening, of whom 68% had bicuspid aortic valve. Of these, 71% were newly detected abnormalities.[5]

A systolic ejection click is followed by an early peaking systolic murmur, most commonly at the apex. Bicuspid valves are typically initially asymptomatic but are commonly associated with progressive valvulopathy and aortopathy.

Sabet et al studied 542 patients with congenital bicuspid aortic valves undergoing aortic valve replacement. Seventy-five percent of the patients had isolated aortic stenosis, 10% had aortic stenosis with some degree of aortic insufficiency, and 13% had isolated aortic insufficiency. Increased risk of aneurysmal dilatation, coarctation, and dissection of thoracic aorta was also found.[2]

Several observational studies have shown that the patients with a native BAV are at markedly increased risk of aortic valve endocarditis compared with endocarditis of other native heart valves. The relative risk of IE of a BAV was markedly greater than for a tricuspid aortic valve. Recent estimates of the incidence of infective endocarditis (IE) occurring in BAV patients are 2% or 0.3%/year.[6]

The etiology of BAV association with increased frequency of IE is not established. Altered flow patterns across the BAV can result in valvular endothelial injury, including platelet and fibrinogen deposition. This has been postulated to allow seeding of hematogenous bacteria and fungi.[7]

Older age, intravenous drug abuse and multiple comorbidities including end-stage renal disease and indwelling cardiac devices are independent risk factors for endocarditis in patients with BAV.

In the current era, TTE can usually confirm the diagnosis of IE. If there is uncertainty in the diagnosis, TEE can improve the sensitivity and specificity. Due to the high rate of negative blood cultures caused by empiric antibacterial therapy administered prior to the hospitalization in United States, the role of TTE or TEE is particularly important.

For aortic valve IE, surgeons should excise the diseased aortic valve, and clear calcified plaque and infective tissues. Presence of peri-valvular abscesses increases the technical difficulties and risks of surgical treatment.

Current guidelines do not recommend prophylactic antibiotics in patients with BAV undergoing procedures prone to bacteremia. As the majority of patients with BAV are unaware of their BAV status until the occurrence of significant morbidity, estimating the risk of IE in patients with BAV, compared with individuals with tricuspid valves is challenging.

Randomized trials of prophylactic antibiotics in patients with BAV undergoing procedures prone to bacteremia have potential value.

Considering the overwhelming evidence based on multiple published case reports and series, we recommend antibiotic prophy-laxis prior to high risk procedures in patients with known structurally or functionally abnormal bicuspid aortic valves.

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