CASE REPORT

Idiopathic left ventricular apical hypoplasia

Ali Raza, Mohammad Waleed, Matthew Balerdi, Thanjavur Bragadeesh,
Andrew L Clark

Department of Cardiology,
Castle Hill Hospital,
East Yorkshire, UK

Correspondence to
Dr Ali Raza,
aliraza987@hotmail.com

Accepted 13 February 2014

SUMMARY

A 46-year-old man was found to have an abnormal ECG taken during a routine health and blood pressure check. His only symptom was non-specific central chest discomfort, unrelated to exertion. His ECG showed sinus rhythm, a normal axis and poor R wave progression across the chest leads and lateral T wave flattening. An echocardiogram showed a dilated left ventricle with a thin and hypokinetic septum bulging to the right. The apex was 'not well seen' but also appeared thin and hypokinetic. The right heart and valves were normal. The patient was further investigated for left ventricular hypoplasia.

BACKGROUND

There are only 22 previous cases reported. Presenting symptoms are non-specific and include chest discomfort, fatigue and shortness of breath. Patients can present with arrhythmias and other rare features, so the underlying diagnosis of idiopathic left ventricular apical hypoplasia (ILVAH) can be easily missed.

CASE PRESENTATION

A 46-year-old man was found to have an abnormal ECG taken during a routine health and blood pressure check in August 2011. His only symptom was non-specific central chest discomfort, unrelated to exertion. Blood pressure was 165/82 mm Hg, with no other abnormalities on physical examination.

INVESTIGATIONS

His ECG ([figure 1](#)) showed sinus rhythm, a normal axis and poor R wave progression across the chest leads and lateral T wave flattening. An echocardiogram showed a dilated left ventricle (LV) with a thin and hypokinetic septum bulging to the right. The apex was 'not well seen' but also appeared thin and hypokinetic. The right heart and valves were normal. His chest pains were not thought to be cardiac in origin and hypertensive medication was started. Given that the LV was dilated, he underwent diagnostic coronary angiography. In addition, given the poor-quality echocardiographic images, cardiac MRI was performed.

The MRI showed that the LV was dilated at 72 mm in diastole. The apex was dyskinetic and there was a lipomatous structure within the LV apex. The right ventricle (RV) was wrapped around the abnormal left ventricular apex ([figures 2 and 3](#)).

The patient was diagnosed as having ILVAH. A 24 h Holter showed a single salvo of four beats of ventricular ectopy.

OUTCOME AND FOLLOW-UP

Currently, the patient is under regular follow-up by the cardiology team.

DISCUSSION

Isolated left ventricular apical hypoplasia is a congenital abnormality characterised by: (1) a truncated and spherical left ventricular configuration with rightward bulging of the interventricular septum;

Figure 1 Presenting ECG at the rapid access chest pain clinic showing poor R wave progression and lateral T wave inversion.

To cite: Raza A, Waleed M, Balerdi M, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-202532

Figure 2 T2-weighted, vertical long axis two-chamber frame of steady state free precession (SSFP). Note the spherical left ventricle (A), the apex replaced by fat (B; see also figure 3 image 2), and papillary muscle arising from the abnormal apex (C).

(2) deficiency of the myocardium within the LV apex with adipose tissue infiltrating the apex; (3) both mitral papillary muscles originating in the flattened anterior apex; and (4) elongation of the RV which wraps around the deficient LV apex.¹

Figure 3 Image 1—T2-weighted, steady state free precession (SSFP), four-chamber image. Note the 'wrap-around' right ventricle (D) that occupies the space normally reserved for the left ventricular apex. Image 2—fat suppression sequence in the four-chamber view showing the left ventricular apex completely suppressed (E).

There are only 22 previous cases reported. Presenting symptoms are non-specific and include chest discomfort, fatigue and shortness of breath.² Patients can present with arrhythmias, as described by Tumabiene *et al.*³ In one patient,⁴ ILVAH was associated with patent ductus arteriosus treated in childhood. There is one case of sudden death reported in a previously well 19-year-old man,⁵ but he had other significant comorbidities including severe pulmonary hypertension. Otherwise, in most patients the condition has been discovered incidentally.

The pathogenesis of isolated left ventricular apical hypoplasia is unclear. Fernandez-Valls *et al.*² proposed that the defective LV apical development might be attributable to relatively inadequate LV to RV dilation during intrauterine organogenesis.

ILVAH is a rare condition. A small and unusually shaped LV on echocardiography should prompt the clinician to consider ILVAH. However, the correct management and the prognosis of ILVAH are unknown.

Learning points

- ▶ Idiopathic left ventricular apical hypoplasia (ILVAH) is a rare condition.
- ▶ A small and unusually shaped left ventricle on echocardiography should prompt the clinician to consider ILVAH.
- ▶ However, the correct management and the prognosis of ILVAH are unknown.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Van Hecke TE, Decker J, Leonowicz N, *et al.* Isolated left ventricular apical hypoplasia. *Congenit Heart Dis* 2011;6:646–9.
- 2 Fernandez-Valls M, Srichai MB, Stillman AE, *et al.* Isolated left ventricular apical hypoplasia: a new congenital anomaly described with cardiac tomography. *Heart* 2004;90:552–5.
- 3 Tumabiene KD, Maggali AE Jr, Chiong L, *et al.* A plump and fatty heart: isolated left ventricular apical hypoplasia. *Echocardiography* 2012;29:1540–8175.
- 4 Haffajee JA, Finley JJ, Brooks EL, *et al.* Echocardiographic characterization of left ventricular apical hypoplasia accompanied by a patent ductus arteriosus. *Eur J Echocardiogr* 2011;12:E17.
- 5 Irving CA, Chaudhari MP. Fatal presentation of congenital isolated left ventricular apical hypoplasia. *Eur J Cardiothorac Surg* 2009;35:368–9.

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