Cardiology in the Young

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Stress cardiomyopathy in dextrocardia with situs inversus and anomalous coronary arteries

David W. Baker, Yue Y. Deng and Amir-Hadi Nojoumian

Cardiovascular Department, Tamworth Hospital, Tamworth, New South Wales, Australia

Brief Report

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Author for correspondence:
David W. Baker, Cardiovascular Department, John Hunter Hospital, Lookout Road, New Lambton Heights 2305, New South Wales, Australia. Tel: (02) 49 214 200; Fax: (02) 49 214 210; E-mail: David.Baker@health.nsw.gov.au

Abstract

We present a 53-year-old male with the rare constellation of stress cardiomyopathy, dextrocardia with situs inversus and anomalous coronary anatomy. This case highlights the difficulties faced when managing patients with uncommon disorders and demonstrates a rare overlap of acquired and CHD.

Case presentation

A 53-year-old male farmer was referred to hospital by his general practitioner after an episode of chest pain and a positive point-of-care troponin. The chest pain occurred while shearing sheep the day prior and was central, tight, non-radiating, relieved by rest and associated with dyspnoea. There was mild ongoing chest discomfort the next morning.

Past medical history included dextrocardia with situs inversus, paroxysmal atrial fibrillation, polymyalgia rheumatica and bilateral shoulder repair. Three years prior, he had a normal exercise stress test and an echocardiogram showing preserved left ventricular function and normal valve function. He had no regular medications or allergies, was a non-smoker and consumed four to six standard drinks per day, living with his wife and two children.

On examination, vital signs were within normal limits. There was a right-sided non-displaced apex beat. Auscultation revealed dual heart sounds and a soft ejection systolic murmur over the left upper sternal edge. Cardiovascular examination was otherwise normal.

The initial electrocardiogram was performed with limb and chest leads not reversed for dextrocardia, showing a negative P wave and QRS complex in lead I and reverse R wave progression across the precordial leads. After appropriate re-positioning, the electrocardiogram revealed pathological anterolateral T wave inversion. Troponin was elevated at 977 ng/L. A chest x-ray confirmed dextrocardia and situs inversus (Fig 1).

The patient was admitted to the coronary care unit for telemetry and commenced on aspirin, clopidogrel, enoxaparin, atorvastatin and bisoprolol for the provisional diagnosis of non-ST elevation myocardial infarction.

Echocardiography revealed moderate left ventricular systolic dysfunction with mid to apical akinesis and basal sparing suggestive of stress cardiomyopathy. There was no significant valvular pathology.

Coronary angiography was performed via the right radial artery. A variety of catheters (Judkins, Tiger, and multi-purpose) were used due to difficult coronary ostia engagement. Anomalous coronary anatomy was identified with the left anterior descending artery and right coronary artery originating from a single ostium within the right coronary sinus. The left circumflex artery was identified only after aortogram, arising from the left coronary sinus. There was minor non-obstructive coronary disease and a mid to apical wall motion abnormality on left ventriculogram (Fig 2). A provisional diagnosis of stress cardiomyopathy was made.

A CT coronary angiogram was performed to further assess the coronary anatomy (Fig 2). It confirmed the anomalous aortic origin of a coronary artery, being the left anterior descending artery arising from the proximal right coronary artery originating from the right coronary sinus. It coursed anteriorly over the right ventricular outflow tract, down the anterior left ventricular wall, terminating in the distal interventricular groove. There was no intramural course of the left anterior descending artery. The right coronary artery was dominant and supplied a high rising acute marginal, large posterolateral, and posterior descending artery. The left circumflex ostium was in the left (posterior) coronary sinus and it supplied the posterolateral wall of the left ventricle.

The patient was discharged on perindopril 2 mg daily and bisoprolol 2.5 mg daily. At 4-week follow-up, the patient had improved markedly but still complained of mild fatigue.

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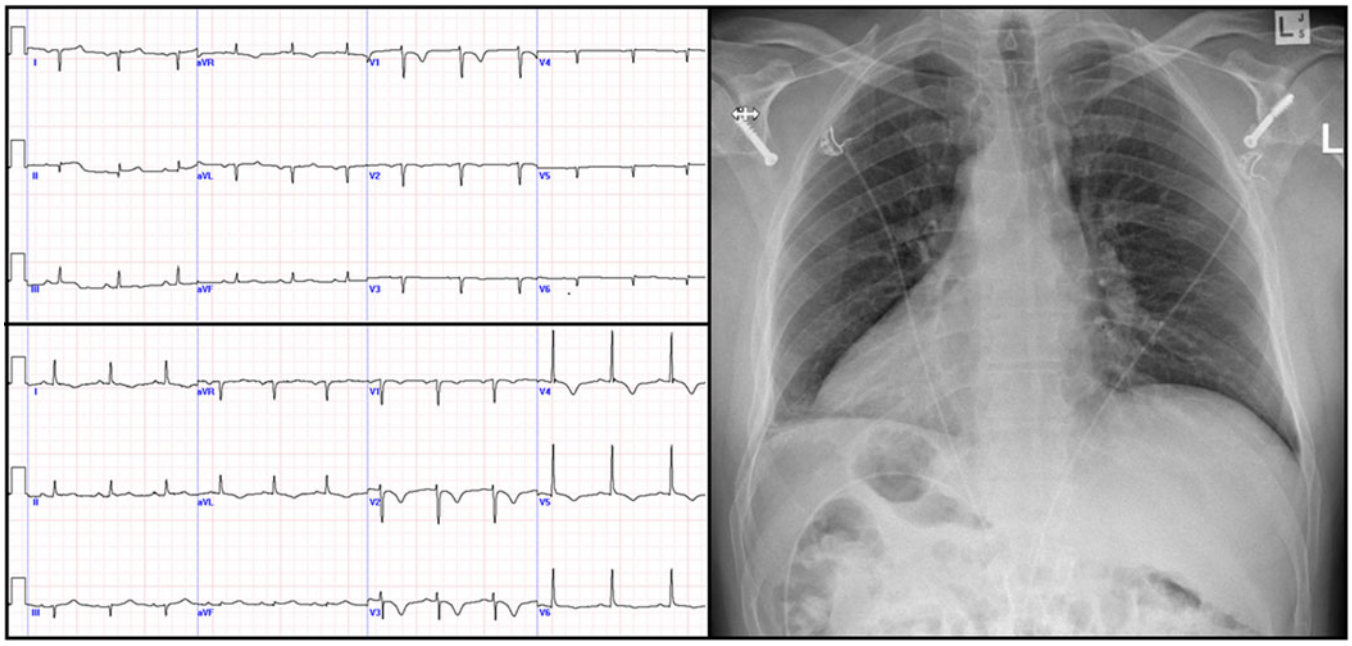

Figure 1. Electrocardiogram with standard lead positioning (top left) and then after re-positioning for dextrocardia with right precordial leads and limb lead reversal (bottom left). (Right) Chest x-ray showing dextrocardia and situs inversus.

A repeat echocardiogram at 1 month showed normalisation of left ventricular function, supporting the diagnosis of stress cardiomyopathy.

Discussion

Dextrocardia is defined as a right-sided heart secondary to abnormal embryonic lateralisation. It is a rare condition, seen in 1 in 8000 to 25,000 live births[1]. This case highlights the difficulties faced when encountering rare disorders, especially CHD.

The initial 12-lead electrocardiogram was performed without appropriate reversal of limb and chest leads, despite the diagnosis of dextrocardia being known. If the error was not corrected, as in this case, the possibility of a significant delay in diagnosis of an acute coronary syndrome exists.

Coronary angiography is more difficult in dextrocardia. The individual's anatomy, approach to vascular access, imaging strategy, and catheter selection all need to be considered. A recent review suggests that the radial approach is effective (successful in 17 of 18 cases) with low risk of significant complications. The review did not compare radial to femoral access; however, recommended femoral access only when the radial approach is unsuccessful[2]. In the normal population, radial compared to femoral approach is associated with similar efficacy and lower complication rates[3], but this should be extrapolated with caution to the dextrocardia population where abnormal anatomy may lead to longer procedures, more frequent catheter exchanges, and potentially more complications. Various coronary catheters have been reported as successful. The Judkin's catheters have been the most commonly used for diagnostic angiography[2]. There is no consensus in regards to catheter selection and should be determined by local operator experience, keeping in mind catheter manipulation is reversed. The most common fluoroscopy strategy is to mirror-image the standard left–right angulations, although rarely post-processing technology has been used to invert the images to normal[4].

Further difficulty was met in this case due to the anomalous aortic origin of a coronary artery. There is no literature describing the prevalence of anomalous coronaries in dextrocardia. It is unclear if it is increased compared to the normal population, where it has a reported prevalence of 0.64%[5]. It has been demonstrated that the risk of additional cardiac malformations is lower in dextrocardia with situs inversus as compared to situs solitus (5 versus 90%); however, coronary abnormalities were not been specifically assessed[1,6].

Stress cardiomyopathy is responsible for around 2% of patients undergoing coronary angiography for suspected acute coronary syndrome, predominantly affecting women in their sixth decade[7]. The diagnosis of stress cardiomyopathy among CHD patients is extremely rare. It has only been reported twice in dextrocardia, with both patients being situs inversus, and one having Kartagener's syndrome[8,9]. Although patients with dextrocardia and situs inversus typically have normal longevity, it is worth considering as patients with more complex CHD are surviving longer, we will encounter further overlap of CHD and acquired heart disease, namely atherosclerosis, or such as in this exceptional case, stress cardiomyopathy.

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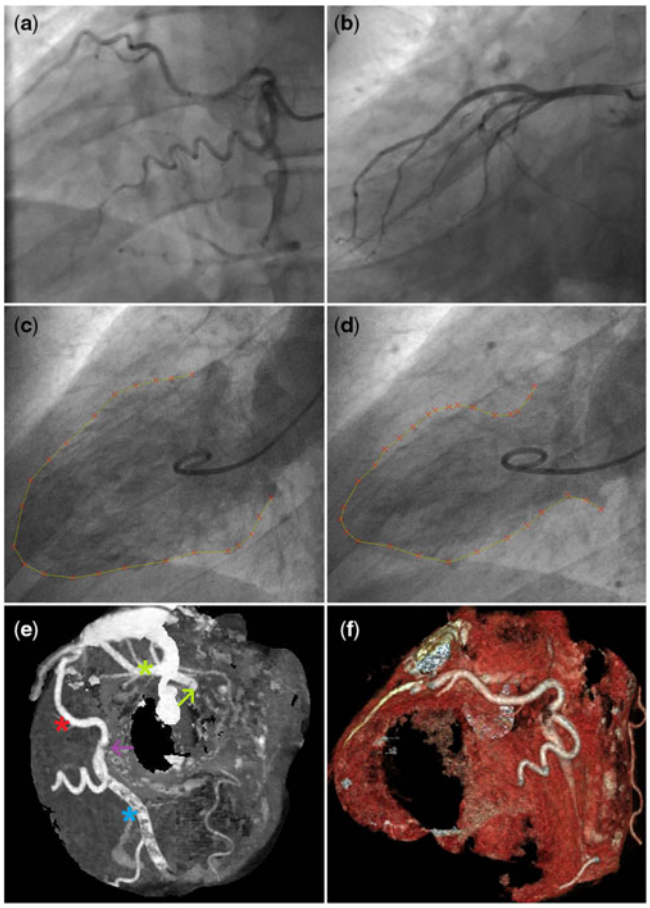

Figure 2. Coronary angiography showing the anomalous left anterior descending artery arising from right coronary artery (Panel ***a***; PA) and the left circumflex artery (***b***, LAO 6:CAU 42). Left ventriculogram in LAO position suggestive of stress cardiomyopathy (***c*** and ***d***). CT coronary angiography reconstructions in panels (***e*** and ***f***). Panel (***e***) demonstrates the left anterior descending artery (red asterix) and right coronary artery (blue asterix) arising from a single ostium (purple arrow) in the right coronary cusp. The left circumflex artery (green asterix) and ostium (green arrow) arise from the left coronary cusp. CAU = caudal; LAO = left anterior oblique.

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Conflicts of interest. None.

Ethical standards. The patient provided informed consent for publication.

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