Case study: coronary artery disease in bicuspid aortic valve

Studio di un caso: coronaropatia nella valvola aortica bicuspe

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Background. Bicuspid aortic valve (BAV) occurring in 1% to 2% of the population increases the risk of aortic valve dysfunction and the most frequently observed pathologies associated with BAV are aneurysms of the aorta and acute dissection.

Aim. We describe a possible pathogenetic role of BAV when it is associated with coronary artery disease.

Description of the case. 66-year-old man with BAV associated with left ventricular hypertrophy and coronary artery disease.

Conclusion. A role of the BAV in the pathogenesis of the death can be mediated by ventricular hypertrophy and ischemic heart disease resulting in cardiac failure.

Key words: Cardiac death, Bicuspid aortic valve, Coronary artery disease

INTRODUCTION

Bicuspid aortic valve is a disease with variable clinical presentation and complications. It increases the risk of aortic valve dysfunction, but in the literature are described associations of BAV both with aortic stenosis and LV hypertrophy than with aortic regurgitation and dilated left chambers. Nigam et al. suggest that the evolution towards aortic stenosis or aortic insufficiency is due to modulate gene expression by microRNAs. But this shows the multiple pathological findings of the disease.

The pathogenetic role of BAV, when it is associated with other diseases, it has a synergistic effect in the pathophysiology of heart failure. BAV associated with coronary artery disease (CAD) is described only as possible chronic complication after a cardiac allotransplantation and univariate predictor of death in bicuspid aortic valve-sparing root replacement.

We present the autoptic outcome of a 66-year-old man with BAV associated with LV hypertrophy and coronary artery disease (CAD) and through a literature review we explain its possible role.

CASE REPORT

A 66-year-old man, the length of 183 cm and weighing approximately 90 kg (BMI: 26,86). No history of hypertension, diabetes or other diseases. Only recently palpitations. Adipose tissue with a regular distribution and normal muscle tropism. Skull without appreciable preternatural motility of the bone surface. Slightly globular abdomen for fat. Dissected the soft parts we founded the integrity of the rib cage, diaphragm and bowel loops with the abdominal cavity dry.
Removed the sternal plastraon lungs were expanded, without adhesions or liquid in the pleural cables. Pericardium were intact and to the opening of which were found fex cc of serous fluid. Heart of globular form (480 g weight, 11 cm longitudinal diameter, 12 cm transverse diameter, 5 cm anteroposterior diameter). The lumen of the left coronary in the proximal portion of anterior descending artery were stenotic with calcified walls. The thickness of the apical and free wall of the left ventricle were approximately 2 cm. The thickness of the interventricular septum 2.5 cm. To the section of the rest of the heart with transverse sections, whitish discolored area was at the level of the septal myocardium and anteromedial wall of the left ventricle (Fig. 1). At the opening of the outflow way of the LV was only two cusps instead of three (Fig. 2). Lungs with increased dimensions and consistency (right lung weight 890 g - left 800 g) and crackling. Large, medium and small bronchi with mucosal hyperemic and slightly raised in folds. To the full-thickness section of lungs parenchyma was congested, a finding corroborated by squeezing with frothy bleeding.

**DISCUSSION**

A pathogenetic role of BAV in the systolic overload of the LV can be explained by valvulopathy. Studies about the dynamics of the bicuspid aortic root indicate that the bicuspid aortic valve is morphologically stenotic and the flow through it is turbulent even if there is no transvalvular pressure gradient and the valve is clinically considered “normal” 2. The development of aortic stenosis occurs in a similar fashion to that seen in patients with trileaflet leaflet calcification, initiated by endothelial dysfunction and involving inflammation, lipoprotein deposition, calcification, and ossification of the aortic side of the valve leaflets 7, but in BAV the folding and creasing of the valves and the turbulent flow are felt to contribute to development of fibrosis and calcification 8.

![Fig. 1. Discolored septal area after cardiac apex section.](image1)

![Fig. 2. Bicuspid aortic valve after opening of left ventricle.](image2)
But a study about left ventricular mechanics in cases of isolated non-stenotic BAV with non-dilated aorta showed that BAV might affect LV systolic functions in a fashion independent from the valvular dynamics and aortic elasticity then that BAV is not only a valvular disease, but possibly a ventricular disease 9. In our patient the stenotic finding was evident, and the concentric LV hypertrophy, caused by a systolic overload of the left ventricle, resulted in an increase of the volumetric size of the heart and simultaneously in a reduction of the amount of blood received into the ventricular cavity 10. However, the stenosis of the anterior descending coronary artery contributed with myocardial necrosis septal to the reduction in systolic function of the heart, accelerating the cardiac failure. A calcific stenosis that, even if tightened, was limited to a small portion of the proximal segment of the anterior descending corona-

ry artery. This suggests that if LV hypertrophy had been no, and therefore increased demand, ischemic heart disease was less severe. Mohamed et al. examined the expression of nitric oxide synthase and suggest dysregulation of the nitric oxide system in patients with bicuspid aortic valves 11. Stasis of blood in the pulmonary circulation caused a pulmonary hypertension and subsequently the edema of the pulmonary parenchyma observed at autopsy 12 13.

CONCLUSION

According to the findings of postmortem examination, we can not exclude a role of the BAV in the pathogenesis of the death because the ischemic heart disease resulting in cardiac failure affected a ventricle hypertrophied by BAV.

La valvola aortica bicuspide che interessa l’1-2% della popolazione incrementa il rischio di disfunzione della valvola aortica e le patologie più frequentemente osservate associate alla valvola aortica bicuspide sono l’aneurisma e la dissecazione acuta dell’aorta. Gli Autori descrivono un possibile ruolo patogenetico della valvola aortica bicuspide quando risulta associata a coronaropatia. Il caso riguarda un soggetto di sesso maschile di 52 anni con valvola aortica bicuspide associata ad ipertrofia ventricolare sinistra e coronaropatia. Il ruolo della valvola aortica bicuspide nella patogenesi dell’evento morte può essere mediato dall’ipertrofia ventricolare e dalla cardiopatia ischemica con conseguente insufficienza cardiaca.

Parole chiave: Morte cardiaca, Valvola aortica bicuspide, Coronaropatia

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