Orthotopic Heart Transplantation for Ankylosing Spondylitis Masquerading as Nonischemic Cardiomyopathy

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Described herein is a 48-year-old man who underwent orthotopic heart transplantation because of severe heart failure considered clinically due to idiopathic dilated cardiomyopathy, but examination of the operatively excised native heart disclosed classic features of ankylosing spondylitis. Orthotopic heart transplantation for this condition has not been reported previously. **(Am J Cardiol 2019;123:1732−1735)**

We recently studied the heart of a patient who had undergone orthotopic heart transplantation (OHT) for presumed idiopathic dilated cardiomyopathy and examination of the operatively excised heart disclosed it to have classic morphologic features of ankylosing spondylitis.[1,2] The patient clinically had aortic regurgitation, complete heart block, and periodic low back pain. Search of PubMed failed to disclose any report of OHT for ankylosing spondylitis. A description of this patient is the purpose of this report.

Case Description

A 48-year-old male roofer, who was born in March 1970, had been well until May 2011 (age 41) when he developed the sudden onset of dyspnea and was hospitalized. His systolic blood pressure was about 200 mm Hg, his coronary arteries were free of obstructive lesions, and his left ventricular ejection fraction was about 15%. He was started on valsartan, carvedilol, isosorbide dinitrate, and amlodipine, but despite these medicines, he had frequent episodes of acute heart failure. During one episode in March 2015, he was found to have abnormal kidney function and an atrophic left kidney (cause unknown) that was excised. The main artery to the right kidney was found to be stenotic and a stent was inserted. At that time, he developed complete heart block and a dual chamber pacemaker was inserted. In July 2016, he was started on peritoneal dialysis and 2 months later, hemodialysis. In July 2017, cardiac resynchronization therapy defibrillator was inserted.

In April 2018, he developed cardiogenic shock and pulmonary edema and was transferred to Baylor University Medical Center at Dallas. On arrival, his blood pressure was 160/80 mm Hg. A precordial murmur was not heard but his respirations were extremely rapid. The electrocardiogram (Figure 1) showed atrial-sensed ventricular-paced rhythm and total 12-lead QRS voltage of 152 mm (10-mm standard).[3] The echocardiogram (Figure 2) showed the left ventricular chamber to be severely dilated, the ejection fraction to be about 20%, and severe aortic regurgitation to be present. At cardiac catheterization, the cardiac index was 1.5 L/min/m^2. Certain laboratory findings are listed in Table 1.

He underwent combined heart and kidney transplant in May 2018. The native heart weighed 675 g (Figures 3 and 4). The left ventricular cavity was considerably dilated longitudinally: the distance from the base of the right aortic valve cusp to the apex was 9.5 cm. The anterior mitral leaflet was severely thickened by dense fibrous tissue, and the posterior mitral leaflet was normal. The bases of each aortic cusp were thickened by similar fibrous tissue which extended cephalad onto the aorta in the areas of the commissures. The epicardial coronary arteries were free of atherosclerotic plaques.

Discussion

Described herein is a 48-year-old man who underwent OHT because of severe heart failure attributed clinically to idiopathic dilated cardiomyopathy. Study of his explanted native heart, however, disclosed classic (specific) morphologic findings of ankylosing spondylitis,[1,2] distinctive and different from other cardiac conditions (Figure 5). Before OHT, echocardiogram disclosed severe aortic regurgitation. Although the degree of aortic regurgitation in our patient was severe by echocardiogram, a precordial murmur was not detected while in severe heart failure, probably the result of his rapid respiratory rate and his obesity (body mass index 33 kg/m^2). A precordial murmur had been present earlier when he was not in heart failure. His pulse pressure when hospitalized at our institution was 80 mm Hg.

Aortic regurgitation appears to occur in about 20% of patients with ankylosing spondylitis[4] and it usually appears after the appearance of the orthopedic consequences, although the reverse occurs, as in the present patient, on occasion. The severe thickening of the anterior mitral leaflet in ankylosing spondylitis in the

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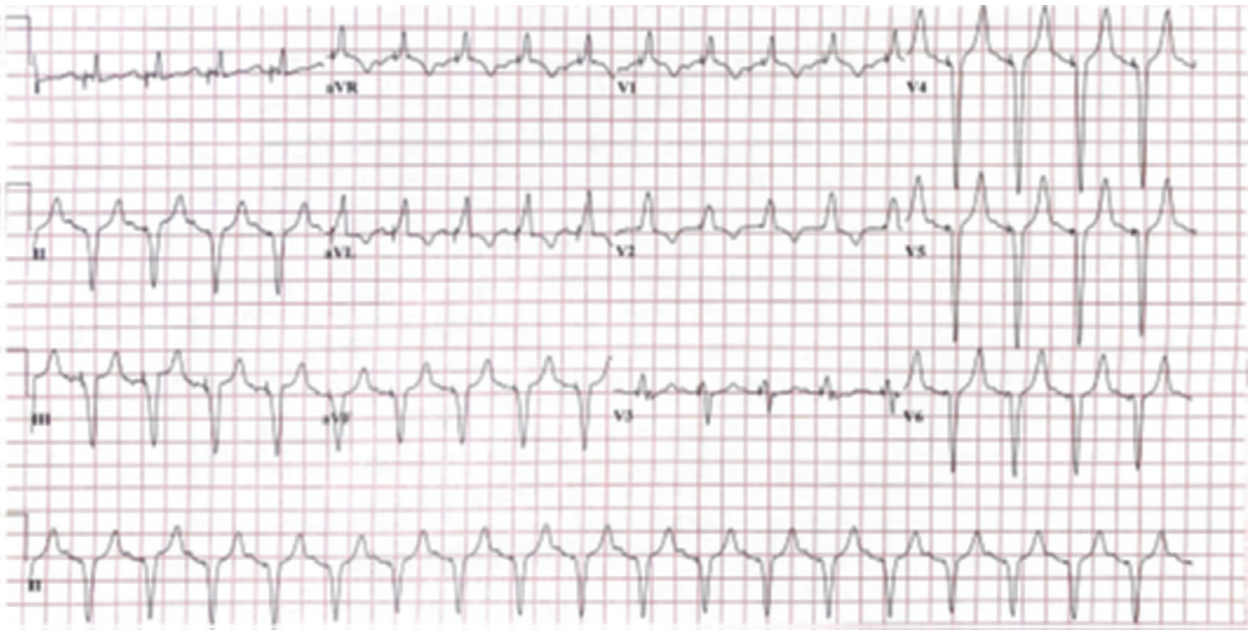

Figure 1. Electrocardiogram, recorded at the time of presentation, showing atrial-sensed ventricular-paced rhythm, biventricular pacemaker, and the total 12-lead QRS voltage of 152 mm.

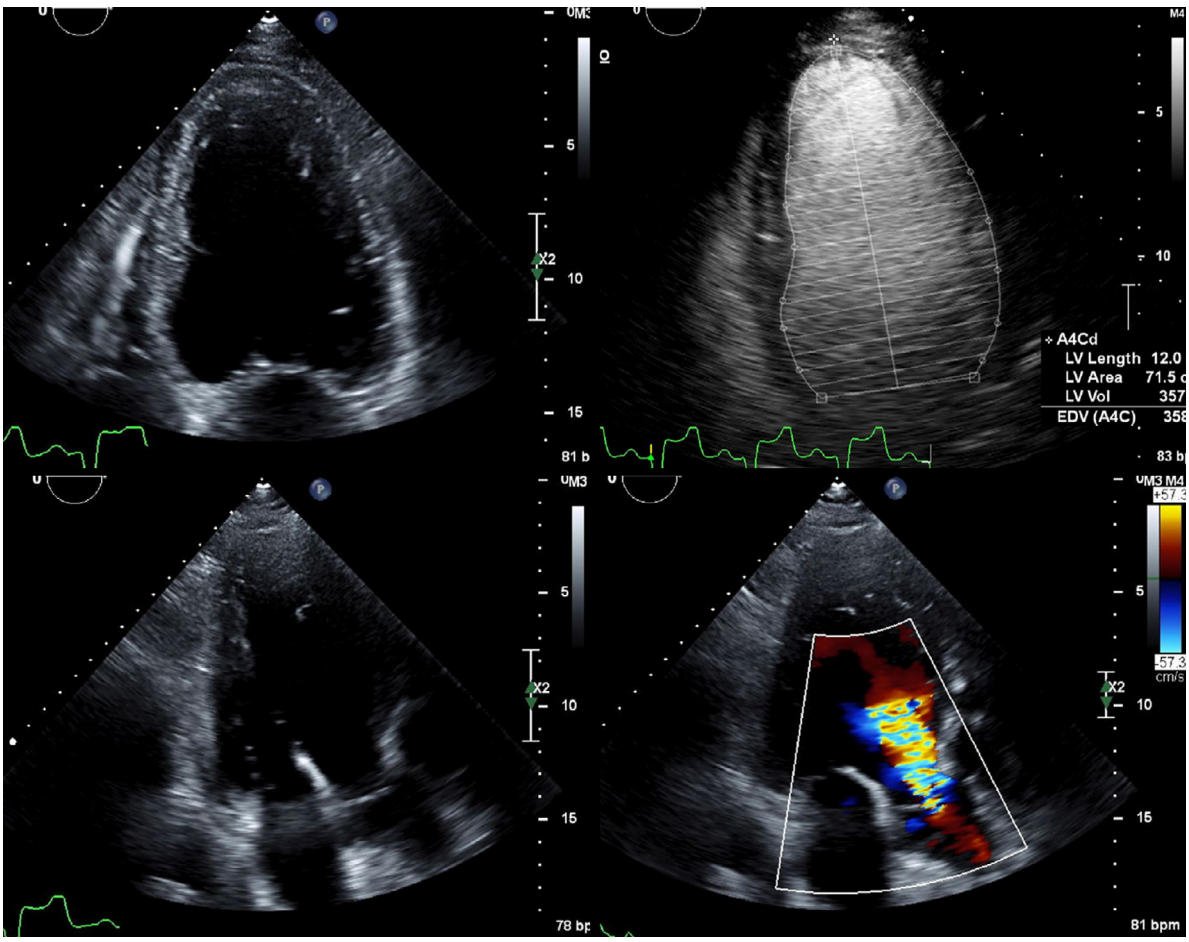

Figure 2. *Top left*: Apical 4-chamber view showing severely dilated, elongated left ventricle (LV) with normal right ventricular (RV) size and systolic function. An ICD lead is seen in the RV (arrow). *Top right*: Apical 4-chamber end-diastolic frame with ultrasound contrast. LV end-diastolic volume was 358 ml with LVEF 19% by biplane Simpson's method. *Bottom left*: Apical long-axis view showing severely thickened, restricted anterior mitral leaflet (yellow arrow). The posterior leaflet (white arrow) was of normal thickness and motion. *Bottom right*: Apical long-axis view with color Doppler imaging showing severe aortic regurgitation (AR).

Table 1
Pertinent admission laboratory findings in the patient described

B-type natriuretic peptide (pg/ml)	1895
Creatinine (mg/dl)	18
Blood urea nitrogen (mg/dl)	77
Estimated GFR (ml/min/1.73 m^2)	3
Sodium (meq/L)	136
Potassium (meq/L)	5.2
Calcium (mg/dl)	8.7
Magnesium (mg/dl)	2.2
Phosphorous (mg/dl)	2.0
Total cholesterol (mg/dl)	219
Low density lipoprotein cholesterol (mg/dl)	151
High density lipoprotein cholesterol (mg/dl)	35
Triglyceride (mg/dl)	241
Hemoglobin A1c (%)	5.8
Rheumatic factor ([IU]/ml)*	8
ANA*	Negative
HLA-B 27*	Negative
C-reactive protein (mg/dl)*	0.5

GFR = glomerular filtration rate.
* Test performed 6 months after the orthotopic heart transplant.

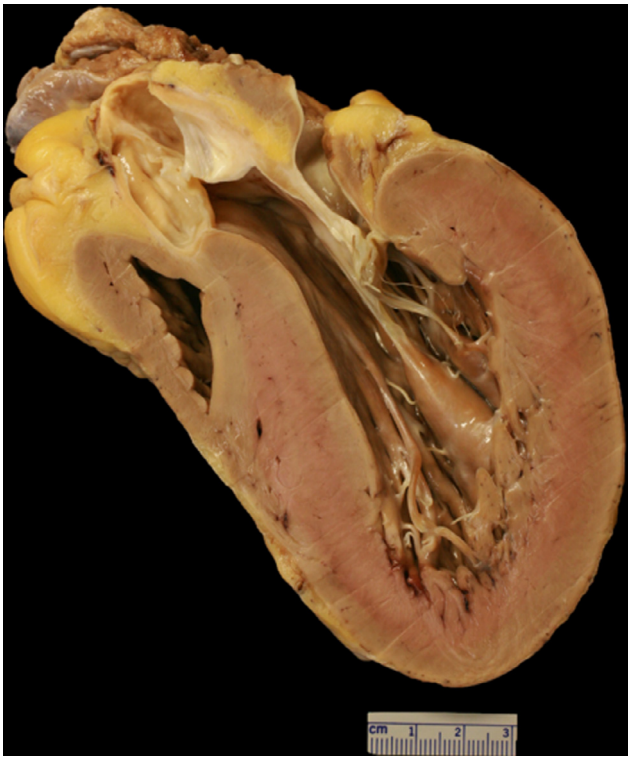

Figure 3. Shown here is the heart of a 48-year-old man showing a dilated left ventricular cavity with thickened left ventricular walls, enlarged papillary muscles, and thickened anterior mitral leaflet. The posterior mitral leaflet is normal (not thickened).

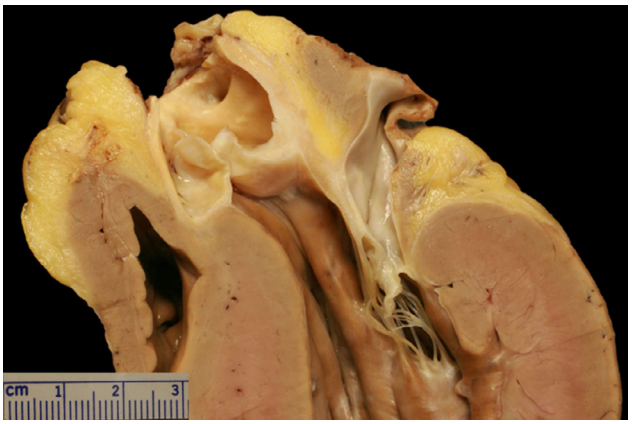

Figure 4. Shown here is a closer view of the mitral and aortic valve showing the remarkably thickened anterior mitral leaflet which is extending into the base of the posterior aortic valve cusp. The posterior mitral leaflet is normal.

absence of thickening of the posterior mitral leaflet as shown in the present patient is diagnostic (Figure 5). Although all 8 patients (all men) with ankylosing spondylitis studied by Buckley and Roberts[1] at necropsy had extremely severe aortic regurgitation, only one of the 187 patients with ankylosing spondylitis studied clinically by Klingberg et al[4] had "severe" aortic regurgitation; 24 others had "mild", and 9 had "moderate" aortic regurgitation.

Interview of the patient and his wife 3 months after the OHT revealed that the patient indeed had had low back pain periodically for years, but he attributed it to his kidney disease rather than to the arthritic problem. Thus, the cardiac features of ankylosing spondylitis in this patient probably appeared after the clinical onset of his orthopedic back problem. Lateral chest radiograph, however, did not show changes of ankylosing spondylitis.

The dense fibrous tissue − characteristic of ankylosing spondylitis − was present in the membranous ventricular septum just above the location of the atrioventricular node and its presence in that location appears to be the cause of the patient's complete heart block diagnosed initially about 2 years before the OHT.

We were unable to find a previous publication of a patient with ankylosing spondylitis having an OHT.

Disclosures

The authors have no conflicts of interest to disclose.

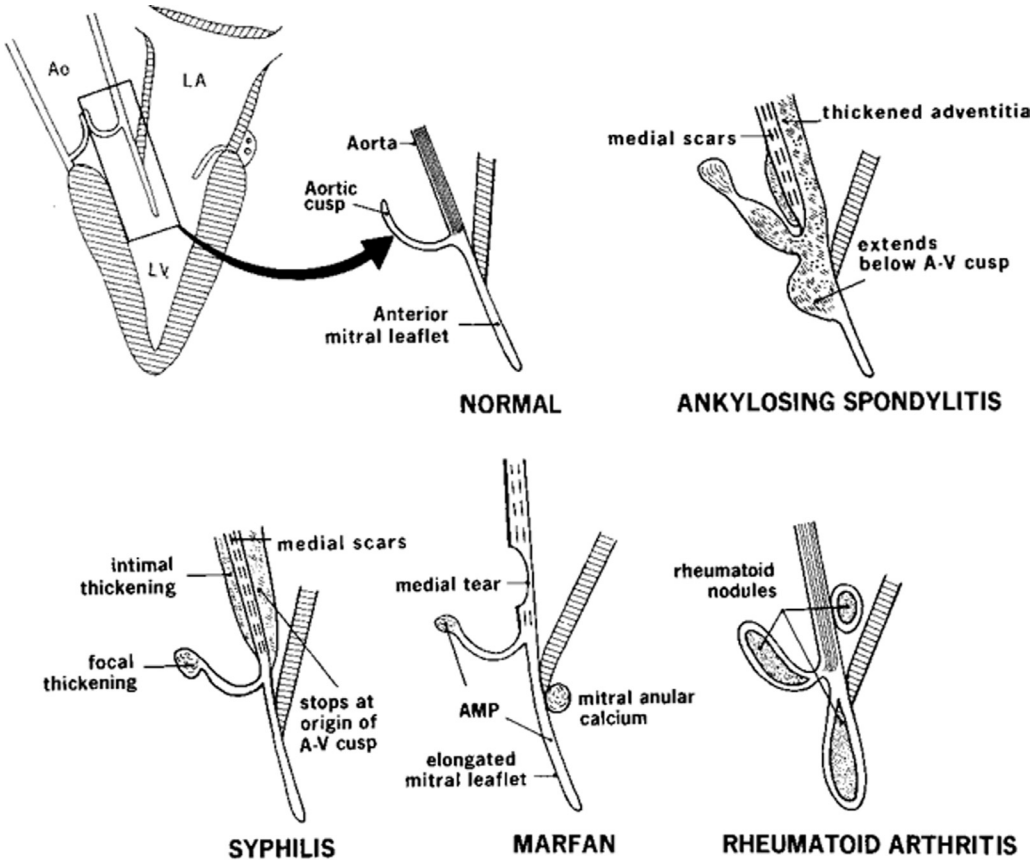

Figure 5. Diagram showing the distinctive morphologic features of 4 different cardiac conditions including *ankylosing spondylitis*. In *cardiovascular syphilis*, the aortic wall behind the sinuses of Valsalva is spared and the adventitial scar tissue does not extend below the aortic valve or involve mitral valve or ventricular septum. Only the distal margins of the aortic valve cusps are thickened in syphilis, not the proximal portions which are always involved in ankylosing spondylitis. In *rheumatoid arthritis*, the distinctive nodules similar to subcutaneous nodules, may infiltrate pericardium, myocardium and mural and valvular endocardium. If the valvular tissue is involved, regurgitation usually of only mild degree results. In the *Marfan syndrome*, aortic regurgitation is a consequence of disease of aortic wall, not of aortic valve; the aorta is thinner, and usually contains intimal-medial tears. The ascending aorta is diffusely involved, and dilatation of the aortic root causes the aortic regurgitation, which is usually severe. The mitral and rarely the aortic valve cusps may be redundant in patients with the Marfan syndrome.

Abbreviations: Ao = aorta; A-V = atrioventricular; LA = left atrium; LV = left ventricle.

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