

Syncope in a Previously Healthy Young Adult: Undiagnosed Aortic Stenosis, Dilation and Coarctation Requiring Emergent Surgery

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Abstract Aortic stenosis is a valvular heart disease that needs particular attention; due to the fact that asymptomatic patients may go undiagnosed for periods of time long enough to cause severe morbidity. On the other hand, aortic stenosis frequently is accompanied with other cardiac pathologies, such as a post-stenotic dilation, and coarctation. Imputed as a possible causative factor for sudden cardiac death, among other, aortic stenosis is a common finding in the setting of a bicuspid valve. We present the case of a previously young adult that suffered an episode of syncope during a football game. Exertion and sport activities are well-known situations that might provoke an abrupt manifestation of the latent valvular pathology. We dealt with an aortic stenosis of severe grade, accompanied with dilation and coarctation of aorta, in an individual with bicuspid aortic valve. A major heart surgery intervention was performed, with replacement of the valve, correction of the dilated arch and removal of the coarcted segment. Close follow-ups and monitoring are warranted even in asymptomatic patients, when the stenosis is uncovered as an incidental finding, during routine checkups.

Keywords: Aortic stenosis, left ventricular hypertrophy, dilation of aorta, coarctation, bicuspid aortic valve, syncope

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1. Introduction

Aortic stenosis (AS) is a relatively frequent valvular heart disease that causes important hemodynamic problems, related to the obstruction of the blood outflow from the left ventricle. Bicuspid valves are a common finding in younger patients presenting with aortic stenosis. The condition seems congenital, with males affected predominantly, and with an overall prevalence of 1-2% [1]. Bicuspid aortic valve (BAV) is responsible for high frequency of aortic wall abnormalities, and aortic dilation seems to be so importantly present, to warrant corrective interventions. BAV patients with a high risk of aortic complications have been identified, among which those with aortic coarctation (corrected or not), with aortic stenosis, with severe valvular dysfunction or systemic hypertension, and individuals with positive familial history for aortic dissection or rupture [2].

The impairment of cusp opening in the setting of AS will lead to a pressure overload, with a compensatory left ventricular hypertrophy (LVH), followed from a fall in the ventricular compliance [3]. Angina may result from the increased wall stress (especially in the setting of a hypertrophied myocardial tissue), and the situation might

progress to heart failure if the systolic function of the left ventricle declines progressively. These occurrences are not the only one accompanying BAV complicated with valve stenosis; in fact the list is much longer, with coarctation of aorta, coronary arteries anomalies, aneurysm of sinus of Valsalva and of the ascending aorta, and other congenital heart defects [4].

Valve stenosis and its severity is an important parameter to be carefully evaluated and quantified. Gorlin formula calculated the valve area from the ratio of the cardiac output to the square root of the valve gradient; normal values suggested for the aortic valve arise from 2.5 to 3.5 cm², and the validity of this formula remains unaltered more than sixty years after the first application [5]. Echocardiography is an important and reliable diagnostic tool when professionally performed, since it allows the evaluation of the presence and of the severity of AS, and other concomitant anomalies [6]. Cardiac catheterization is feasible as well, but the valve might be impossible to cross when critically diseased, and the risk of embolic stroke following the procedure cannot be underemphasized [7].

2. Case Report

3. Discussion

We dealt with a case of syncope in a previously young adult, an episode that was clearly stress-provoked (during a football game). The differential diagnosis of syncope is extremely detailed and out of the scope of the present paper; however, the clinical and radiological images of a combined BAV, AS, post-stenotic dilation of aorta, and coarctation were highly suggestive of a serious medical condition. Such a combination (BAV, AS, dilation of aorta and coarctation) is reported in the casuistics, and mutual influences in between the valve morphological characteristics with the presence, severity and outcome of aortic dilation or coarctation have been scrutinized [15,16].

Asymptomatic AS is not a rare condition; from the moment AS becomes symptomatic, authors suggest an average survival of less than 2-3 years. Indications for surgery are angina, CHF and syncope, but treating clinicians should take into account the clear influence and correlation between AS and the risk of sudden cardiac death [17]. Another controversy when detecting occasionally an asymptomatic AS is related with the unclear character of the progression of the disorder [18]. Worries regarding sports activity in patients with BAV and the potential of the latter for unleashing potentially fatal cardiac disorders have been formulated [19].

In all cases, a careful monitoring is warranted, particularly in patients with previous fainting episodes, with a positive family history for aortic diseases, or in particular physiological situations such as pregnancy [20]. In a recent publication of our centre's data, we compared the results of aortic surgery between two subgroups of patients, the second of which had a simultaneous coronary artery bypass graft; meanwhile the first subgroup had aortic valve surgery alone, concluding that a combined surgical intervention did not affect the long-term results of either from the procedures [21]. When it comes to surgical approaches, there is however a consistent trend of specialized facilities in reporting better results, with increasingly lower morbidity and mortality, even for interventions of a major importance, such as the aortic surgery. This optimistic overview has led to conceptual changes as well: fast-track elective open aortic surgery, with minimally invasive surgery and anesthesia, early oral feeding and ambulation, is advocated as another valid option, with similar good results [22].

Abbreviations

AS (aortic stenosis); **LVH** (left ventricular hypertrophy); **BAV** (bicuspid aortic valve); **CHF** (cardiac heart failure).

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