

Bicuspid Aortic Valve and Unruptured Asymptomatic Aneurysm of Sinus Valsalva in a 67-Year-Old Patient: Case Report

Altmış Yedi Yaşında İki Yapraklı Aort Kapağı ve Belirti Vermeyen Yırtılmamış Sinüs Valsalva Anevrizması Bulunan Olgu

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ABSTRACT Sinus valsalva aneurysm is a rare clinical condition which is mostly congenital. Association of sinus valsalva aneurysms with other cardiac pathologies can be seen, however, association of sinus valsalva aneurysm with a bicuspid aortic valve is rare. Complicated (symptomatic and/or ruptured) sinus valsalva aneurysms; are diagnosed early with ease with their associated arrhythmic, embolic and coronary complications, on the other hand, diagnosis is difficult in asymptomatic cases and in these individuals, transthoracic echocardiography is an important modality. Sinus valsalva aneurysms, especially when associated with bicuspid aortic valve, are generally diagnosed earlier. In this case report we present a 67-year-old patient with an asymptomatic and unruptured aneurysm of sinus valsalva associated with bicuspid aortic valve.

Key Words: Sinus of valsalva; aortic aneurysm; aortic valve; echocardiography; tomography, X-ray computed

ÖZET Sinüs valsalva anevrizması nadir görülen, çoğunlukla konjenital olan bir klinik durumdur. Sinüs valsalva anevrizmaları diğer kardiyak patolojilerle birlikte görülebilir, ancak iki yapraklı aort kapağı ve sinüs valsalva anevrizması birlikteliği nadir görülür. Komplike (sempptomatik ve/veya yırtılmış) sinüs valsalva anevrizmaları; aritmik, embolik ve koroner komplikasyonları nedeni ile kolay ve erken tanı alır ancak asemptomatik vakalarda tanı zordur ve bu hastalarda tanıda transtorasik ekokardiyografinin yeri oldukça önemlidir. Sinüs valsalva anevrizmaları, özellikle iki yapraklı aort kapağı ile ilişkili olduklarında, genellikle erken yaşta tanı alırlar. Bu olgu raporunda 67 yaşında, bicuspid aortik kapak ile ilişkili asemptomatik, yırtılmamış sinüs valsalva anevrizması sunuyoruz.

Anahtar Kelimeler: Valsalva sinüsü; aort anevrizması; aort kapağı; ekokardiyografi; tomografi, X-ray bilgisayarlı

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Sinus valsalva aneurysm is a rare congenital anomaly which can be associated with other cardiac abnormalities. These aneurysms are mostly congenital but secondary aneurysms are also encountered. Uncomplicated sinus valsalva aneurysms are generally asymptomatic unless they cause compression on an adjacent structure, thrombus formation within the aneurysm, with subsequent embolic events or infective complications. Continuous murmur is detected during cardiac auscultation as a result of fistulous connection to cardiac chambers. Association of sinus valsalva aneurysm with bicuspid aortic valve is an extremely rare clinical entity. In this case

report, we present a case of an unruptured sinus valsalva aneurysm associated with bicuspid aortic valve in a patient who remained asymptomatic until the age of 67 years.

CASE REPORT

A 67-year-old man who has hypertension was admitted to our outpatient clinic for routine clinical follow-up. He had no history of fatigue, dyspnea, palpitation or chest pain. His past medical history revealed long standing hypertension with inadequate control. Physical examination revealed blood pressure of 150/90 mmHg, and heart rate of 76 bpm. No evidence of valvular regurgitation and/or stenosis and heart failure sign was apparent on auscultation. The ECG was in sinus rhythm without any abnormal signs. Chest X-ray film was normal. In order to rule out end organ damage of hypertension, transthoracic echocardiography was performed. Transthoracic echocardiogram showed bicuspid aortic valve associated with unruptured right sinus valsalva aneurysm on parasternal short axis and subcostal views (Figure 1A, 1B). There was only mild aortic regurgitation and no other concomitant congenital anomaly was present. Due to frequent association of bicuspid aortic valve and other cardiac and coronary anomalies with bicuspid aortic valve,¹ and to further evaluate the aortic root, cardiac computerized tomography (CT) was performed as recommended by guidelines.² Cardiac CT confirmed the diagnosis of right sinus valsalva aneurysm (Figure 2A, 2B). There were no associated cardiac and coronary anomalies (Figure 2C). Surgical correction was recommended to the patient but

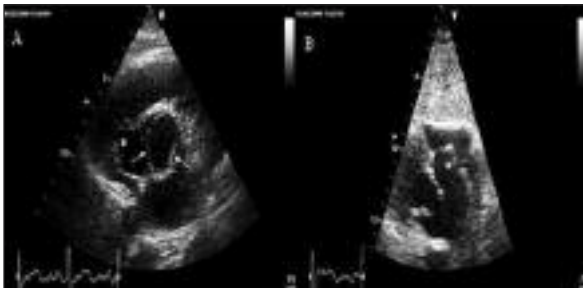


FIGURE 1: A) Parasternal short axis view showing bicuspid aortic valve associated with right sinus valsalva aneurysm. B) Sinus valsalva aneurysm from subcostal view.

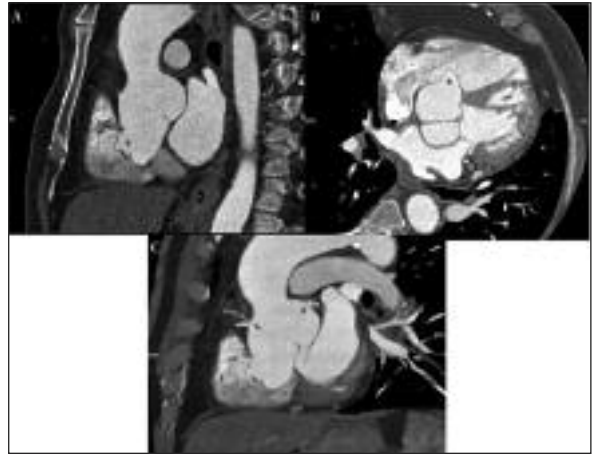


FIGURE 2: A) Sagittal view of cardiac computerized tomography reveals sinus of Valsalva aneurysm with well defined borders. B) Axial view reveals sinus of Valsalva aneurysm associated with bicuspid aortic valve. C) Coronal oblique view of cardiac computerized tomography shows normal coronary anatomy.

unfortunately he refused. Therefore, close medical follow up was planned.

DISCUSSION

Bicuspid aortic valve is one of the most common congenital cardiac anomalies affecting 1 to 2% of the population.³ Patients may remain asymptomatic for a long time during their life span or aortic stenosis or combination of stenosis and regurgitation of the aortic valve may occur at their third or fourth decades.⁴ The sinus valsalva aneurysm is a disorder mostly seen congenitally, but may also be secondary to other disease processes like syphilis and trauma.^{5,6} Congenital sinus valsalva aneurysm has been reported to be associated with other cardiac congenital anomalies such as ventricular septal defect (most common), atrial septal defect, coarctation of aorta, Ebstein's anomaly. However, association of sinus valsalva aneurysm and bicuspid aortic valve is an extremely rare clinical entity. Among associated cardiac lesions, reported coincidence of bicuspid aortic valve and sinus valsalva aneurysm in preoperative patients were 9% in a single center.⁷

The association of bicuspid aortic valve with other congenital cardiac anomalies was mostly explained by disintegration and lack of continuity of the aortic media and annulus fibrosa (aortic valve

ring).⁸ Although progressive degeneration of connective tissue of the aortic root was thought to be the main pathophysiologic mechanism in our patient, arterial hypertension and ageing might also contribute to aortic root dilatation in our patient, particularly above the sinotubular junction.⁹

Unruptured sinus valsalva aneurysm may become symptomatic due to right ventricular outflow obstruction, coronary artery compression, aortic regurgitation, complete heart block or resistant ventricular tachycardia.¹⁰ Reported mean age at the diagnosis of sinus valsalva aneurysm is 39.1 years in the literature.¹¹ However, our patient remained asymptomatic until the age of 67 years despite association of bicuspid aortic valve and large sinus valsalva aneurysm. Aortic valve showed only minimal aortic regurgitation and there were no arrhythmic, embolic and/or coronary complications. Cardiac CT demonstrated large right sinus valsalva aneurysm with well defined borders and no connection with adjacent cardiac structures was found. Also, no coronary anomaly was present on cardiac CT.

Sinus valsalva aneurysms causing symptoms and ruptured aneurysms mandate prompt and urgent surgical correction. Uncomplicated aneurysms have a relatively lower surgical mortality and morbidity,⁷ however survival in patients with ruptured sinus valsalva aneurysm ranges between 1 to 4 years without any intervention.^{7,12} Although symptomatic and/or ruptured sinus valsalva aortic aneurysms require surgical intervention,¹² surgery

indication in patients with asymptomatic uncomplicated sinus valsalva aneurysm is somewhat controversial. There are different reports regarding the natural history of asymptomatic sinus valsalva aneurysms. According to these reports, some of which remain silent for long time period but some actually show progressive clinical deterioration.^{11,13} Therefore, algorithms were established for the management of sinus valsalva aneurysms. Surgery is a clear indication for patients who are symptomatic or for those having a ruptured aneurysm, but in uncomplicated aneurysms, therapy must be individualized. Semi-annually follow up to detect any possible enlargement, infection and rupture of the aneurysm is generally recommended for patients with asymptomatic unruptured sinus valsalva aneurysms with tricuspid aortic valves, however, surgery is recommended to those with sinus valsalva aneurysms associated with bicuspid aortic valves.³ There is no specific recommendation addressing patients with sinus valsalva aneurysms in current guidelines, however in patients with dilated ascending aorta or root associated with a bicuspid aortic valve, serial evaluation with transthoracic echocardiography, cardiac computed tomography or magnetic resonance is recommended.² Although, elective surgical correction was recommended to our patient in an attempt to prevent future possible cardiac complications, he did not accept surgery and close medical follow up was planned. Bearing in mind the genetic component of the both anomalies, family screening for aortic valve and ascending aortic disease was also recommended.

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