Left Ventricular Torsion by Two-Dimensional Speckle Tracking Echocardiography in Patient with A-Type Amyloid Heart Disease

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Amyloidosis is a clinical disorder caused by extracellular and or intracellular deposition of insoluble abnormal amyloid fibrils that alter the normal function of tissues. Amyloid A amyloidosis is the most common form of systemic amyloidosis worldwide The heart may be affected in systemic AL amyloidosis, but also more rarely in A amyloidosis.

In this report, we presented a patient with A-type amyloid heart disease who had dynamic left ventricular outflow tract obstruction mimicking hypertrophic obstructive cardiomyopathy. We also investigated left ventricular torsion, untwisting rate and left ventricular longitudinal global strain by two-dimensional speckle tracking echocardiography. (J Am Soc Echocardiogr 2011;24:818.e5-818.e9.)

Keywords: Cardiac amyloidosis, Left ventricular outflow tract obstruction, Two-dimensional speckle-tracking echocardiography, Torsion

Amyloidosis is a clinical disorder caused by extracellular and/or intracellular deposition of insoluble abnormal amyloid fibrils that alter the normal function of tissues.[1] Amyloid A (AA) amyloidosis is the most common form of systemic amyloidosis worldwide.[2] In A amyloidosis, the kidneys, liver, and spleen are the major sites of involvement. The heart may be affected in systemic amyloid light chain amyloidosis, but also more rarely in AA amyloidosis.[1,3]

In this report, we present the case of a patient with A-type amyloid heart disease who had dynamic left ventricular (LV) outflow tract (LVOT) obstruction mimicking hypertrophic obstructive cardiomyopathy (HOCM). We also investigated LV torsion, untwisting rate, and LV longitudinal global strain using two-dimensional speckle-tracking echocardiography.

CASE PRESENTATION

A 33-year-old male patient with no known previous disorder was admitted to our hospital with the complaints of dyspnea, anorexia, nausea, and vomiting. On physical examination, his blood pressure was 110/70 mm Hg, and his pulse rate was rhythmic and 76 beats/min. He had a grade 1/6 systolic murmur along the left sternal border that increased with performance of the Valsalva maneuver and hepatosplenomegaly 2 cm. electrocardiography revealed normal sinus rhythm and T negativity in leads V_4 to V_6 derivations and down-sloping ST-segment depression. Hematologic results were normal. Biochemical investigation showed serum creatinine of 10.1 mg/dL, blood urea nitrogen of 70 mg/dL, sodium of 128 mEq/L, and potassium of 6.5 mEq/L, and blood gas analysis revealed metabolic acidosis. Urinalysis showed albuminuria (800 mg/L in 24 hours). To assess the clonality of immunoglobulins, serum protein electrophoresis and serum and urine immunoelectrophoresis were performed. We did not detect any amonoclonal antibodies. Renal ultrasonography showed bilaterally increased dimensions and echogenicity in the kidneys. On follow-up, the patient's urine output decreased. He was scheduled for hemodialysis because of uremic symptoms, refractory acidosis, and hyperkalemia.

The patient improved after hemodialysis, and transthoracic echocardiography was performed. Echocardiography revealed that LV function was normal (ejection fraction, 60%), and there was minimal pericardial effusion and concentric LV hypertrophy (septal thickness, 2.6 cm; posterior wall thickness, 2.3 cm; LV mass index, 208 g/m^2); also, granular sparkling was widely observed, particularly on the interventricular septum (Figure 1, Video 1). Although there was no systolic anterior motion or premature closure of the aortic valve, we investigated the LVOT gradient because of the presence of septal thickness. There was a gradient across the LVOT of 20 mm Hg at rest and 75 mm Hg after performance of the Valsalva maneuver. Transmitral flow was found to be consistent with a restrictive pattern (ratio of transmitral early diastolic to late diastolic velocity, 2.7; deceleration time, 97 msec; E/E′ ratio, 21; flow propagation velocity, 23 cm/sec; Figure 2). In addition, LV torsion was 8°, peak basal rotation was −2.4°, peak untwisting velocity was 25°/sec, and LV longitudinal global strain was −7 on speckle-tracking echocardiography (Figures 3A and 3B and Videos 2 and 3). To evaluate right ventricular systolic function globally, tricuspid annular motion was assessed and found to be 2.3 cm (Figure 4).

Amyloidosis was considered because of the presence of LV concentric hypertrophy and granular sparkling images and the determination of restrictive flow pattern. Amyloid collections were found in aspiration of abdominal fat tissue, which were stained by Congo red and crystal violet stains. Renal biopsy was done, and incubation of kidney tissue with potassium permanganate prevented amyloid AA from staining with Congo red. The patient was diagnosed with

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Abbreviations
AA = Amyloid A
HOCM = Hypertrophic obstructive cardiomyopathy
LV = Left ventricular
LVOT = Left ventricular outflow tract

A-type (formerly AA-type, secondary) amyloidosis. The patient was interrogated for familial Mediterranean fever. He was diagnosed as having familial Mediterranean fever because of recurrent episodes of fever, arthritis, and peritonitis.

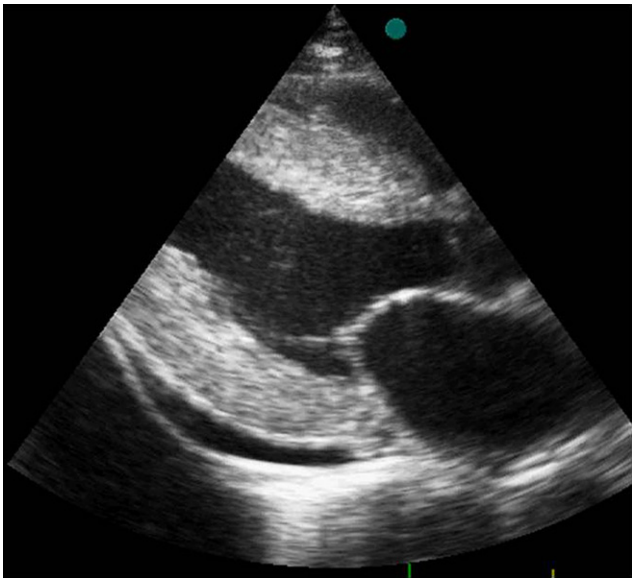

Figure 1 Parasternal long-axis view showing marked septal hypertrophy and also hypertrophy of the LV posterior wall.

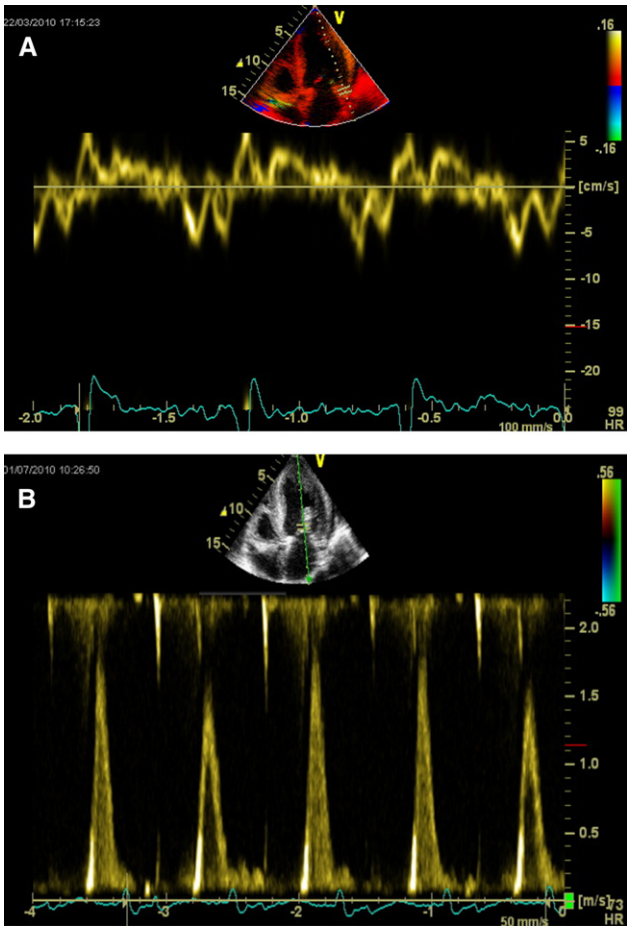

Figure 2 Pulsed Doppler and color Doppler tissue imaging showing a restrictive pattern.

DISCUSSION

AA amyloidosis is the most common form of systemic amyloidosis worldwide. The precursor protein in A amyloidosis is a normal-sequence apolipoprotein serum amyloid A, now called "A," which is an acute-phase reactant produced mainly in the liver in response to cytokines.[2] In A amyloidosis, the kidney, liver and spleen are the major sites of involvement. Heart involvement is seen most often in primary amyloidosis. The heart is affected very rarely in A amyloidosis.[1,3]

Amyloid infiltration of the heart may frequently mimic as other cardiac disorders. The classical cardiac manifestation in amyloidosis is thought to be a restrictive cardiomyopathy.[3] Although dynamic LVOT obstruction is classically seen in HOCM, it can also be seen rarely in cardiac amyloidosis.[4] Mookadam *et al.*[5] reported a case of senile amyloidosis with LVOT obstruction. LVOT obstruction has also been described in amyloidosis with multiple myeloma and type I Gaucher's disease.[5] To our knowledge, a case report of A-type cardiac amyloidosis due to familial Mediterranean fever causing dynamic LVOT obstruction has not appeared in the literature before. In the present case, there was only a small LVOT gradient (20 mmHg) at rest, but the LVOT gradient increased to 75 mmHg after Valsalva maneuver.

We could not perform endomyocardial biopsy. It is suggested that typical noninvasive findings, together with the demonstration of amyloid in another organ, are sufficient for a diagnosis.[1,5]

Peak untwisting velocity of 25°/sec was detected by speckle-tracking echocardiography. One study has shown that LV untwisting is delayed in HOCM; this finding probably contributes to diastolic dysfunction.[6] Wang *et al.*[7] also reported that dynamic obstruction leads to delayed untwisting in HOCM.

One study has found that transmitral flow indices consistent with restrictive patterns are important predictors of survival in cardiac amyloidosis.[8] In the presented case, transmitral Doppler flow studies detected a restrictive LV filling pattern. We also investigated LV torsion, untwisting rate, and LV longitudinal global strain using speckle-tracking echocardiography. The parameters we found were low. LV torsion and subsequent untwisting play an important role in diastolic filling. Park *et al.*[9] reported that LV torsion and untwisting were markedly reduced in patients with cardiac amyloidosis and severe diastolic dysfunction. Decreased LV torsion is markedly compromised with severe elevation of filling pressures. They also found significant reductions of LV torsion and untwisting in patients with cardiac amyloidosis and severe diastolic dysfunction compared with patients with HOCM and severe diastolic dysfunction. Therefore, we suggest that speckle-tracking parameters can be used for distinguishing cardiac amyloidosis with dynamic LVOT obstruction from HOCM.

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