Case Report

Quadricuspid Aortic Valve by Transthoracic Echocardiography

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A 55-year-old woman seeks medical attention with palpitations, atypical chest pain and dyspnea upon exertion, lasting for 12 months. ECG showed left ventricular hypertrophy.

Transthoracic Doppler echocardiogram confirmed hypertrophy of the left chambers and showed a quadricuspid aortic valve, with equal-sized cusps, in addition to a moderate regurgitant central flow.

Upon echocardiography, the quadricuspid valve is identified by its characteristic “X” form during diastole and rectangular aspect during systole. Heart failure symptoms are presumably caused by valve insufficiency due to abnormal leaflet coaptation.

Considering the possibility of progressive regurgitation, follow up on the patient was performed with periodic echocardiographic control. Symptoms responded to treatment.

Discussion

Semilunar valve alterations are among the most common cardiac malformations. In contrast to bicuspid aortic valve – present in 2% of population – the finding of a quadricuspid aortic valve is extremely rare, with an incidence between 0.008 and 0.033%¹, ³, ⁴.

The first report of a quadricuspid aortic valve, in 1862, comes from necropsy data; and the first echocardiographic finding is from 1984³. Since then, the frequency of this diagnosis has been increasing.

Normally, it presents itself as an isolated condition but may be associated with interventricular septum defect, patent ductus arteriosus, coronary ostium alterations and other valvular defects as well¹, ⁴.

Aortic regurgitation, caused by abnormal leaflet coaptation, is common. Some authors suggest fibrous thickening¹, ⁴, while others propose unequal distribution of transvalvar stress – because of the fourth cusp¹ –, as the pathophysiological mechanism for valvular insufficiency. Stenosis is rare.

On echocardiography, the quadricuspid valve is identified by its characteristic “X” configuration during diastole (different from the “Y” of normal tricuspid aortic valve) and rectangular aspect during systole¹, ³, ⁴. According to the Hurwitz and Roberts⁴ classification, considering the size of leaflets, only 12% of quadricuspid aortic valves have four equal-sized leaflets.

There is no agreement about which, transthoracic or transesophageal, is the best echocardiographic technique for this diagnosis. However, it is known that the use of Doppler has made the examination more sensitive in

Key words

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Figure 1 - Quadricuspid aortic valve during systole, showing the typical rectangular aspect (A); and during diastole, demonstrating the “X” configuration of the four closed leaflets (B).

Figure 2 - Doppler image of regurgitant flow.

demonstrating the retrograde flow through the valve, even when there is no clinical repercussion, facilitating early diagnosis of aortic regurgitation.

Taking into account the technological development of echocardiographic equipment, and its availability as a non-invasive diagnostic tool, it is feasible to detect, follow and diagnose possible complications of quadricuspid aortic valves even in asymptomatic individuals.

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References


