Rev Port Cardiol. 2015;34(5):357.e1–357.e5

Revista Portuguesa de Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

CASE REPORT

High left ventricular outflow tract gradient: Aortic stenosis, obstructive hypertrophic cardiomyopathy or both?☆

Inês Almeida*, **Francisca Caetano, Joana Trigo, Paula Mota, António Leitão Marques**

Serviço de Cardiologia, Centro Hospitalar e Universitário de Coimbra – Hospital Geral, Coimbra, Portugal

Received 31 July 2014; accepted 10 October 2014
Available online 18 May 2015

KEYWORDS
Aortic valve stenosis;
Hypertrophic cardiomyopathy;
Left ventricular outflow tract obstruction;
Echocardiography;
Doppler echocardiography

Abstract The authors report the case of a patient diagnosed with both hypertrophic cardiomyopathy and aortic stenosis. Due to clinical deterioration, additional investigation was performed, and a high left ventricular outflow tract gradient was identified. Correct identification of the condition causing the symptoms was challenging, and involved several imaging techniques, the contribution of transesophageal echocardiography being crucial. The final diagnosis of severe aortic stenosis led to successful valve replacement surgery. The presence of these two conditions in the same patient has been documented, although it is uncommon. This association poses particular diagnostic and therapeutic challenges, which are discussed in this paper.

PALAVRAS-CHAVE
Estenose aórtica;
Cardiomiopatia hipertrófica;
Obstrução do trato de saída do ventrículo esquerdo;
Ecocardiografia;
Ecocardiografia *Doppler*

Gradiente elevado no trato de saída do ventrículo esquerdo: estenose aórtica, miocardiopatia hipertrófica obstrutiva ou ambas?

Resumo Os autores apresentam o caso de uma doente com os diagnósticos de miocardiopatia hipertrófica e estenose aórtica, na qual foi identificada a presença de um gradiente elevado ao nível do trato de saída do ventrículo esquerdo. O reconhecimento da patologia responsável pela sintomatologia foi desafiante, com envolvimento de várias técnicas de imagem, tendo sido fundamental a contribuição do ecocardiograma transesofágico. O diagnóstico final de estenose aórtica severa conduziu à referenciação para cirurgia de substituição valvular, com sucesso. A presença destas duas patologias em simultâneo num mesmo doente é conhecida, embora

☆ Please cite this article as: Almeida I, Caetano F, Trigo J, et al. Gradiente elevado no trato de saída do ventrículo esquerdo: estenose aórtica, miocardiopatia hipertrófica obstrutiva ou ambas? Rev Port Cardiol. 2015;34:357.

* Corresponding author.
E-mail address: inesalm@gmail.com (I. Almeida).

incomum. A sua combinação cria importantes desafios diagnósticos e terapêuticos, os quais serão objeto de discussão neste artigo.

Introduction

Aortic stenosis (AS) and hypertrophic cardiomyopathy (HCM) are two conditions that can cause hemodynamic gradients in the left ventricular outflow tract (LVOT).[1] In both cases the presence of significant obstruction has clinical, therapeutic and prognostic implications.[1–3]

The presence of both of these conditions in the same patient has been documented, although it is uncommon. This association poses particular diagnostic and therapeutic challenges.[4] Meticulous echocardiographic assessment is required for correct identification of the cause of the obstruction,[5] although this can be complicated, and the result can lead to different therapeutic options.[6,7]

This case report aims to discuss the complexity of such cases.

Case report

A 68-year-old woman, with a history of hypertension, dyslipidemia, obesity and breast cancer (treated by left mastectomy and adjuvant chemotherapy and radiotherapy in 1998), was referred for cardiology consultation in April 2011 to investigate chest pain; she had no other cardiovascular symptoms. On physical examination, auscultation revealed a grade III/VI systolic murmur audible at the right second intercostal space, crescendo-decrescendo and radiating to the carotids; the murmur became less intense with the Valsalva maneuver and on standing up, and increased with squatting.

A previous electrocardiogram had shown sinus rhythm with voltage criteria for left ventricular hypertrophy without overload (Figure 1), while transthoracic echocardiography (TTE) (described as ''technically very difficult'') had revealed concentric hypertrophy of the left ventricle (LV) with no wall motion abnormalities and with preserved global systolic function and a calcified aortic valve (AV) with moderate stenosis (mean left ventricle/aorta [LV/Ao] gradient of 21 mmHg).

Given the patient's low pretest probability of coronary artery disease, coronary computed tomography angiography was performed, which identified mild coronary calcification (calcium score 54 Agatston units) with no endoluminal obstruction, and also revealed thickening (22 mm) of the interventricular septum (IVS). Suspicion of HCM prompted investigation by magnetic resonance imaging (MRI), which confirmed the diagnosis of asymmetric HCM with hypertrophy of the basal and mid IVS (22 mm), all other walls being of normal thickness; non-dilated LV with ejection fraction of 69% and LV mass index of 78 g/m^2; moderately dilated left atrium (area 33 cm^2); and no late gadolinium enhancement (Figure 2). The patient presented no risk factors for sudden cardiac death and genetic study for Fabry disease was negative; screening for classic mutations in sarcomere protein genes is in progress.

At 18-month follow-up she presented worsening functional capacity with dyspnea on moderate exertion (New York Heart Association class II). TTE was repeated and showed marked LV hypertrophy of the basal IVS and good global systolic function; an apparently tricuspid AV, calcified, with reduced opening, that could not be assessed by planimetry; and a calcified mitral valve with systolic anterior motion (SAM). Doppler study revealed accelerated flow beginning in the LVOT, with peak velocity at mid-systole and peak and mean LV/Ao gradient of 49 mmHg and 32 mmHg, respectively, supporting the hypothesis of a fixed obstruction (Figure 3). No late-systolic velocity peak was observed, with or without the Valsalva maneuver.

In view of the limitations of TTE, transesophageal echocardiography (TEE) was performed, which revealed a malformed AV with marked calcification and fusion of the noncoronary and left coronary leaflets, with an area estimated by planimetry of 0.6 cm^2 (0.27 cm^2/m^2) (Figure 4). Color Doppler study clearly differentiated laminar flow in the LVOT and turbulent flow through the AV throughout systole, confirming the suspicion of obstruction of the valve only (Figure 4).

Invasive hemodynamic study showed a peak-to-peak LV/Ao gradient of 52 mmHg and no intraventricular gradient, and excluded significant coronary artery disease.

A 22-mm Medtronic Hall mechanical valve was implanted surgically in aortic position. At six-month follow-up the patient presented improved functional capacity and TTE revealed a normally functioning aortic valve.

Discussion

This case report highlights the difficulties of investigating a patient with both HCM and AS, particularly in assessing the severity of each condition and determining which is functionally more important. Identifying the cause of the high LVOT gradient as AS led to the patient being referred for valve replacement surgery, which resolved the obstruction and improved symptoms.

Assessment of such patients is based on a thorough echocardiographic assessment of the LVOT region.[4,5] Color and pulse wave Doppler study are essential to locate the