3/07/22 Morning Report with @CPSolvers

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CC: Fatigue and exertional dyspnea

HPI: 74yo M presents with a 2-month history of fatigue and exertional dyspnea. He used to walk 3 miles daily but has cut back over the past few months, initially limited by back pain but now also due to SOB. He denies chest discomfort or palpitations. He has positional lightheadedness but no syncope. He had been on losartan 100 mg daily and it was reduced to 25 mg daily due to orthostasis. He denies orthopnea or paroxysmal nocturnal dyspnea but has noted ankle edema at the end of the day. He denies cough.

PMH: CAD- 10y prior, he presented with angina and had a stress test showing anterior ischemia, resulting in angiogram and LAD PCI. A-fib s/p ablation 5y prior with sick sinus syndrome requiring pacemaker
PMR-diagnosed 6 months prior, on a slow prednisone taper. CTS, bilateral, with surgery 3 years prior. Lumbar spinal stenosis with surgery 8y prior.

Meds:Aspirin 81 mg/Atorvastatin 40 mg
Losartan 25 mg/
Prednisone 4 mg

Fam Hx:his father passed away in his sleep at the age of 61; the cause of death was presumed to be a "heart attack."

Soc Hx: He was born and raised in Chicago and is a retired civil engineer. He lives at home with his wife. He has 2 children and 2 grandchildren. He has never smoked cigarettes and does not drink alcohol or use illicit drugs.

Vitals: **T**: **HR**: 92 **BP**: 106/80 **RR**: **SpO_2**: 95% room air
Exam:
Gen: Thin, no distress
Neck: JVP 12 cm H2O
CV: RRR, no murmurs, +S4 gallop
Pulm: lungs clear to auscultation
Abd: soft, nontender, no organomegaly
Extremities/Skin: warm with trace ankle edema

Notable Labs & Imaging:
Hematology:
WBC: 5.6 Hgb: 11.2 Plt: 210

Chemistry:
Na: 140 K: 4.5 Cr: 1.3 with eGFR 50
Troponin I 0.3 ng/mL BNP 540 ng/mL

Imaging:
Angiogram: patent LAD stent, no other disease
Echocardiogram – LV septum 1.9 cm, PV posterior wall 2.0 cm (severely increased LV wall thickness), increased RV wall thickness, EF 48%, peak global longitudinal strain -10% (low) with apical sparing, RVSP 52 mm Hg.
Technetium pyrophosphate scan –positive scan
Monoclonal light chain screen: no monoclonal band by serum/urine immunofixation electrophoresis but kappa/lambda ratio 2.3
Endomyocardial biopsy: amyloid by Congo red staining, LC/MS consistent with TTR amyloid. Genetic testing: V122I mutation

Final diagnosis: Amyloidosis
Patient started on tafamidis and enrolled in a clinical trial of a new agent, eplontersen.

Problem Representation: 74yo M with a hx of CAD with PCI LAD 10y/o and afib s/p ablation 5y/o +SSS requiring pacemaker and PMR recently and on Prednisone presenting with chronic fatigue and exertion dyspnea with diminished functional status due to dyspnea with peripheral edema developing towards the end of the day.

Teaching Points (Seyma Yildirim⭐):

- **Exertional dyspnea**: Start w/ pathophysiology! -> heart (muscle (e.g. CM), artery, valves, electrical system, pericardium) - lung - anemia - decondition
  - Heart: muscle (CM), CAD (dissection, atherosclerosis, thrombotic), Valves (aortic stenosis, mitral regurgitation), electrical (Bradyarrhythmia, aFib)
- **Framework for CAD+ Afib** => Coronary, arrhythmia?, Sick-Sinus syndrome
- **Examination:** Consider JVP(elevated?); S3 is usually a bad sign!, Lung (Crackles present?: e.g. due to atelectasis, refractory HF)
- **First clues:** Prednisone (fluid retention -> CM), fam hx (heart attack: MI, arrhythmia?), JVP+ BNP elevation → consider CM, rather no arrhythmia
- **Thick walls on Echo:**
  - 1.**more muscle** (LVH -> athletes, aortic stenosis, HTN, HCM) or
  - 2. **infiltrating heart dz** (adult: e.g. Fabry, Amyloid, Hemochromatosis, lipid storage dz, sarcoidosis)
- **Collecting the clues for infiltration in terms of amyloidosis:** Troponin elevation due to infiltration of heart muscle, Low RR: autonomic dysfunction (infiltration of nerves) + Carpal tunnel syndrome
  - **Consider:** Protein infiltrates heart (restrictive CM w/ HFpEF), nerves (sensory neuropathy), GI-tract (diarrhea, constipation), muscle etc.
- **Amyloidosis** involving heart → 2 main forms are **TTR** (mutation or wild-type), **AL** (plasma cell dyscrasia) → **restrictive CM**
- **Dx of amyloidosis:** Global longitudinal strain (**apical sparing**), **Technetium pyrophosphate scan** (only if light chains are negative!), **monoclonal light chain screen** (serum, urine, Serum-free light chains, Immunofixation)
  - monoclonal light chain can be missed in SPEP -> immunofixation is more sensitive
  - Biopsy (Congo-red stain) is not always necessary!
- **Volume overload:** heart (valvular, pericardial lesion?, obstructive lesions like myxoma)- liver - kidney
- **Treatment of amyloidosis:** Silencer and Stabilizer!
- Infiltrative heart dz: perform cardiac MRI as a general screen