68-year-old male with a 1-month history of dyspnea on exertion

Brandon Byrd, MS3



Echocardiogram













PET Scan





SPECT



















Cardiac Amyloidosis



Echocardiogram



Parasternal short-axis view

Parasternal long-axis view

Concentric left ventricular wall thickening with increased echogenicity of the myocardium

Reduced systolic and diastolic function





Widened interventricular septum

Left ventricular hypertrophy





Late gadolinium enhancement



PET Scan



Mild increased radiotracer cardiac uptake



SPECT



Diffuse myocardial tracer uptake

HEALTH

RADIOLOGY

Cardiac Amyloidosis

Amyloidosis: A heterogeneous group of diseases characterized by extracellular accumulation of abnormal fibrillar protein deposits (amyloid)

- Types of amyloidosis that affect the heart
 - Immunoglobulin light chain amyloidosis (AL),
 - Familial transthyretin-related amyloidosis (ATTR)
 - Senile systemic amyloidosis (wild-type TTR)
 - Secondary amyloidosis (AA)
 - Isolated atrial amyloidosis (atrial natriuretic peptide)

Prognosis depends on type of amyloid and extent of cardiac and systemic involvement



Imaging Findings

Echocardiogram

- Concentric thickening of the left ventricle, an abnormal bright echotexture, and/or asymmetric thickening of the septum
- Bi-atrial enlargement, valvular thickening, right ventricular hypertrophy, and small pericardial effusion may be observed
- With a small left ventricular cavity, low-gradient aortic stenosis is suspicious for hereditary amyloidosis
- +/- systolic and diastolic dysfunction

Contrast-enhanced cardiac MRI

- Helpful in infiltrative cardiomyopathies; gadolinium-based contrast can reveal abnormal patterns suggestive of cardiac amyloidosis
 - T1 signal may significantly be increased with amyloid deposition
- Inability to "null" or blacken the myocardium has been suggestive of cardiac amyloidosis.
- Left ventricular late gadolinium enhancement is very common
- Other features include, patchy myocardial scarring, right ventricular thickening, bi-atrial enlargement, and delayed enhancement of respective structures

SPECT (single-photon emission computed tomography)

- Tc-99m PYP bone tracer uptake graded visually with scores ranging from 0 to3
 - 0 no cardiac uptake
 - 1 cardiac uptake < rib uptake
 - 2 cardiac uptake = rib uptake
 - 3 cardiac uptake > rib uptake
- Contralateral lung uptake ration > 1.5 at 1 hour is also diagnostic
- Planar SPECT confirms myocardial tracer retention



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