Multimodality imaging of isolated left ventricular apical hypoplasia

Greg Starmer*, John F. Younger, and Peter Stewart

Department of Cardiology, The Royal Brisbane and Women’s Hospital, QLD 4006, Australia

*Corresponding author. Tel: +61 402918164, Fax: +61 7 33989818, Email: g_starmer@hotmail.com

A previously well 62-year-old male presented after 7 days of worsening dyspnoea following a recent long distance flight. The electrocardiography documented atrial fibrillation and poor precordial R-wave progression. A computed tomographic pulmonary angiogram revealed pulmonary oedema and a truncated left ventricle (LV) with apical fatty infiltration (Panel A). Left ventriculography confirmed a truncated apex and severe systolic dysfunction, whilst angiography showed a small left anterior descending artery terminating at the deficient LV apex (Panels B and C). On transthoracic echocardiography, a large, normally functioning, right ventricle (RV) wrapped around the spherical LV (Panel D). Three-dimensional transoesophageal echocardiography confirmed the ‘banana-shaped’ RV and ‘apical’ origin of the papillary muscles (Panels E and F). T1-weighted and fat saturation cardiac magnetic resonance imaging outlined fat within the myocardium at the LV apex (Panels G and H). Late gadolinium enhancement suggested fibro-fatty change (Panel I).

These findings are consistent with isolated LV apical hypoplasia, a newly recognized cardiomyopathy, previously described in only 10 individuals, aged between 3 months (suggesting a congenital basis) and 50 years. Four features define this condition: (i) a truncated, spherical LV; (ii) fatty infiltration of the LV ‘apical’ myocardium; (iii) an elongated RV wrapping around the truncated LV; and (iv) apical papillary muscle insertion. The embryological pathogenesis is speculative. Seven of the 10 described patients presented either asymptotically or with mild symptoms. A single, rapidly fatal, case has been described. One group has described benign 5-year outcomes in two individuals. Our patient responded to standard heart failure therapy and remains stable 12-month post-discharge.

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2011. For permissions please email: journals.permissions@oup.com.