85-year-old male presenting with worsening fatigue, dyspnea on exertion, and weight gain

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Cardiac MR- Coronal Scout
T1- Axial TSE
Short Axis Late Gadolinium Enhancement
Cardiac Amyloidosis
RV Wall Thickening Measuring up to 5mm

Intraventricular Septal Thickening Measuring up to 2.3cm

LV Wall Thickening Measuring up to 1.6cm
Diffuse, patchy enhancement consistent with infiltrative disease such as amyloidosis.
Diffuse sub-endocardial enhancement, consistent with infiltrative disease such as amyloidosis.
Cardiac Amyloidosis

- Cardiac amyloidosis is protein deposition and aggregation of amyloid within the myocardium.
  - Involvement can be sub-endocardial or transmural depending on the type of amyloid disease.

- There are two types of amyloid disease which make up 95% of cardiac amyloidosis:
  - Light Chain Amyloidosis (AL)
  - Transthyretin Amyloidosis (ATTR)
Cardiac Amyloidosis

• Amyloid cardiomyopathy typically manifests as symptoms of heart failure, which include:
  – Dyspnea, fatigue, edema, angina, pre-syncope.

• Severe cardiac amyloidosis does lead to HFpEF because the thickened ventricular myocardium results in impaired diastolic function/filling.

• Additional cardiac manifestations include small vessel disease, conduction system disease, and pericardial disease.
Cardiac Amyloidosis

- Echocardiogram shows LV wall thickness and EKG shows low voltage amplitudes.

- Cardiac MRI is also useful in diagnosis.
  - Typical findings include late gadolinium enhancement with diffuse, patchy involvement.
  - Cardiac MRI is highly sensitive (93%) and specific (70%).

- Endo-myocardial biopsy is nearly 100% sensitive, and is used for definitive diagnosis.
Cardiac Amyloidosis

• Management of the disease depends on the type of amyloidosis the patient has.
  – AL can be treated with alkylating agents, proteasome inhibitors, immuno-modulators, and anti-CD 38 monoclonal antibody.
  – ATTR can be treated with TTR blockers, silencers, and fibril disrupters.

• Regardless of the type of amyloid disease, patients with HFpEF are treated symptomatically.
  – Treatment includes salt restriction and diuresis.
References


