Indolent cardiac angioma mimicking hypertrophic obstructive cardiomyopathy and causing right ventricular outflow tract obstruction

Martin R. Brown1,2*, Andrew Dettrick3, George Javorsky1, Scott C. McKenzie1,2, and David G. Platts1,2

1Advanced Heart Failure and Transplant Unit, The Prince Charles Hospital, Rode Road, Chermside, Queensland 4032, Australia; 2School of Medicine, University of Queensland, Brisbane, Queensland 4072, Australia; and 3Anatomical Pathology, Pathology Queensland, The Prince Charles Hospital, Rode Road, Chermside 4032, Australia

* Corresponding author: Tel: +61 7 3139 4000; fax: +61 7 3139 4426, Email: martin_brown@health.qld.gov.au

A 20-year-old female with a 12-year history of hypertrophic obstructive cardiomyopathy (HOCM) with progressive right ventricular outflow obstruction (RVOTO) and mild left ventricular outflow obstruction (LVOTO) was referred to our institution for consideration of septal myomectomy due to symptomatic outflow tract obstruction. The patient had New York Heart Association Class III dyspnoea, abdominal bloating, nausea, and vomiting with palpitations and presyncope on standing or walking up stairs. Examination revealed sinus rhythm, blood pressure 100/60 mmHg, and normal jugular venous pressure. A loud ejection systolic murmur was present at the left sternal edge and posteriorly which was accentuated by valsalva manoeuvre. The remainder of examination was normal.

ECG demonstrated sinus rhythm, right axis deviation, right ventricular hypertrophy, and inferior Q waves. Transthoracic echocardiography (TTE) showed marked asymmetric interventricular septal hypertrophy 39 mm, RVOTO (peak/ mean) 94/51 mmHg, and LVOTO 9/4 mmHg with Doppler colour and Definity™ microsphere contrast revealing a highly vascularized septum (Panel Aa–c) (Supplementary data online, Videos 1–4). Cardiac MRI demonstrated severe septal hypertrophy and basal septum late gadolinium enhancement (Panel Ad–f).

The patient underwent an uncomplicated left and right ventricular septal myomectomy (3 cm resections). Histological analysis of the excised myocardium revealed a diffuse vascular proliferative process consistent with cardiac angioma (diffuse capillary haemangioma) (Panel Ag–i).

At follow-up 1 month later, the patient’s symptoms had resolved. TTE showed an LV ejection fraction 67%, IVSd 29 mm, and LVOT/RVOT mean gradients of 5/3 mmHg, respectively (Supplementary data online, Video 5). One year post-myomectomy, the patient was asymptomatic with unchanged gradients.

Panel A: (a) Parasternal long-axis transthoracic echocardiogram (TTE) demonstrating septal hypertrophy and (b) colour Doppler flow within hypertrophied septum (long arrow). (c) Parasternal short-axis TTE showing septum projecting into RVOT (arrow head). Cardiac MRI in four chamber (d) and short-axis (e) demonstrating asymmetric septal hypertrophy and with late gadolinium enhancement (LGE) (f). Macroscopic appearance of resected septum (g). (h) Diffuse proliferation of small vessels between cardiac myocytes (H&E, original magnification ×40). (i) CD31 immunohistochemistry highlighting vascular endothelium.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2013. For permissions please email: journals.permissions@oup.com