Significant biventricular obstruction in non-syndromic hypertrophic cardiomyopathy

Iranna Hirapur, Vikram B Kolhari, Navin Agrawal, Ravindran Rajendran

Department of Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bangalore, Karnataka, India

Correspondence to

Dr Navin Agrawal,
drnavinagrwal@gmail.com

Accepted 9 September 2014

DESCRIPTION

Although hypertrophic cardiomyopathy (HCM) is classically considered a disease of the left ventricle (LV), we present a rare case of biventricular HCM presenting in middle age.

A 57-year-old man without a family history of HCM and sudden death presented with presyncope and New York Heart Association (NYHA) class II breathlessness. Clinical examination was normal except for ejection systolic murmur. ECG showed biventricular hypertrophy with deep T-wave inversions. Echocardiography demonstrated pronounced asymmetric LV hypertrophy and thickened right ventricle (RV) muscular components (figure 1A–C; videos 1–3). Doppler revealed a peak LV mid-cavity gradient of 109 mm Hg and subvalvar obstruction of the RV outflow tract with a peak gradient of 138 mm Hg (figure 2A–C). The patient underwent successful surgical myectomy and has been stable during the course of follow-up. The histopathological

Video 1 Parasternal long axis view showing biventricular hypertrophy.

specimen revealed myofibril disarray as is expected in a case of HCM (figure 3). Genetic testing for Noonan's syndrome and three of the most common mutations were performed which were negative.

Figure 1 (A) Parasternal long-axis view showing asymmetric septal hypertrophy. (B) Parasternal short-axis view showing biventricular hypertrophy. (C) Parasternal long-axis view showing hypertrophied muscle bundles in right ventricular (RV) outflow tract with colour flow turbulence (LV, left ventricle; RVOT, right ventricular outflow tract).

CrossMark

To cite: Hirapur I, Kolhari VB, Agrawal N, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-206271

Video 2 Parasternal short axis view at mid cavity level showing significant bi-ventricular hypertrophy.

Figure 3 Photograph of the histopathological specimen showing myofibril disarray.

Video 3 Parasternal short axis view at the level of right ventricular outflow showing turbulence in the color Doppler signal across the RVOT suggesting RV outflow obstruction.

Figure 2 (A) M-mode across left ventricle (LV) showing significant biventricular hypertrophy. (B) Continuous wave Doppler showing an LV mid-cavity gradient. (C) Continuous wave Doppler across right ventricular outflow tract showing significant gradient.

Learning points

- ▶ Biventricular obstruction can occur in cases of non-syndromic hypertrophic cardiomyopathy, which should be kept in mind while evaluating the patient. Rare cases may present very late in the course of the disease, as in our case.
- ▶ The occurrence of significant right ventricular outflow obstruction can alter the management of the patient and can make percutaneous management extremely challenging and the results sometimes unsatisfactory.
- ▶ Surgical correction can provide a lasting outcome where myectomy is performed in both the chambers.
- ▶ The effect of the surgery on conduction blocks in cases with biventricular hypertrophy needs to be evaluated and can complicate an otherwise haemodynamically successful procedure. An extra bit of caution needs to be taken.

Others were not performed due to financial constraint on the part of the patient.

Biventricular HCM and valvular pulmonary stenosis is relatively common in Noonan's syndrome. However, most cases present early and this condition is rare in adults without a

dysplastic pulmonary valve.^{1 2} It appears that biventricular HCM in young patients can initially be localised to only the RV.³ In contrast to LV outflow obstruction, which is characteristically dynamic and due to mitral valve systolic anterior motion, RV obstruction is the result of a static and fixed impediment by excessive hypertrophy of muscular components. RV cardiomyopathy may or may not correlate with LV. While the genetics of RV involvement have not been well characterised, histological findings appear similar to those in the LV, suggesting similar pathogenesis. Surgical correction often gives successful and lasting outcome.⁴

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Hayashi S, Tojyo K, Uchikawa S, *et al.* Biventricular hypertrophic cardiomyopathy with right ventricular outflow tract obstruction associated with Noonan syndrome in an adult. *Jpn Circ J* 2001;65:132–5.
- 2 Chirillo F, Zecchel R, Stritoni P. Biventricular hypertrophic cardiomyopathy with alone obstruction to right ventricular outflow. *Heart* 2002;87:565.
- 3 Fournier C, Bache R, Valette H, *et al.* Hypertrophic myocardiopathy with isolated obstruction of the right ventricle. *Ann Cardiol Angeiol (Paris)* 1985;34:71–4.
- 4 Maron BJ, McIntosh CL, Klues HG, *et al.* Morphologic basis for obstruction to right ventricular outflow in hypertrophic cardiomyopathy. *Am J Cardiol* 1993;71:1089–94.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow