

An Unusual Case of Bicuspid Aortic Valve Masquerading as NSTEMI

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Abstract

Case Report

Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly with a prevalence between 0.5% and 2% in the general population [1], and a male predominance of approximately 3:1 [2]. There is a wide spectrum of presentation ranging from a severe symptomatic form in the neonatal period to incidental detection later in life, including adulthood [2]. Possible symptoms include chest pain, dyspnea, and syncope, which are related to complications. These complications can involve aortic valve stenosis or incompetence, endocarditis, aortic aneurysm formation, and aortic dissection. While most long-term complications associated with BAV manifest later in life, children and adolescents may experience early onset valvular dysfunction or aortic dilation [2]. Here, we report the case of a 36-year-old female with an unusual revelation mode of BAV during an acute coronary syndrome (NSTEMI).

Keywords: Bicuspid aortic valve (BAV), male predominance, NSTEMI, valvular dysfunction.

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INTRODUCTION

Bicuspid aortic valve (BAV) disease is the most common congenital heart defect, with a prevalence between 0.5 and 2%. There is a male predominance with a male to female ratio 3:1 [1, 2]. Although the clinical presentation of patients with BAV can range from severe valve disease in neonatal period to asymptomatic valve or thoracic aortic disease in old age, symptoms typically emerge in adulthood. The clinical manifestations relate to the function of the aortic valve (stenosis or incompetence), the aortopathy (dissection), and acquired complications such as endocarditis [2].

Transthoracic echocardiography (TTE) continues to be the main technique for diagnosing and monitoring bicuspid aortic valve (BAV) and aortic dilation in children and adolescents. Assessing the morphology of the aortic valve and the patterns of cusp fusion is crucial, as these factors can predict future complications and prognosis [3].

Here, we report the case of a 36-year-old female with an unusual revelation mode of BAV during an acute coronary syndrome (NSTEMI).

CASE PRESENTATION

A 36-year-old female patient, at low cardiovascular risk. Visited Emergency department complaining of prolonged midsternal, non-radiating chest pain. With a history of angina (Canadian Cardiovascular Society (CCS) Class II) that began ten days prior. She was not on any medications and had no significant family history of cardiac disease.

On physical examination, her blood pressure was 110/74 mmHg, her heart rate was 105 beats/min, and a grade IV/VI crescendo-decrescendo late-peaking murmur in aortic area that radiated to the carotids. These physical examination signs were consistent with aortic stenosis. There was no signs of heart failure or other specific clinical signs.

A 12-lead EKG revealed an irregular sinus rhythm at 107 beats/min with left ventricle hypertrophy and repolarization abnormality (Figure 1). Laboratory work revealed an increase of myocardial necrosis markers while thyroid, renal, and liver function tests were unremarkable.

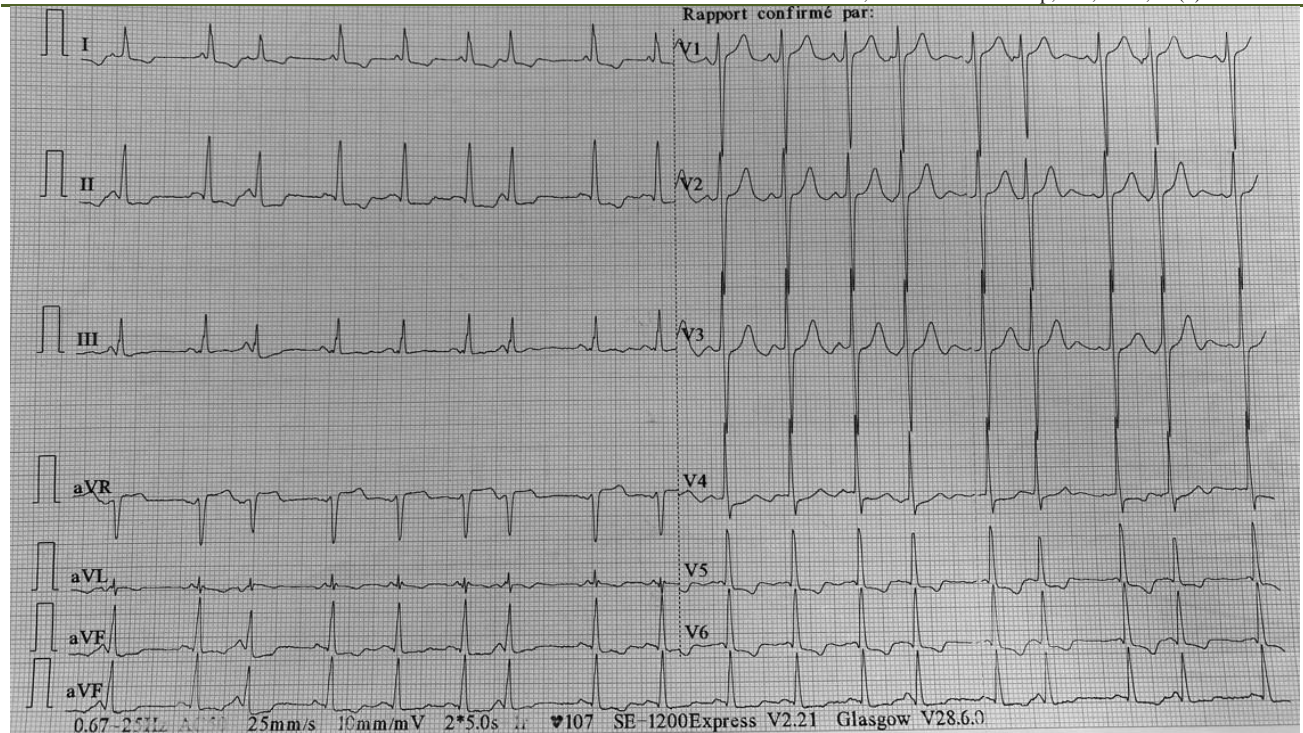


Figure 1: 12-lead EKG

Echocardiography (TTE) revealed an ejection fraction of 61%, suspected bicuspid aortic valve with critical aortic stenosis and peak velocity of 4,9 m/sec, a mean gradient of 56 mmHg, and an indexed valve area of 0.3 cm², all of which were suggestive of critical aortic stenosis (Figure 2). A transesophageal echocardiography

(TEE) further confirmed the presence of a Type 0 bicuspid aortic valve (Figure 3) without initial aortic dilation. An additional investigation involving a thoracic CT scan was conducted, yielding concordant results (Figure 4). Elective coronary angiography done a day later revealed normal coronaries.

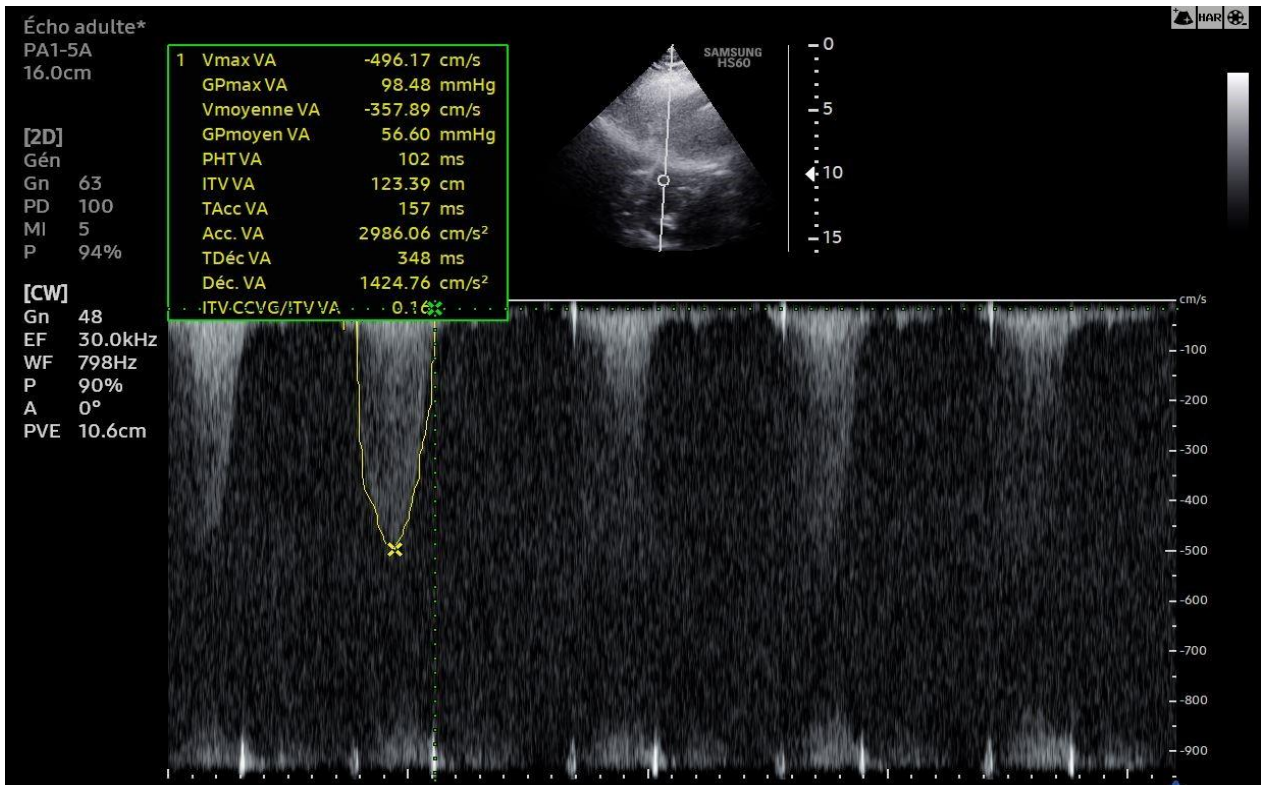


Figure 2: Continuous wave doppler showing criteria of critical aortic valve stenosis

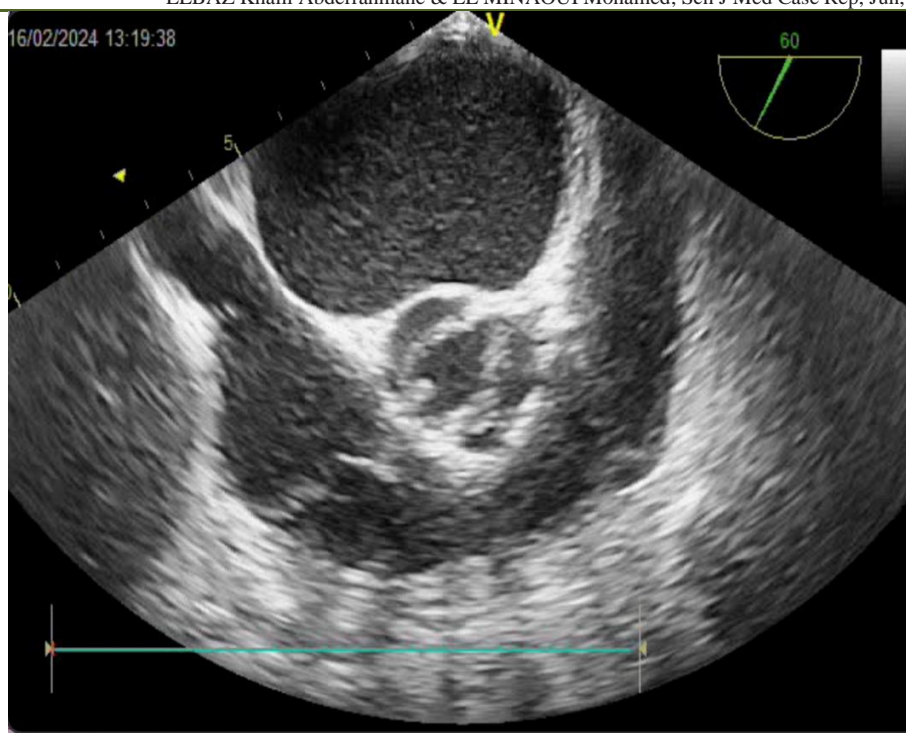


Figure 3: Transesophageal short-axis view of a type 0 bicuspid aortic valve (BAV)

Acutely, the patient was treated with double antiplatelet therapy (aspirin plus clopidogrel), heparin and statin. The early use of non-invasive examinations, such as TTE and TEE, enabled the diagnosis and

management to be rectified. The patient was then put on beta-blockers and referred for surgery. She subsequently underwent uncomplicated aortic valve replacement surgery and is doing well.



Figure 4: Sagittal CT scan image of the thoracic aorta. Note the absence of dilation of the thoracic initial aorta

DISCUSSION

BAV, the most common congenital heart defect in children and adults with a prevalence between 0,5 and 2% [1]. BAV occurs with the fusion of two aortic valve (AV) cusps usually with a distinct raphe at the site of the fused commissure.

The Sievers classification for BAV has been introduced to sub-divide BAV into different morphological phenotypes, based on number of raphe and spatial position of cusps or raphe [4]. The phenotype of BAV helps predict the future complications and prognosis and may guide the follow-up. Transthoracic echocardiography (TTE) remains a

primary modality for diagnosis and surveillance of BAV and aortic dilation [3].

The presentation of patients with BAV can range from severe valve disease in neonatal period to asymptomatic valve or thoracic aortic disease in old age, symptoms typically emerge in adulthood. Clinical manifestations relate to the function of the aortic valve (stenosis or incompetence), the aortopathy (dissection), and acquired complications such as endocarditis.

In childhood, BAV disease is often asymptomatic. It is estimated that only 1 in 50 children will have clinically significant valve disease by adolescence [5]. Aortic stenosis, resulting from a small valve orifice, can manifest in children with BAVs. Likewise, pure aortic incompetence due to a prolapsed leaflet may also occur during childhood. As individuals reach adulthood, the abnormal shear stress eventually leads to valve calcification. In some cases, there is additional dilation of the aortic root [6, 7]. Recent series have demonstrated that cardiac event rates are higher when one or more of the following risk factors are present: age over 30 years, moderate or severe aortic stenosis, and moderate or severe aortic incompetence [8, 9].

The diagnosis of BAV involves auscultatory findings such as an ejection sound, typically best heard at the apex. Associated murmurs of aortic stenosis, incompetence, or coarctation of the aorta may be present when these lesions occur. In the current era, confirmation of the diagnosis is usually achieved through transthoracic echocardiograms [3]. If there is uncertainty in diagnosis, a TEE can improve visualization of the leaflets. In certain cases, alternative cardiac imaging modalities like cardiac magnetic resonance imaging or computed tomography may aid in confirming BAV anatomy. However, more commonly, these imaging techniques are utilized to visualize the thoracic aorta [3].

In literature, there are reports of cases involving acute myocardial infarction in patients with bicuspid aortic valve, attributed to the embolization of calcific material from the valve. This can occur during catheterization or may happen spontaneously [10, 11].

This case illustrates an uncommon presentation of aortic stenosis and bicuspid aortic valve (BAV) as an acute coronary syndrome (ACS), highlighting the importance of a meticulous physical examination and prompt utilization of non-invasive diagnostic techniques, such as echocardiography. This is especially crucial in young patients with low cardiovascular risk. The mechanisms behind the elevation in cardiac biomarkers remain unclear. Potential explanations include coronary embolization of valvular calcium material, an imbalance between oxygen supply and demand secondary to aortic stenosis, or a reduced coronary flow reserve and diastolic perfusion time due to ventricular hypertrophy.

CONCLUSION

BAV is the most common congenital heart defect and has a wide spectrum of presentations. It can be discovered under a variety of circumstances including incidental auscultatory finding of a murmur or through paraclinical investigations, it can also be diagnosed in emergencies, such as when acute coronary syndrome is suspected. Simple, rapid, and non-invasive methods, such as clinical examination and ultrasound, are employed to identify and screen for aortic valve disease, particularly in young patients with low cardiovascular risk.

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