CASE REPORT

Bicuspid aortic valve with dissecting aortic aneurysm in an African

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ABSTRACT

Bicuspid aortic valve (BAV) is a relatively rare congenital heart disease (CHD) in Africans compared to caucasians and it is an independent risk factor for aortic aneurysm and dissection. We therefore report a case of a 51-year-old farmer who presented with clinical features of heart failure (HF). He had a diastolic murmur loudest in the aortic area. Echocardiography revealed BAV with regurgitation. The aortic root and ascending aorta were dilated with dissection flaps which mimicked a double aortic valve. Computed Tomography angiography of the aorta confirmed Debakey's type I (Stanford A) dissecting aortic aneurysm (DAA). He was managed for HF due to chronic aortic regurgitation and referred for surgery. BAV is a relatively rare CHD in Africans and is a strong risk factor for DAA. Long-term monitoring of the aorta and timely surgical intervention are recommended when indicated. SAHeart 2020;17:208-210

INTRODUCTION

Bicuspid aortic valve (BAV) is a congenial heart disease (CHD) with 2 functional aortic valve cusps instead of 3. It is common in caucasians, but relatively rare in Africans.[1,2,3] It is associated with aortic wall abnormalities (aortopathy) and may occur in isolation or in combination with other CHD. BAV is a risk factor for life-threatening complications such as infective endocarditis, stroke, aortic aneurysm and dissection (AD). Dissecting aortic aneurysm (DAA) is an abnormal dilatation of the aorta with separation of the inner and middle layers of the aortic wall.

BAV has not been well documented as a CHD or as a risk factor for DAA in our setting (West Africa). Other CHD such as ventricular and atrial septal defects have been extensively reported, but BAV remains under-reported, hence the need for this report. Ranjan, et al.[4] reported a case of BAV in a 37-year-old male with hypertension, while Amin, et al.[5] reported another in a 23-year-old female with severe aortic stenosis and heart failure, but these were reported abroad (Bangladesh). In the US, Mahajan, et al.[6] reported a case with stroke in a 52-year-old man. This report, though a single case, highlights the rarity of BAV in Africans, and offers the foundation for prospective case series. It also emphasises the associated aortopathies, the need for long-term surveillance of the aorta and timely surgical intervention.

We therefore report the case of a middle-aged African farmer with BAV and Debakey's type I dissecting aortic aneurysm (DAA).

CASE SUMMARY

Approval was obtained from the hospital's Research and Ethics Committee on 16 May 2019 and the reference number is ADM/E 22/A/VOL.VII/14750. Informed consent was obtained from the patient. Mr E.I., a 51-year-old farmer presented with complains of bilateral leg swelling and cough of 3 and 2 months duration respectively. He also had breathlessness on exertion, orthopnoea and paroxysmal nocturnal dyspnea. There was no history of chest, abdominal or back pain. He was not hypertensive and there is no history of trauma, smoking or heart disease in the family.

On examination, he was an acutely ill looking middle-aged man of normal habitus with bilateral pitting pedal edema up to the knees. His pulse rate was 100bpm, regular and of normal volume, with no radio femoral delay and other pulses were palpable and symmetrical. Blood pressure was normal and

similar in both arms. The jugular venous pressure was 6cm elevated above the sternal angle of Louis. The apex beat was displaced to the 6th left intercostal space, lateral to the mid-clavicular line. He had a 3rd heart sound with gallop rhythm. There was an early diastolic murmur loudest in the aortic area and it radiated to the left sternal border. There were fine crepitations in the lung bases and a soft tender hepatomegaly, 6cm below the costal margin.

Echocardiography revealed a bicuspid aortic valve (Figure 1a) with aortic regurgitation and stenosis. The ascending aorta was aneurysmal (6.8cm) with mobile intimal flaps just distal to the aortic valve, which mimicked a double aortic valve (Figure 1b). The chambers were dilated with reduced left ventricular systolic function.

Computerised Tomography (CT) angiography of the aorta (Figure 2a) confirmed De-Bakey's type 1, Stanford type A dissecting aneurysm. There was no coarctation of the aorta. A diagnosis of heart failure (HF) due to chronic aortic regurgitation was made. He was placed on medical therapy for heart failure and referred for surgery (aortic root replacement and repair of dissection). Surgery is being awaited pending availability of funds. His first-degree relatives are to be screened for BAV.

DISCUSSION

This patient has Type 0 BAV (Figure 1a) according to the Sievers and Schmidtke classification.[7] Type 0 has no raphe with a single horizontal or vertical commissure and it is the least prevalent. Types 1 and 2 have raphes at different locations depending on the cusps that are fused. Fusion of left and right coronary cusps is the most common morphology found in about 60% of patients. The morphology of BAV correlates with the haemodynamic lesion (aortic stenosis and/or regurgitation) and the location of the aortic dilatation.[8] He had predominant aortic regurgitation (AR) with mild stenosis. Aortic stenosis is more common in BAV and is usually severe. The severity of the AR is due to prolapse of dissection flaps into the aortic valve in diastole, in addition to poor coaptation from the dilated aortic root and unequal sizes of cusps. Valvular dysfunction is the natural consequence of BAV due to early sclerosis and progressive fibro calcification of the cusps.

The ascending aorta was 6.8cm dilated (Figure 1b). Aneurysm of the aortic root and ascending aorta are consistent aortic wall abnormalities associated with BAV with a prevalence of 30% - 70% of cases.[8] It may be present at birth and progresses at a rate of 0.3 - 1.1mm/year.[9] Other aortopathies include coarctation of the aorta and interrupted aortic arch. Aortic aneurysm is due to degenerative changes in the media. Abnormal flow pattern in the ascending aorta and genetic factors may also be contributory. Aortic aneurysm is a significant predictor of aortic dissection, rupture and sudden death. He also had (DeBakey type 1) aortic dissection (Figure 2). BAV has a 6.14% lifetime risk of AD which is 9 times higher than the general population.[10] This risk is related to aortic dimensions and persists even after valve replacement. However, the functional status of the aortic valve is not a factor.

Our patient is a male adult, and this is not surprising because BAV has a strong male predominance and is usually clinically

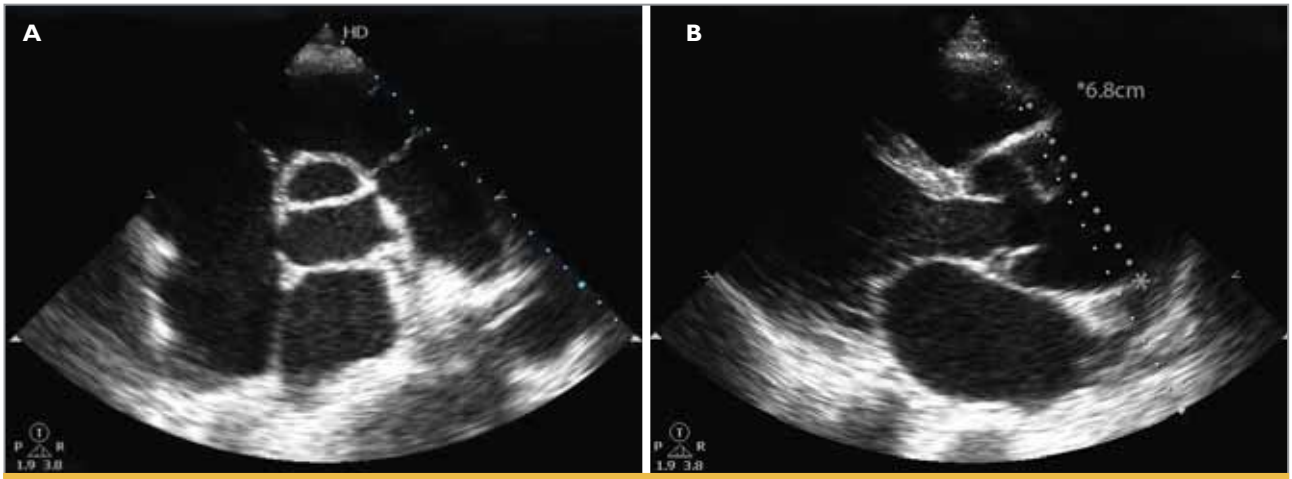

FIGURE 1: (A) **2D trans-thoracic echocardiography**; bicuspid aortic valve in diastole, with anterior, posterior cusps and horizontal commissure. (B) **2D trans-thoracic echocardiography**; ascending aortic aneurysm, intimal dissection flaps above the aortic valve (double aortic valve sign).

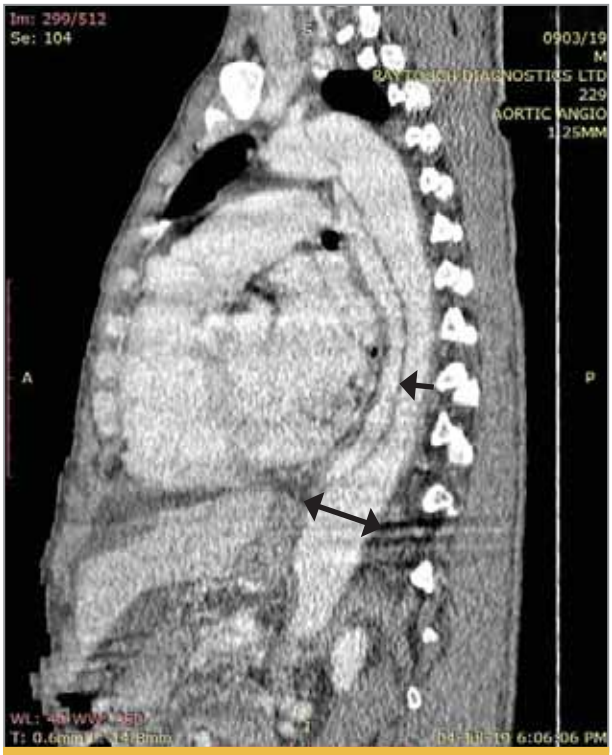

FIGURE 2: CT angiography of the aorta (para-sagittal reformatted image); dilated aorta (double arrow heads), intimal flap (single arrow head) extending from the arch of the aorta, descending and abdominal aorta. De-Bakey's type I (Stanford A) dissecting aortic aneurysm.

silent until after the third or fourth decade of life.[11] The absence of chest or abdominal pain is not the typical presentation of AD, but about 10% of AD are painless.[12] The absence of pain must have contributed to the late presentation.

In conclusion, BAV is a relatively rare CHD in persons of African ancestry and is a strong risk factor for aortic aneurysm and dissection. Therefore, serial monitoring of the aorta with prompt surgical intervention are recommended to prevent these potentially fatal complications.

Conflict of interest: none declared.

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