CASE REPORTS

Anterior leaflet on the leash: unusual cause of congenital severe mitral regurgitation

Piotr Hoffman, Maria Kordybach*, Jacek Różański, Mariusz Kuśmierczyk, and Mirosław Kowalski

Institute of Cardiology, Alpejska 42, 04-628 Warsaw, Poland

Received 14 December 2008; accepted after revision 7 February 2009; online publish-ahead-of-print 26 February 2009


Introduction

Isolated mitral regurgitation is infrequently found in adults and accounts for <1% of congenital anomalies of the heart. Transthoracic echocardiography remains the most useful diagnostic tool allowing precise definition of anatomy, function, and sequelae of the lesion. We report a young female patient with a 10 years history of mitral regurgitation in whom transthoracic echocardiography showed an extraordinary valve anomaly leading to severe regurgitation.

Case report

A 23-year-old female student, with a history of congenital mitral regurgitation was referred to Congenital Heart Disease Department for the evaluation of valvular disease. She has remained entirely asymptomatic for many years. Her exercise tolerance was very good allowing riding bicycle and other unrestricted physical activities. However, recently she noted post-exercise palpitation and shortness of breath.

A physical examination revealed vivid pulsation of the cardiac apex which was shifted leftward and inferiorly. A loud systolic murmur (grade III/IV) was audible at the apex radiating to the left. Neither signs of pulmonary congestion nor any other symptoms of overt heart failure were found.

A 12-lead ECG showed normal sinus rhythm with a 1 mm depression of the ST-segment in the V4–V6 leads.

A transthoracic echocardiography revealed grossly abnormal mitral valve. Thickened anterior leaflet of the mitral valve was hooked to the left atrial roof by a thick thread tissue of the same echogenicity as leaflet tissue and normal chordae tendineae (Figure 1). This was attached to the anterior mitral leaflet in the A2 region. In systole, the additional structure was pulling the anterior leaflet into the left atrial cavity.

Figure 1. A thick fibrotic tissue (arrow) connecting left atrial roof and the anterior leaflet of the mitral valve. Transthoracic echocardiography—parasternal, long axis.

* Corresponding author. Tel: +48 22 343 4263; fax: +48 22 343 4538. E-mail address: mkordybach@iakard.pl

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2009. For permissions please email: journals.permissions@oxfordjournals.org.
precluding proper coaptation of the leaflets. This mechanism was responsible for severe valvular incompetence. Colour Doppler showed regurgitant flow towards the inferior wall of left atrium and depressed systolic wave in pulmonary veins. Effective regurgitant orifice was 0.9 cm². Left ventricle (LV) was dilated (LVEDD 69 mm, LVESD 40 mm) as well as the left atrium (LAD 50 mm). LV function was well preserved (EF 72%). Mild tricuspid regurgitation enabled the evaluation of RVSP which amounted to 46 mmHg.

Because of recent onset of symptoms (dyspnea and palpitation during exertion), the patient was referred to Cardiosurgery Department, where repair of the valve was performed. Mechanism of the regurgitation was fully confirmed and abnormal chorda was resected. There was still mitral valve incompetence in the water test due to anterior leaflet prolapse. Three artificial chordae were placed to anterior leaflet and a 28 mm Edwards Lifesciences Annuloplasty Mitral Ring was implanted. Competent valve was obtained in the water test.

A post-surgery transthoracic echocardiography showed perfect function of the mitral leaflets without regurgitation. Moreover, the LV and left atrium dimension returned to normal (LVEDD 56 mm, LVESD 33 mm, LAD 32 mm). On a post-operative echo study, RVSP was 35 mmHg. The patient was discharged 10 days after surgery. In a few months follow-up, she remains in excellent condition.

Reference