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Case Report

Severe Bicuspid Aortic Valve Stenosis, Single Right Coronary Artery and Main Stem-Equivalent Atherosclerosis: A Functional Triad Causing Effort-Induced Syncope

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Introduction

Syncope is a common hospital presentation with a broad underlying differential diagnosis. We report a novel functional triad of severe biscuspid aortic stenosis, single right coronary artery and stenosis of the main stem-equivalent in a patient presenting with exercise-induced syncope. These three pathologies are proposed to be functionally synergistic, leading to hypoperfusion of the myocardium on exertion.

A 59 year-old man was admitted to the emergency department (ED) following a sudden collapse while playing soccer. After 20 minutes of the game he felt sweaty and unwell and had paused to rest. Without further warning he was seen to collapse to the ground with loss of consciousness. His teammate, a police officer, was unable to feel a carotid pulse and immediately commenced chest compressions. After two minutes there was spontaneous return of circulation and he was found to be conscious and alert on arrival of the ambulance service.

In the ED he was afebrile, with a blood pressure of 119/68 mmHg, heart rate of 83 bpm and oxygen saturations of 95% on air. Cardiovascular examination revealed a harsh ejection systolic murmur over the aortic valve area radiating to the carotid arteries and precordium. There was mild chest wall tenderness at the site of chest compressions.

The admission ECG showed sinus rhythm with no ischemic changes. Chest X-ray and routine blood tests (including troponin I) were normal. Transthoracic (and subsequent transoesophageal) echocardiography demonstrated severe stenosis of a heavily calcified bicuspid aortic valve (peak gradient 72 mmHg, mean gradient of 40 mmHg, valve area of 0.69 cm^2; figure panels A, B, C). Left ventricular size and function was normal, with no regional wall motion abnormalities. There was no evidence of aortic root dilatation or coarctation.

The patient underwent coronary angiography as part of the surgical work-up for aortic valve replacement. Unexpectedly, this revealed a congenital coronary artery anomaly, with a single coronary artery arising from the right sinus of Valsalva (D, E, F). After providing the posterior descending and posterior left ventricular branches, its course continued in the left atrioventricular groove before giving rise to the obtuse marginal branches and subsequently the left anterior descending artery (Lipton classification R-IIA).

Coronary angiography also identified at least moderate atherosclerotic disease of the single coronary artery, prior to the origin of the OM branches in the main-stem equivalent part of the vessel (arrowed, E). Due to the unusual anatomy, these findings were confirmed by gated coronary CT angiography prior to surgery (Toshiba Aquilion CX) (G, H).

The patient underwent uncomplicated mechanical aortic valve replacement and coronary artery bypasses grafting (left internal mammary artery to left anterior descending and a saphenous vein graft to the circumflex; panel I) and was well at two-month follow-up.

To our knowledge, this is the first report of a patient presenting with the combination of severe bicuspid aortic stenosis, a single right coronary artery (RCA) and atherosclerosis of the main stem-equivalent. We hypothesize that these three pathologies are functionally synergistic, leading to hypoperfusion of the myocardium on exertion, resulting in the clinical presentation of effort-induced syncope in this patient.

Anomalies of the coronary arteries affect approximately 1% of the population, although single coronary artery is rare, in 0.02-0.07% in published series [1]. The functional significance of a single RCA is debated and highly variable. A retro-aortic course, with compression of the RCA between the aorta and the main pulmonary artery, has been described as a cause of angina, syncope and sudden death. While an anterior course is relatively benign, a single RCA may predispose to effort-related ischemia even in the absence of coronary artery disease, due to inadequate myocardial blood supply to a large myocardial territory [2].

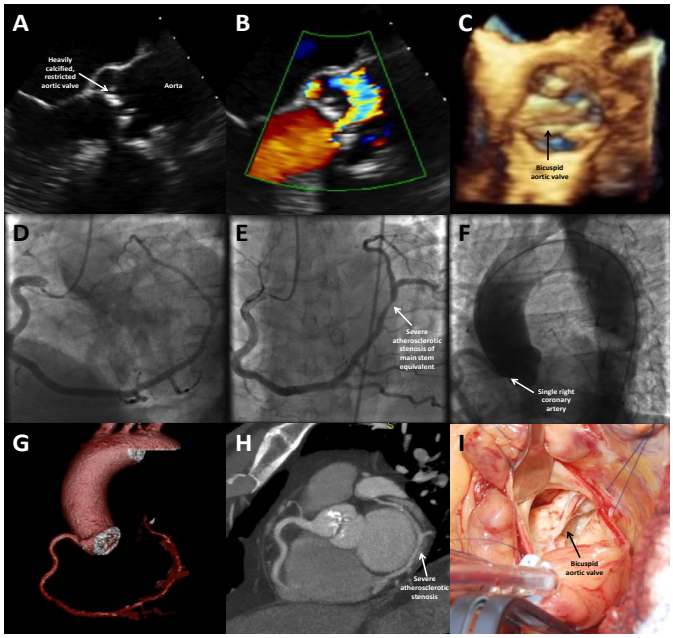
Bicuspid aortic valve (BAV) is frequently associated with anomalies of the coronary arteries, underpinning their common developmental pathway. While the anatomic association is well reported, pathophysiological synergy between BAV stenosis and anomalous coronary arteries is extremely rare, and is not previously reported in a patient with a single RCA [3,4].

While inducible ischemia distal to the main stem disease could not be confirmed due to the severity of aortic stenosis, this seems likely to be contributory, given the large distal myocardial territory supplied. Confirmation by measurement of fractional-flow reserve at angiography could have contributed to the assessment of the flow limiting nature of this stenosis, but since the patient was undergoing AVR this was deemed to be unnecessary.

In conclusion, functional synergy between the triad of aortic stenosis, single coronary artery and main stem-equivalent stenosis is proposed as an unusual cause of exertional syncope. Conventional surgical management with valve replacement and bypass grafting was curative.

Figure Legend

Transoesophageal echocardiography showed a heavily calcified bicuspid aortic valve with severely restricted opening of both leaflets and turbulent blood flow (A–2D x-plane; B–colour Doppler; C–3D en face). Coronary angiography and aortography (D–LAO projection; E–PA projection; F–aortogram) revealed a single RCA arising from the right sinus of Valsalva, with no evidence of aortic coarctation. Prior to the origin of the OM branch, in the main stem-equivalent part, there was at least moderate atherosclerotic disease (E, arrowed) of the single RCA, confirmed on CT coronary angiography (G, H). The everely stenotic bicuspid aortic valve was visualized directly with the patient on cardiopulmonary bypass at the time of aortic valve replacement and coronary artery bypass grafting (I).



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