

Non-compliant left atrium masquerading as severe mitral regurgitation on cardiac catheterisation haemodynamics

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CASE

A 68-year-old woman was referred to our cardiac clinic with a 4-month history of progressive dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, pedal oedema and fast regular palpitations. Further enquiry revealed diarrhoea and weight loss

ABSTRACT

A prominent v wave on a capillary wedge or left atrial pressure tracings classically signify significant mitral regurgitation. However, infiltrative disease involving the left atrium and left atrial scar from prior catheter ablation or Maze procedures, may produce a similar waveform due to noncompliance of the left atrium.

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of about 20kg. On clinical examination she was wasted, pale, had bilateral pitting pedal oedema and periorbital purpuric rash. Her jugular venous pressure was elevated, the apex beat was not displaced and there were no murmurs. The electrocardiogram showed small complexes on the limb leads, non-specific T wave inversion, and atrial flutter with a variable atrioventricular block (Figure 1). The echocardiogram showed a small pericardial effusion, left ventricular hypertrophy, bi-atrial enlargement, and no valvular regurgitation (see online publication for video supplement).

She underwent invasive haemodynamic assessment: The invasive haemodynamics revealed restrictive physiology with

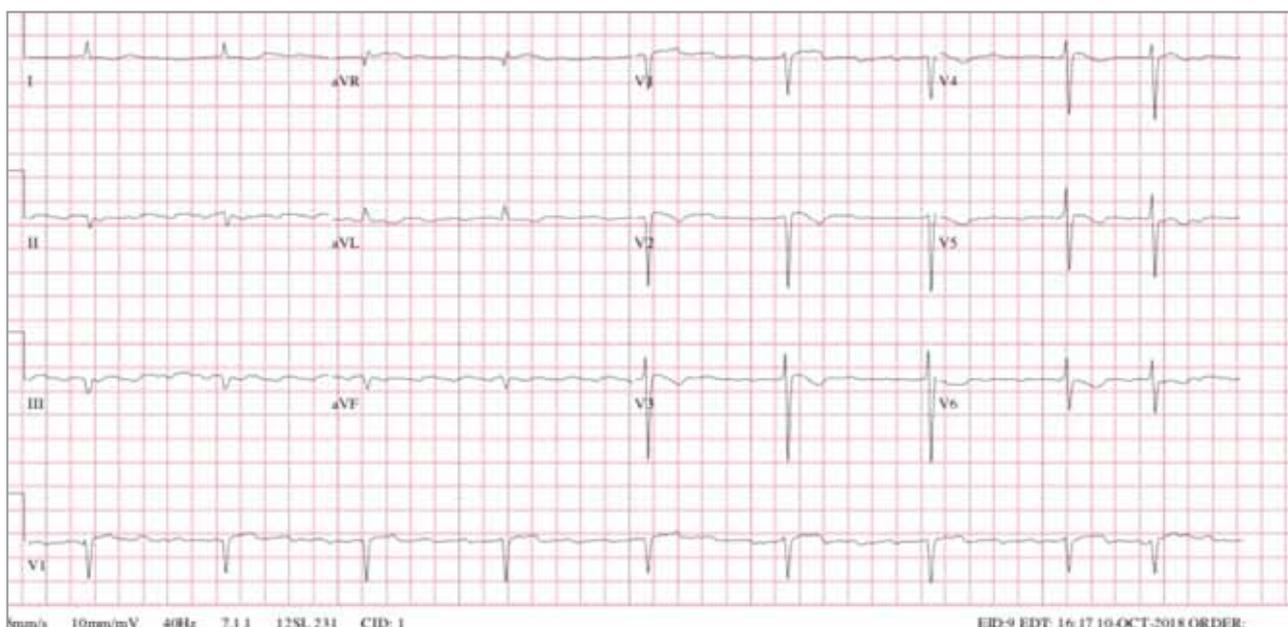


FIGURE 1: Electrocardiogram shows atrial flutter with a variable atrioventricular block, ventricular rate of 54/minute, and small complexes in the limb leads.



VIDEO 1: The echocardiogram showed a small pericardial effusion, left ventricular hypertrophy, bi-atrial enlargement, and no valvular regurgitation.

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raised right atrial pressures with prominent x and y descents, raised right ventricular diastolic pressures with a dip and plateau (square root sign). Similarly, the left ventricular diastolic pressures were elevated with a dip and plateau (Figure 3).

We call attention to the pulmonary capillary wedge (PCWP) tracing of elevated mean wedge pressure and a large V wave (Figure 2) and a simultaneous PCWP and the left ventricle end-diastolic tracing, which shows a large V wave gradient (Figure 4). These findings are classically suggestive of significant mitral valve regurgitation. However, in this scenario, a large V wave and a V wave gradient in the absence of significant mitral regurgitation is characteristic of a stiff non-compliant left atrium, suggestive of amyloid infiltration of the left atrium.

She later developed small bowel obstruction and endoscopy and biopsy of the small bowel was performed. The demonstration of perivascular and interstitial amorphous eosinophilic



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FIGURE 2: Pulmonary wedge pressure tracing with a V wave of 54mmHg, and mean pressure of 36mmHg; these are elevated pressures.



FIGURE 3: Left ventricular diastolic tracing shows diastolic pressures of 10mmHg - 29mmHg, and a dip and plateau (square root sign) characteristic of restrictive haemodynamics.

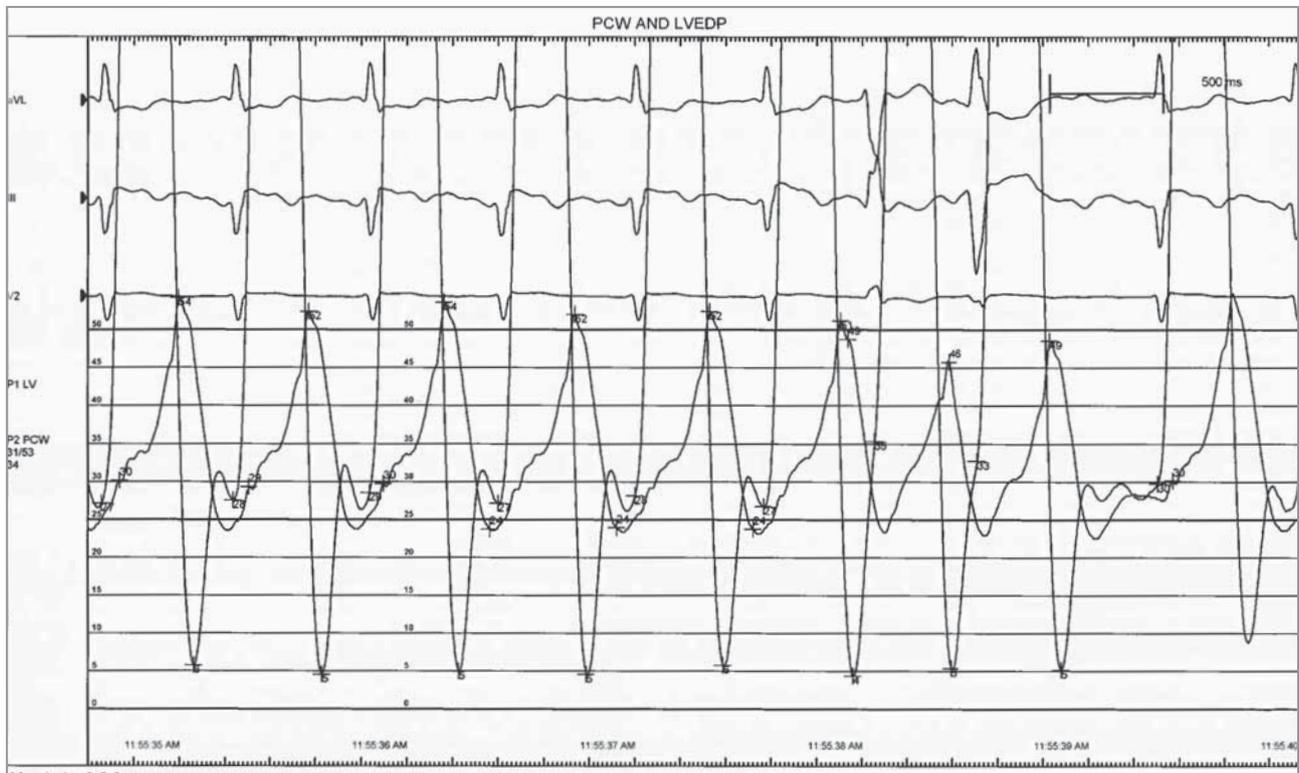


FIGURE 4: Simultaneous PCWP and left ventricular diastolic tracing shows a V wave gradient suggestive of a non-compliant left atrium.

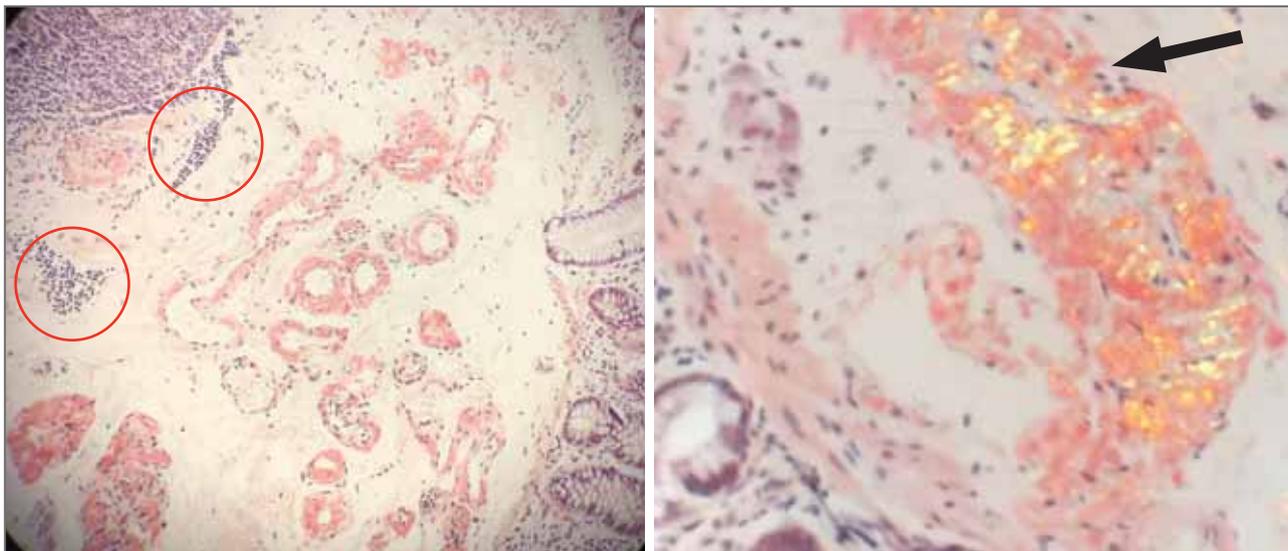


FIGURE 5: Duodenal biopsy shows perivascular and interstitial amorphous eosinophilic amyloid deposits (red circles), and apple-green birefringence highlighted on Congo red staining and polarised light microscopy (Black arrow) (**FIGURE 6**).

amyloid deposits, highlighted on Congo red staining, was consistent with cardiac amyloidosis and the bone marrow biopsy confirmed multiple myeloma (Figures 5 and 6). She died 2 months later on multiple myeloma therapy.

Conflict of interest: none declared.

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