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Case Report

Bicuspid Aortic Valve, Bovine Aortic Arch and early Atherosclerosis: When one plus one equals three

Abstract

Despite that even normally functioning bicuspid aortic valves can have abnormal transvalvular-flow patterns, resulting in regional increases in wall shear stress and aortic dilatation, most often are preserved during ascending aorta replacement. On the other hand, the most common normal variant of aortic arch branching, known as bovine aortic arch, may be considered a risk factor for the development of ascending aorta dilatation, as recent studies have shown. However, data are lacking in the literature regarding the involvement of bicuspid aortic valve and/or bovine aortic arch in early atherosclerosis development.

We present a case of a young female patient with bicuspid aortic valve, bovine aortic arch and advanced carotid atherosclerotic disease disproportionate to her cardiovascular risk profile and solely explained by hemodynamic alterations produced by those two 'normal' anatomical variations. We present a case of a young female patient with bicuspid aortic valve, bovine aortic arch and advanced carotid atherosclerotic disease disproportionate to her cardiovascular risk profile and solely explained by hemodynamic alterations produced by those two 'normal' anatomical variations.

Case Report

A 36-year old white female, housewife, presented in the preventive cardiology and antihypertensive unit of our clinic, with a two-month history of stage III systemic hypertension (HTN) with associated palpitations. Despite initial treatment with barnidipine 10mg OD, her home blood pressure (BP) remained poorly controlled (160/80 and 140/75 mmHg in the morning and evening, respectively).

She reported both maternal and paternal history of hypertension at a young age, while her father had also suffered from an ischemic stroke at the age of 55, which was attributed to long-lasting and untreated severe HTN.

As far as her personal history is concerned, we should emphasize that she is a woman of normal weight (BMI: 24.6 kg/m²), without abdominal obesity (waist circumference 82cm), with normal menstruation and two uncomplicated childbirths, with no history of HTN during pregnancy, occasional smoker the last five years (3-5 cigarettes/week) and without any previous drug therapy. During an athletic preparticipation screening, almost 1.5 years prior to her visit in our unit, she was found to have a soft systolic murmur and the echocardiographic study revealed dilatation of the ascending thoracic aorta (45

mm) and a bicuspid aortic valve (BAV) with fusion of the right and left coronary cusps without any signs of aortic stenosis or regurgitation. The CT angiography that followed confirmed the presence of an ascending aortic aneurysm (48 mm) and the patient was referred for surgical consultation. One month later the patient underwent replacement of the ascending thoracic aorta with a tubular Dacron graft with preservation of her native valve. The postoperative course was uneventful. Moreover, the echocardiographic screening of her 1st degree relatives did not reveal any valve or ascending aorta pathology.

A year and a half after her surgery, the patient discovered high BP values in random home BP measurements which led her visit our unit. On-treatment office blood pressure was 154/93 mmHg and 142/80 mmHg, in the right and left arm, respectively, while 24-hour ambulatory BP monitoring (ABPM) showed slightly increased average values of SBP, during the day and night (135/76 mmHg, 127/74 mmHg, respectively), with SBP loads exceeding 45% and inadequate nocturnal fall of SBP (6.3%) defining her as a non-dipper.

Complete clinical evaluation, as well as her glycemic, metabolic and lipid profile (LDL-Cholesterol: 123 mg/dl, HDL-C: 56 mg/dl, Lp (a): 7.3 nmol/l) were normal.

The patient's relative 10-year cardiovascular risk according to HeartScore was 4 times greater compared with normal subjects of the same sex and age, mainly due to smoking (explaining 65% of her total CVD risk).

The 12-lead ECG showed sinus rhythm, 70 bpm and normal electrical axis without ST-T abnormalities or signs of LV hypertrophy. The fundoscopic examination revealed findings of advanced hypertensive retinopathy (stage II-III according to Sheie classification). The echocardiogram revealed concentric left ventricular hypertrophy (LVMI 100 gr/m^2 , RWT: 0.775), normal LV diastolic function (LAVI: 17.9 mL/m^2 , mitral E/A: 1.62, Dec Time: 189 msec) and a BAV with fusion of the right and left coronary cusps, with an excellent hemodynamic profile (V_{max} : 1.35 m/sec, mean PG: 4.31 mmHg) without any signs of regurgitation or calcification of the commissures. Ascending aortic graft and aortic arch diameter (28.3 mm) were normal, while an anomalous common origin of the innominate and the left common carotid artery known as bovine aortic arch (BAA), was noted (Figure 1).

Duplex vascular ultrasonography demonstrated mild and diffuse intimal-media thickening (IMT) of the lower extremity arteries with small (<5% stenosis) calcified atherosclerotic plaques at the level of femoral arteries, while abdominal aorta and renal arteries were normal. However, carotid ultrasound revealed diffuse IMT thickening of the carotid arteries, more advanced on the left side, with a small longitudinal atherosclerotic plaque (1.5 mm thick) to the right bulb, and a sizable (30% stenosis) longitudinal isoechogenic homogeneous plaque at the level of the left bulb extending to the left internal carotid artery causing approximately 60% stenosis (Figure 2). Based on the above findings the patient underwent a treadmill exercise test according to Bruce protocol (achieving 13.5 METS), which was negative for ischemia and with normal inotropic and chronotropic response.

Considering the advanced atherosclerotic findings of this young female patient, we decided, apart from reinforcing her antihypertensive treatment with olmesartan/amlodipine fixed combination 40/10 OD and nebivolol 5 mg OD, the addition of atorvastatin 20 mg OD and acetylsalicylic acid 100 mg OD, in order to reduce her actual cardiovascular risk.

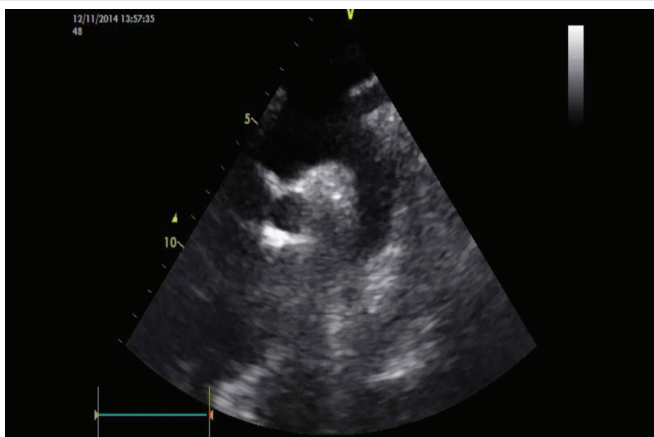


Figure 1: Suprasternal echocardiographic view. Bovine aortic arch.

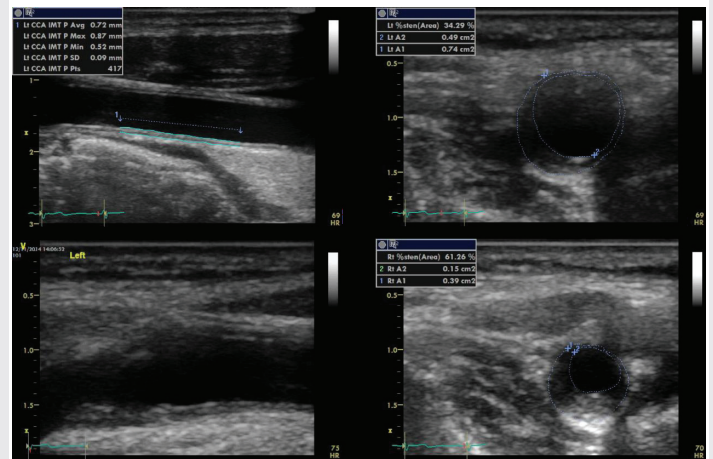


Figure 2: Carotid ultrasonography reveals diffuse left common carotid intima media thickening (A), long uniform isoechogenic with regular surface atherosclerotic plaque (B) which produces 30% stenosis at the bulb level (C) and 60% stenosis at the level of the left internal carotid artery (D).

Discussion

We present a case of a young female patient with BAV and surgically corrected bicuspid aortopathy, BAA, newly diagnosed essential hypertension, generalized early atherosclerosis and carotid atherosclerotic disease. Are those entities interrelated?

BAV is the most common congenital heart defect in adults, affecting 1.3% of the population worldwide [1]. Although aortic stenosis and aortic regurgitation are the most common complications of BAV, dilatation of any or all segments of the proximal aorta from the aortic root to the aortic arch, known as bicuspid aortopathy, is also present in approximately 50% of the affected persons. The development of aortopathy and its specific types has been attributed to genetic and hemodynamic factors. The concept that abnormal valve dynamics lead to bicuspid aortopathy is supported by the observation that even normally functioning BAV, as in the case of our patient, can result in regional wall shear stress abnormalities, which predispose to aortic aortopathy.

Recent ESC guidelines for aortic diseases recommend that surgery should be considered in patients with a BAV who have aortic root aneurysm with maximal ascending aortic diameter ≥ 55 mm without other risk factors and ≥ 50 mm with the presence of at least one of the following risk factors: coarctation of the aorta, systemic HTN, family history of dissection or an expansion rate of the ascending aortic aneurysm (AAA) of ≥ 3 mm per year. However, guidelines emphasize that the decision for surgery in patients with small body size, must be based on the aortic diameter indexed to BSA, with a threshold ≥ 27.5 m^2 [1]. Accordingly, the referral for cardiovascular surgery of our patient was appropriate, considering her ascending aortic dilation index equal to 30 mm/m^2 .

Considering the 'unexplained' advanced carotid findings of our patient, the preservation of the 'normal' functioning BAV, without valve tricuspidization or replacement in concert with aortic graft placement may possibly aggravate even

further local hemodynamic alterations. There are studies that have shown that proximal aortic graft by itself leads to great haemodynamic alterations and development of systolic hypertension [3].

Despite the lack of evidence in current literature concerning the association of BAV with the appearance of early atherosclerosis, data from our unit show numerous cases of young patients with BAV and early carotid atherosclerosis, which cannot be related and explained based on their family history and the conventional cardiovascular risk factors. Probably, this discrepancy can be attributed to the rarely used carotid ultrasound screening of young patients with BAV and bicuspid aortopathy. To our knowledge only few reports exist correlating BAV with systemic atherosclerosis indices, such as carotid to femoral pulse wave velocity, augmentation index, flow-mediated dilatation and metalloproteinase-2 levels in plasma [4,5].

In the majority of patients (74%), there are three great vessels branching from the aortic arch: the innominate artery, the left carotid artery and the left subclavian artery. However, there are many other possible anatomic configurations of the aortic arch – the most common being the BAA configuration, in which the innominate and the left carotid artery originate from a common stem of the aortic arch. The term refers to both a shared origin of the left common carotid and innominate artery (most common), and an origin of the left common carotid from the innominate artery (less common) [6].

Assumed to have little physiological consequence, the principal significance of identifying a BAA has been for the planning of interventional and surgical procedures. However, Malone et al found recently a strong association between BAA and aortic aneurysm in patients over 70 years old [7]. The authors tried to explain this association using Coanda effect [8]. They proposed that the wide origin of the combined innominate and left common carotid arteries leads to a streaming phenomenon, which would marginalize high velocity blood flow and potentially increase vessel wall shear stress. Increased shear stress has been linked to ascending aortic wall injury and dilatation. The last 3 years several studies have demonstrated the association of BAA with dilatation of the ascending aorta and suggested that BAA should be considered a risk factor for aortic dilatation [9-12]. Furthermore, Eleftheriades JA et al have proceeded with histological assessment studies of the aortic wall in patients with BAV and BAA and found that both entities seem to affect vascular wall architecture [13].

Up to date, the association of BAA with carotid atherosclerosis was assessed only in one study of Baadh et al in a small number of patients, which showed no significant correlation [14]. However, the last few years, there is intense research interest in the influence of the geometry and morphology of local blood flow on the development of carotid atherosclerosis. Studies from Phan T et al in 2012 revealed that the angle between the common and internal carotid artery as well as the angle between internal and external carotid artery are significant and

independent risk factors for the development of atherosclerotic carotid plaques [15]. In the case of BAA, the origin of the left common carotid artery from the innominate artery may have a significant effect on the geometry of the carotid bulb, altering regional blood flow dynamics and thus contributing to an *increased susceptibility* for the development of atherosclerotic carotid disease.

Conclusion

Complete cardiovascular system evaluation often reveals “normal” anatomical variants, such as bovine aortic arch and normally functioning bicuspid aortic valve, as significant etiological factors of early atherosclerosis development. To our knowledge this is the first case report connecting those two entities with aortic dilatation and early carotid atherosclerosis. Consequently, a thorough evaluation of the entire arterial tree in such individuals should be an integral part of the diagnostic and therapeutic investigation.

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