Dynamic mild subaortic left ventricular obstruction caused by an accessory mitral valve attached to the anterior mitral valve in a young pregnant woman

Antonio D’Aloia, Enrico Vizzardi*, Ermanna Chiari, Francesco Fracassi, Gregoriana Zanini, Pompilio Faggiano, and Livio Dei Cas

Unità Operativa di Cardiologia, Spedali Civili, Brescia e Cattedra di Cardiologia, Università di Brescia, Brescia, Italy

Received 24 April 2007; accepted after revision 20 May 2007; online publish-ahead-of-print 23 August 2007

Accessory mitral valve tissue is an extremely rare congenital cardiac anomaly of embryologic development of the endocardial cushion. This anomaly is often associated with left ventricular outflow tract obstruction (LVOTO). A 26-year-old pregnant female was referred to our Department of Cardiology with exertional shortness of breath and tachycardia. Transthoracic and transesophageal echocardiography revealed a flexible circular (1.3 × 1.4 cm), mobile structure attached to the ventricular side of anterior mitral valve leaflet, with chordal attachments structure from anterior papillary muscle. This picture is compatible with a parachute-like accessory mitral valve tissue. We performed an echocardiographic exercise test that shows a systolic flow turbulence starting immediately proximal to this structure, resulting in a small increase in left ventricular outflow tract (LVOT) gradient (30 mmHg). Therefore we started low dose of beta-blocker therapy in order to decrease heart frequency and reduce the future risk of a worsening of an LVOT dynamic obstruction.

Transthoracic and transesophageal echocardiography is critical for the differential diagnosis of LVOT and in the management of accessory mitral valve tissue. In patients without rest and only an exertional mild LVOTO and no other cardiac malformations, prophylactic removal of mitral accessory tissue excision is not required; antibiotic prophylaxis for endocarditis can be indicated and a regular follow-up is recommended to identify any progression in LVOTO entity.

**KEYWORDS**
Subaortic left ventricular obstruction; Accessory mitral valve; Pregnancy

**Background**

Accessory mitral valve tissue is an extremely rare congenital cardiac anomaly of embryologic development of the endocardial cushion firstly described by McLean in 1963.1 This anomaly is often associated with left ventricular outflow tract obstruction (LVOTO)1,2 and, most rarely, with interventricular septal defect.3 The obstruction can occur in the early period of life as a result of mass effect due to continued deposition of fibrous tissues within the left ventricular outflow tract (LVOT). The accessory mitral valve tissue is usually diagnosed in the first or second decade of life and the main symptoms are exercise intolerance with dyspnoea, chest pain and syncope.2

An accurate echocardiographic evaluation of this entity and assessment of its hemodynamic consequences are very important for an appropriate therapeutic decision to be made.

**Case report**

A 26-year-old pregnant female was referred to our Department of Cardiology with exertional shortness of breath and tachycardia. She was a normally developed young woman, without history of heart disease. At admission blood pressure was reduced (100/70 mmHg), with moderately high heart rate (100 beat/minute). At auscultation an ejection type, 2/6 systolic murmur was heard. Biochemical and hematological parameters were in the normal range. The electrocardiogram showed sinus tachycardia (100'). Chest X-ray was normal.

Transthoracic and transesophageal echocardiography revealed a flexible circular (1.3 × 1.4 cm), mobile structure attached to the ventricular side of anterior mitral valve leaflet (AML), with chordal attachments structure from anterior papillary muscle. This picture is compatible with
a parachute-like accessory mitral valve tissue, without cleft in the AML. Other echocardiographic findings were normal except for a mild mitral regurgitation, and sign of LVOTO (rest normal LVOT pressure gradient) was not detected (Figures 1 and 2, movie 1 and 2).

We performed an echocardiographic exercise test in order to exclude a dynamic and worsening of LVOTO. At color Doppler examination, there was a systolic flow turbulence starting immediately proximal to this structure, resulting in as small increase in LVOT gradient (30 mmHg), but without symptoms.

Therefore we started low dose of beta-blocker therapy in order to decrease heart frequency and reduce the future risk of a worsening of the LVOT dynamic obstruction.

Discussion

Accessory mitral valve tissue is a congenital cardiac anomaly that often manifests itself in the first decade of life, and usually requires surgical reparation. Clinical manifestations are exercise intolerance, chest pain, syncope, tachycardia and they are usually due to left ventricular outflow tract obstruction.\(^2\) Rarely it can be associated to thromboembolic accidents (two cases are described in literature in the same patient).\(^4\) Transthoracic and transesophageal echocardiography is critical for the differential diagnosis of LVOT and in the management of accessory mitral valve tissue.\(^5,6\) Surgical removal is indicated in patients presenting significant LVOTO, even if they are asymptomatic, in patients ongoing correction of other congenital malformations or exploration of an intracardiac mass.\(^2\) In our patient we did not find a significant LVOTO and no other abnormalities were present.

In conclusion in patients without rest and only an exertional mild LVOTO and no other cardiac malformations, prophylactic removal of mitral accessory tissue excision is not required; antibiotic prophylaxis for endocarditis can be indicated\(^6\) and a regular follow up is recommended to identify any progression in LVOTO entity.

Supplementary material

Supplementary data associated with this article can be found in the online version.

References