IMAGING TOOLS FOR HEART FAILURE

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Disclosures

• No relevant disclosures or conflicts
Case – Mr. CA

• 71 year-old man
• CAD – inferior MI 6 years prior with PCI BMS to R coronary artery
• Hypertension
  • Poorly tolerant of ACEI
• Hypercholesterolemia, ex-smoker
• No diabetes
• Carpal tunnel syndrome
• Decompression surgery for lumbar spinal stenosis
Case – Mr. CA

- Referred for hip replacement surgery and noted to have progressive dyspnea on exertion at preop evaluation
  - Myocardial perfusion scan – Normal LVEF, small prior inferior infarct, no ischemia
- Decompensated HF post hip surgery
- Systolic murmur noted on physical exam
- Echocardiogram performed
E vel 0.9 m/s, E/A 1.1  
Septal e’ 6 cm/s, E/e’ 15  
Lateral e’ 7 cm/s, E/e’ 13

In patients with normal LV EF

- Average E/e’ 14
- e’ velocity reduced
- TR velocity not measurable
- LA vol index 35 mL/m²

- 50% positive
  - Indeterminate diastolic function

Nagueh et al. JASE 2016;29:314
What is the most likely diagnosis?

- Post-operative volume overload without cardiomyopathy
- Ischemic heart disease
- Hypertensive heart disease
- Hypertrophic cardiomyopathy
- Infiltrative cardiomyopathy
Echo Summary

• Technically difficult study

• Normal LV size and EF
  • Increased septal wall thickness
  • Dynamic apical function

• Systolic anterior motion of the mitral valve
  • No significant MR or LVOT obstruction

• Consider cardiac MRI for evaluation of possible Hypertrophic Cardiomyopathy
Cardiac MRI

- Asymmetric basal septal hypertrophy (max thickness 17 mm)
- SAM of the mitral valve
- Mid-wall patchy LGE consistent with fibrosis
- Overall findings consistent with septal variant HCM

Based on findings:
- Nitrate stopped
- β-blocker uptitrated
- Symptoms progress, dyspnea and now exertional chest pain
- What to do?
In Months Following

• Developed worsening severe exertional dyspnea and new angina

• Coronary angiography
  • Severe 3VD

• Left heart cath
  • LVEDP 25 mmHg
  • 50 mmHg gradient between apical and basal LV cavity
  • LVOTO gradient induced with nitrates of 20 mmHg
  • Brockenbrough phenomenon noted

• Referred for CABG +/- myectomy

Brockenbrough Phenomenon
http://www.clevelandclinicmeded.com
Intra-Operative TEE

Esmolol – HR 50 BPM

Isoproterenol – HR 70 BPM

Low Blood Pressure

Increased Blood Pressure
Case — Mr. CA

• CABG x3, no myectomy
• Post-operative prolonged due to persistent hypotension and dyspnea
  • Poorly tolerant of β-blocker and ACEI
• Anemia noted, acute kidney injury
• Repeat echo performed
Case – Mr. CA

- Global longitudinal systolic strain
  - Impaired -8% (normal ≈ -20%)
  - Basal-apical strain gradient
  - Suggestive of cardiac amyloidosis
Strain Analysis – Speckle Tracking Echo

• Change in deformation
• Typically long-axis systolic shortening – longitudinal strain
  • Negative value
Strain For Cardiac Amyloidosis

Diagnosis

Bellavia et al. AJC 2008;101:1039

Prognosis

Bellavia et al. JASE 2010;23:643
Case Mr. CA

• Cardiac amyloid suspected preoperatively, myocardial biopsy requested
• Atrial biopsy positive for green birefringence under polarized light by Congo Red stain

• What comes next?
  • Bone marrow biopsy
  • Fat biopsy
  • Nuclear imaging
  • Repeat cardiac MRI
  • No further testing needed
Cardiac Amyloidosis

• Large group of disorders caused by extracellular deposition of insoluble abnormal fibrils composed of misfolded proteins of different types

• Most common amyloidosis causing cardiac involvement
  • AL (primary) – Immunoglobulin light chain
  • ATTR – Transthyretin
    • Mutant
    • Wild-type
  • IAA – Isolated atrial amyloid, atrial natriuretic peptide
    • Atrial arrhythmias, rarely causes heart failure
  • AA – Associated with chronic inflammatory conditions, causes more renal amyloidosis, cardiac involvement rare

Falk et al. JACC 2016;68:1323
Cardiac Amyloidosis Types

Primary AL

- Immunoglobulin
- Free light chains
  - Target of serum FLC immunoassay
  - Hidden epitopes
- Heavy chain
- Light chain
- Disulphide bonds

Transthyretin (ATTR)

- "Neurologic"
- "Cardiac"

Familial (Mutant)

- Transthyretin (Mutant)
- Transthyretin (Wild-Type)

Wild-Type

- Transthyretin (Wild-Type)

Rapezzi et al. Eur Heart J 2013

Merlini et al. NEJM 2013;7:583

Courtesy of THAOS Registry
Technetium 99m Pyrophosphate Scan
Nuclear Imaging

• Technetium 99m PYP (NA), or DPD or HMDP (Europe) bone scintigraphy
  • Sensitivity 99% for ATTR CA
  • Specificity 64% for ATTR CA due to none-mild update in AL, AApoA1 CA
    • Improves if a greater intensity of uptake is used as diagnostic criteria
    • Rises to 99% when high intensity uptake is combined with negative immunofixation electrophoresis and free light chain assay

Gillmore et al. Circ 2016;133:2404
Tc 99m PYP Scan

Planar whole body scan

- Uptake ratio >1.5 is >95% sensitive for ATTR
- **Cannot differentiate** Familial from Wild-type

With SPECT

Falk et al. JACC 2016;68:1323
Gillmore et al. Circ 2016;133:2404
CMR for Cardiac Amyloidosis Detection

- Morphologic findings similar to echo
- Late gadolinium enhancement
  - Difficulty nulling the myocardium following injection
  - May be subendocardial, transmural, patchy or absent
  - Extent of LGE reflects prognosis

Boynton et al. JACC CV Img 2016;9:680
Kwong et al. Am J Cardiol 2015;116:622
CMR T1 Mapping

Visual T1 assessment

Quantitative T1 mapping
CMR – T1 Mapping and LGE Imaging

Fontana et al. Circ 2015;132:1570
Cardiovascular Amyloidosis Manifestations and Clues

• ECG – ‘may’ be helpful

• wt-ATTR - ↑ risk of **aortic stenosis**
  • High risk AVR, even for TAVR

• Increased risk of CAD

• Rhythm disturbances
  • Atrial fibrillation
  • Conduction disease
  • Ventricular arrhythmia

• Stroke

• **Imaging is key** for confirming clinical suspicion and guiding diagnostic evaluation
Case - Mr. Cardiac Amyloidosis

- Dx ATTR cardiac amyloidosis – presumed Wild-type
  - Declined genetic testing
- Reduced beta-blocker and stopped ACEI, uptitrated diuretics
- Postop Afib – anticoagulation
- Needed repeat lumbar spinal decompression and carpal tunnel syndrome release surgeries
- Peripheral neuropathy
- Chronic abdominal bloating and discomfort
- Has remained compensated from HF perspective
Cardiac Amyloidosis – Imaging Evaluation

- **Echo** – global longitudinal strain pattern showing apical sparing is highly specific
- **MRI** – LGE imaging very helpful, but different patterns of enhancement may be present
- **Nuclear scintigraphy** – Tc99m-PYP scan very accurate for ATTR
- Look for clinical clues
  - **AL** – periorbital bruising, macroglossia, nephrotic syndrome, peripheral or autonomic neuropathy
  - **ATTR** – peripheral or autonomic neuropathy, carpal tunnel syndrome, spinal stenosis, gastrointestinal complaints
Imaging Tools For HF Workshop

• Thank you

• Questions / comments?

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