Ultrasound for Critical Care Physicians: Now My Heart Is Full

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A 65 year old woman with a history of hypertension and a recent diagnosis of multiple myeloma was admitted to the ICU with septic shock due to *Morganella morganii* bacteremia. She was treated with cefepime, levophed and dobutamine. During treatment she developed symptoms and a chest x-ray compatible with congestive heart failure. A transthoracic echo is shown below (Figures 1 and 2).

Figure 1. Parasternal long echocardiogram of the patient.

Figure 2. Apical four-chamber echocardiogram of the patient.

Additionally a spectral pulsed-wave Doppler study of the mitral inflow velocities is presented (Figure 3).

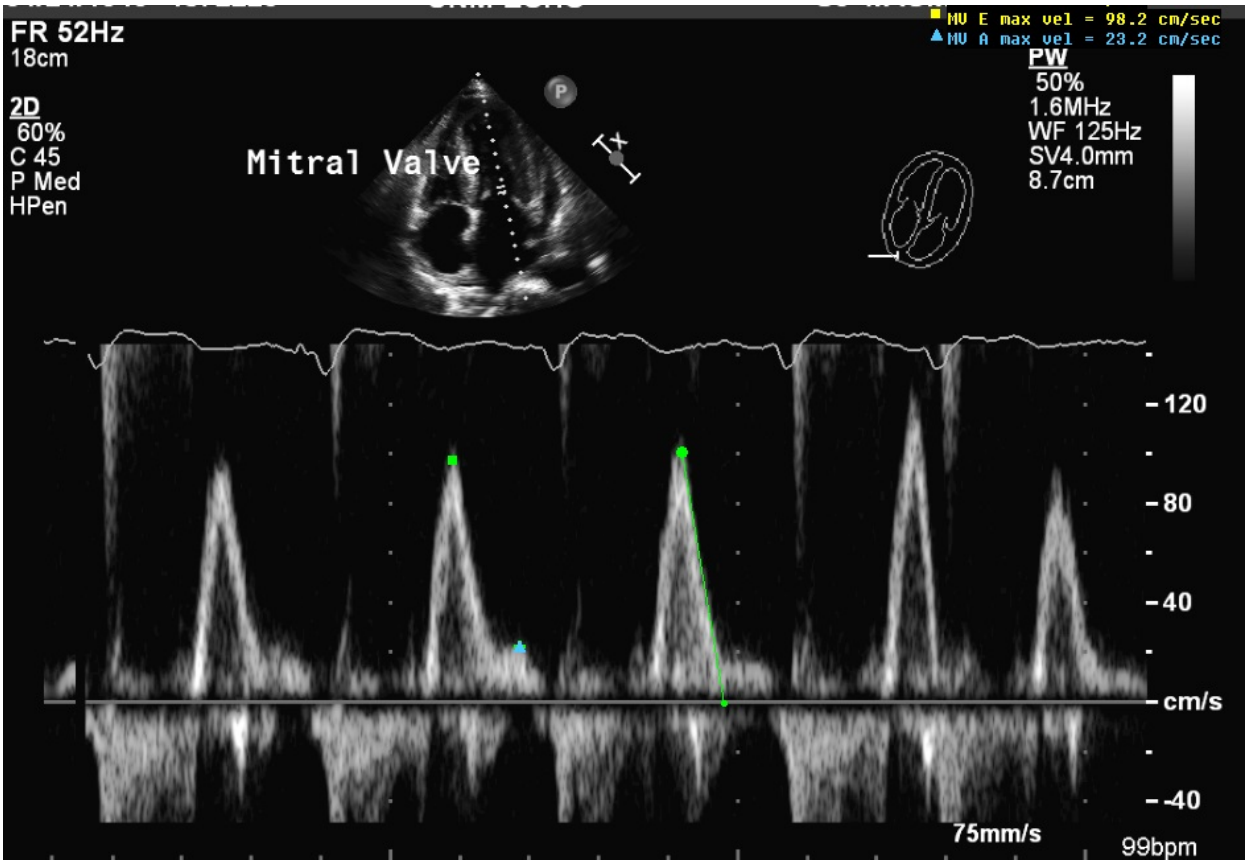

Figure 3. Pulsed-wave spectral Doppler velocities of the mitral-valve inflow of the patient.

What is the ***best*** explanation for the findings seen in on the echocardiogram?

1. Cardiac amyloidosis
2. Hypertensive heart disease
3. Hypertrophic cardiomyopathy
4. Stress (Takotsubo) cardiomyopathy

Correct!
1. Cardiac amyloidosis

The transthoracic echocardiogram demonstrates a mildly reduced ejection fraction (~40-45%) but marked global thickening of the myocardium. This is best appreciated in the parasternal long view (Figure 4).

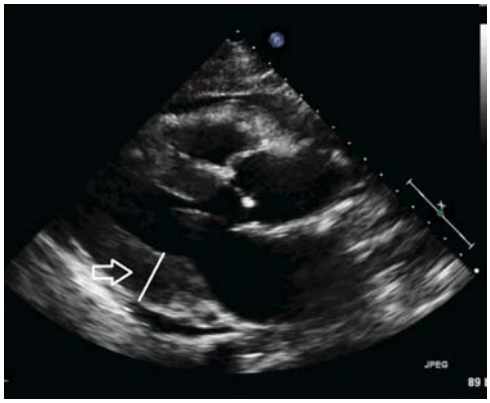

Figure 4. Still image of the patient's parasternal long echocardiogram in diastole, demonstrating thickness of the myocardium (white line). Using the scale on the right it is estimated at least 2 cm.

A small, incidental, pericardial effusion is also noted in this view. The Doppler study reveals a high ratio of early to late filling in the mitral inflows (high e/a ratio) suggestive of a restrictive filling pattern (Figure 5).

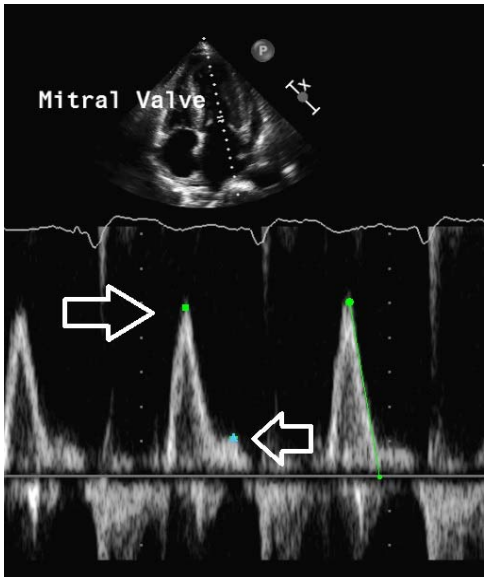

Figure 5. Pulsed wave Doppler of the patient's mitral valve inflow with the right pointing arrow demonstrating the early velocity (E wave) and the left pointing arrow the (A-wave). The ratio of E/A greater than 2:1 suggests a restrictive filling pattern.

Hypertensive cardiomyopathy could also demonstrate increased thickness of the myocardium but not usually a restrictive filling pattern. Hypertrophic cardiomyopathy has multiple forms but is not usually diffuse, or associated with a restrictive pattern. Typically it would usually demonstrate an increase in septal thickness. No mention of obstruction is given. Stress cardiomyopathy typically demonstrates regional differences in contractility usually with apical ballooning. Cardiac amyloidosis is the right answer because of the combination of increased thickness of the myocardium and a restrictive filling pattern.

The patient underwent an endomyocardial biopsy that stained positive with Congo Red consistent with cardiac amyloidosis. Given the patient's depressed left ventricular ejection fraction, bortezomib chemotherapy could not be given to treat her multiple myeloma due its known adverse effect of causing further decreases in left ventricular ejection fraction in patients with existing heart disease (1,2). The patient wished to stop all aggressive treatments and was discharged home with hospice.

In our patient, restrictive cardiomyopathy was due to amyloid deposition in the myocardium from underlying multiple myeloma. Amyloid cardiomyopathy is a rare disease that can be seen with systemic primary or secondary amyloidosis. Echocardiographic features associated with amyloid cardiomyopathy include diastolic dysfunction, left ventricular wall thickening, small ventricular chambers, large atria, thickened ventricular septum, pericardial effusion and "sparkling" appearance of the myocardium due to amyloid and collagen nodules in the heart (3). The speckling or starry sky appearance is no longer specific to amyloid, and is seen in most echocardiograms due to the use of tissue harmonic imaging in modern echocardiography (4). This harmonic imaging mode can however, be turned off on most machines to bring out the classic starry sky appearance.

References

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