Accessory mitral valve as cause of left ventricular obstruction in the adult

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A mitral valve tissue is a rare congenital cardiac malformation and an uncommon cause of left ventricular outflow tract (LVOT) obstruction. To our knowledge, there are about 50 reported cases in the medical literature, most of them diagnosed during childhood in association with other congenital heart defects. In the adult patient, accessory mitral valve is an uncommon feature. We report the case of a 43-year-old man, referred for operation on a subaortic membranous stenosis diagnosed by transthoracic echocardiography (TTE), in whom an obstructive accessory mitral valve was identified during the surgical procedure.

Clinical Summary

A 43-year-old Caucasian man was referred to our center with the diagnosis of membranous subaortic stenosis, which had been made

by TTE. He reported short episodes of palpitations since his 30s, without other symptoms. On physical examination, a grade III/VI harsh systolic murmur, radiating to the neck, was audible at the precordium.

The electrocardiogram showed sinus rhythm and left ventricular hypertrophy with strain pattern. The Doppler echocardiogram demonstrated the presence of a subaortic obstructive membrane (maximum and median gradients of 81 mm Hg and 52 mm Hg,

respectively) and concentric left ventricular hypertrophy. Catheterization with left ventriculography confirmed the diagnosis of LVOT obstruction, and a peak gradient of 62 mm Hg was measured.

The patient underwent cardiac surgery on July 21, 1999. The ascending aorta was opened, and the LVOT was inspected through the aortic valve. A tiny subaortic membrane was visualized and excised. On looking deeper into the LVOT, an abnormal structure was found that looked like mitral leaflet tissue. It was attached to the mitral annulus and to the mitral chordae tendineae by two short chords (Figure 1). When hooked by its free edge, the valve opened in parachute, occluding the LVOT.

That structure was excised, and no intraoperative gradient was registered between the left ventricle and the aorta after the patient was weaned from bypass. Histologic examination of the specimen confirmed it to be valve tissue. A TTE performed 3 months later did not disclose any residual LVOT obstruction.

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Figure 1. A, Aortic aspect. The central part (corresponding to area of attachment to the mitral annulus) is darker in color. On left side, two short cords can be seen. Right portion has a more or less normal valve appearance. B, Ventricular aspect. Typical valve anatomy can be seen.

Discussion

The first case of accessory mitral valve was published in 1959 by MacLean and associates.¹ Since then there have been about 50 cases reported in the medical literature.

A review of the published data showed the following:

- In about two thirds of the cases, accessory mitral valve was associated with other cardiac anomalies, and symptoms had developed in the neonatal or early childhood period.²
- LVOT obstruction is usually a mark of this anomaly; in fact, only 3 reported cases involved nonobstructive forms.³

The widespread use of 2-dimensional echocardiography with color Doppler has greatly improved the recognition of accessory mitral valve tissue. When a discrete subaortic stenosis is also present, however, the accessory valve may pass unnoticed.⁴ During the surgical procedure, because the heart is empty and relaxed, it may even happen that the anomalous valve is missed and a second operation will be needed to relieve the residual obstruction.⁵

In this case the presence of the accessory mitral valve was not recognized by TTE and left ventriculography, and the tiny subaortic membrane was considered to be responsible for the pressure gradient registered between the left ventricle and the ascending

aorta. Although we do not know the relative weights of each of the structures involved in the gradient value registered through the LVOT, we believe, because of the small size of the subaortic membrane, that the accessory mitral valve actually played the major role.

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