Total relief of severe left ventricular outflow obstruction after spontaneous rupture of chordae tendineae in a patient with hypertrophic cardiomyopathy

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In hypertrophic cardiomyopathy (HCM), rupture of mitral chordae tendineae is infrequent and causes acute haemodynamic deterioration. A 38 year old male patient had chordae rupture leading to prolapse of both mitral leaflets and severe regurgitation, without change in symptomatic status. One year before, he had mild mitral regurgitation and a resting left ventricle outflow tract of 105 mm Hg that disappeared in the present evaluation. In this unique case, worsening of mitral regurgitation was counterbalanced by total relief of the severe obstruction. This case report highlights the role of the mitral valve apparatus in the genesis of obstruction in HCM, further stimulating surgical techniques in which mitral repair can be the main procedure.

DISCUSSION

In HCM, ruptured mitral chordae tendineae is an infrequent complication. Few single case reports can be found in the literature1; five cases reported by Zhu et al2 are of note. The rupture can be spontaneous or secondary to infective endocarditis and is generally followed by acute haemodynamic deterioration requiring immediate surgical intervention.

The present case has unique features. Unlike in previously reported cases, the present patient’s symptoms did not worsen. The rupture caused prolapse of the medial portions of both mitral leaflets, originating a curious bidirectional regurgitant jet and, importantly, completely relieving the severe LVOT obstruction. We can speculate that the clinical status was unchanged because the overload imposed by the regurgitation was counterbalanced by the disappearance of the obstructive gradient.

In HCM, the mitral valve apparatus has a critical role in the genesis of LVOT obstruction. A constellation of structural malformations can be present such as enlarged or elongated...
leaflet and abnormal papillary muscle insertions. Coexistence of mitral valve prolapse and HCM is considered infrequent (3%).

The best surgical procedure to alleviate obstruction in HCM has been the subject of debate. Septal myectomy (Morrow’s surgery), combined myectomy and mitral surgery (repair or replacement), and mitral surgery alone have been the most commonly performed. The choice of the appropriate technique is based on careful preoperative and transoperative morphological evaluation, in addition to the surgeon’s preference.

In view of the present case, in which the mitral valve seemed to be the principal and perhaps the only mechanism of obstruction, surgeons must keep in mind the possibility of relieving the obstruction through a single valve repair in selected cases of obstructive HCM.

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