Giant Eustachian valve and left ventricular systolic dysfunction in a patient with non-dilated amyloid cardiomyopathy

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Received 7 April 2007; accepted after revision 28 April 2007; online publish-ahead-of-print 30 June 2007

Amyloid cardiomyopathy is characterized by non-dilated thick-walled left ventricular, thickening of interventricular septum and right ventricular free wall, biatrial enlargement associated with granular ‘sparkling’ appearance of the myocardium. Typically, decreased left ventricular compliance results in abnormal diastolic functions but left ventricular systolic functions are preserved until late in the course of the illness when left ventricle starts to dilate culminating into dilated cardiomyopathy. We present a 77-year-old patient who had typical echocardiographic features of amyloid heart disease, a giant Eustachian valve resembling cor triatriatum dexter and left ventricular systolic dysfunction without associated left ventricular dilatation.

Introduction

We present a patient with typical echocardiographic features of amyloid cardiomyopathy who had left ventricular systolic dysfunction in the absence of ventricular dilatation. This was also associated with a giant Eustachian valve giving the appearance of divided right atrial cavity.

Case report

A 77-year-old female with history of hypertension and chronic atrial fibrillation was brought to the hospital because of swelling and pain in bilateral lower extremities for five days. This was associated with progressively increasing shortness of breath for the last few months. She denied any chest pain, fevers, chills or cough. There was no known history of coronary artery disease or prior myocardial infarction. On examination, she had pulse rate of 88/min, blood pressure of 112/74 mmHg, respiratory rate of 22/min and oxygen saturation of 90% on room air. Jugular venous pressure was elevated around 5 cm above the sternal angle. Cardiovascular examination revealed normal heart sounds with a pansystolic murmur at the apex radiating to the axilla. She also had a holosystolic murmur at left sternal border which increased on inspiration. On auscultation of the chest, she had crackles and decreased air entry at both lung bases. Rest of the systemic examination showed hepatomegaly and bilateral erythematous, tender and swollen lower extremities suggestive of chronic edema with superimposed cellulites.

Laboratory investigations showed mild anemia with elevated white blood cell count. Chest X-ray was remarkable for bilateral vascular congestion and pleural effusions without significant cardiomegaly. EKG showed low voltage complexes in the precordial leads. She was diagnosed with congestive heart failure with cellultes of lower extremities and was started on diuretic therapy and appropriate antibiotics. An echocardiogram was ordered to further evaluate heart failure.

Transthoracic echocardiogram revealed non-dilated thick-walled left ventricle, interventricular septum and right ventricle free wall, biatrial enlargement and thickened mitral and tricuspid valve leaflets associated with moderate mitral and tricuspid regurgitation and trace pericardial effusion (Figures 1 and 2). Ventricular walls had reflectile ‘sparkling’ appearance typical of amyloid heart disease (Figure 1). Doppler examination of mitral-inflow velocity showed prominent E waves with steep deceleration slope and a small E/a wave on tissue Doppler, a pattern suggestive of

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restrictive heart disease. There was a giant Eustachian valve in the right atrial cavity extending from mid-interatrial septum to just above the inferior vena cava simulating a cor triatriatum dexter (Figure 1). This was also associated with prominent chiari network and a small patent foramen ovale with left to right shunting (Video clip 1). Echocardiography also revealed akinetic anterior-lateral, anterior, anterior-septal and inferior-septal walls with severe left ventricular systolic dysfunction and estimated left ventricular ejection fraction of 30% (Video clips 2 and 3).

Patient underwent MRI of her lower extremities to evaluate possible necrotizing fascitis. Unfortunately she developed ventricular fibrillation and cardiac arrest while undergoing MRI and died in spite of aggressive resuscitative measures. Family did not request autopsy.

Discussion

We present a patient of restrictive cardiomyopathy who had severe left ventricular systolic dysfunction in absence of left ventricular dilatation. Though myocardial biopsy was not done, echocardiographic features were quite suggestive of amyloid infiltration. These include thickened left ventricular and right ventricular free walls with 'sparkling' appearance of the myocardium, biatrial enlargement, thickening of valvular leaflets, mitral and tricuspid regurgitation and restrictive pattern on Doppler inflow velocities. Also, our patient had few unusual features. Patients with amyloid cardiomyopathy usually have non-dilated left ventricles with preserved systolic functions until late in the course of their disease. While these patients may progress and develop dilated cardiomyopathy, our patient had abnormal left ventricular systolic functions without any anatomic/hemodynamic pattern of left ventricular dilatation.

Patho-physiological mechanisms responsible for left ventricular systolic dysfunction in our patient are not clear. Though it may be attributed to concomitant coronary artery disease, myocardial ischemia with or without prior myocardial infarction there was no clinical or electrocardiographic evidence of significant coronary artery disease. Moreover, absence of myocardial thinning makes prior myocardial infarction a less likely etiology for left ventricular dysfunction. It is possible that extensive infiltration of amyloid protein by itself may result in impaired myocardial contractility in the presence of normal myocardial perfusion.

Our patient also had a giant Eustachian valve extending from inferior vena cava to mid portion of the interatrial septum giving the appearance of a divided right atrium. This type of Eustachian valve has been described only...
rarely before.\textsuperscript{2} Such a giant valve should be differentiated from cor triatriatum dexter, another rare cardiac malformation which can resemble the former echocardiographically. This is especially important due to the fact that the latter entity may require surgical correction. Cor-triatriatum dexter forms a large obstructive (though perforated) membrane or septum across the right atrium thus dividing it into two distinct chambers. The upper chamber receives blood from both inferior and superior vena cava and directs it towards interatrial septum with shunting of blood from right to left atrium through atrial septal defect, which frequently co-exists in these patients. This results in mixing of desaturated blood with oxygenated blood in the left atrium causing cyanosis. Our patient had none of these features and had a free flow of blood across the Eustachian valve. Though there was a communication between the two atria in our patient, color Doppler demonstrated predominantly left to right shunting across the interatrial septum.

**Supplementary material**

Supplementary date associated with this article can be found in the online version.

**References**