



9/26/24 Morning Report with @CPSolvers

“One life, so many dreams” Case Presenter: Ann Marie Kumfer (@annkumfer) Case Discussants: Rabih (@rabihmgeha) and Hee (@heemun8)



CC: 60 YOM with SOB and orthopnea
HPI: Felt well and was able to walk for several miles up until 2 months ago. 2 nights ago, profound SOB prior to bed that lasted 20 minutes. Breathing better upright, worse supine to the point that he started sleeping in a recliner. Mild LE swelling, weight stable. Swelling is generally worse in the summer and better in the morning, worse after standing on the legs.

ROS:
No fevers or chills. No eye weakness, double vision, dysphagia or food getting stuck, arm/leg weakness, or neck masses. No sleep apnea. No worsening abdominal distension. No thoracic surgery. Works in a car shop, and did report small drop of oil in his mouth before symptoms started. May be mold in the house but reports it is mild. Denies other exposures.

In the ED:
Went to the CT scan and became diaphoretic and SOB. Patient said “I thought I was gonna die.” No oxygen monitor in the room, but when got back to the ED, O2 was 93%

PMH:
HTN (well-controlled)

Meds:
Lisinopril 20mg

Fam Hx:
None
Soc Hx:
No foreign travel. No hx of incarceration
Health-Related Behaviors:
30-pack smoking history (quit 10 yrs ago). 1-2 beers/week
Allergies:
None

Vitals: T: 36.6C HR:70-90s BP: 130/60-90 RR: 16 SpO2: 96% on RA BMI: 40
Exam:
Gen: Sitting on a chair
Eyes: EOMI. No nystagmus.
Neck: No thyroid masses or enlarged lymph nodes.
CV: RRR, no murmurs. Unable to see JVP.
Pulm: Diminished breath sounds, worse on R. No crackles
Abd: nontender, no distension
Neuro: no focal weakness, able to move arms and legs with strong hand grip. Sensation intact. Preserved patellar reflexes
Extremities/skin: 3+ pitting edema on ankles. 1+ pitting edema on tibia

Notable Labs & Imaging:

Hematology:
WBC: 9.4 (normal diff) Hgb: 13 Plt: 450
Chemistry:
Na: 141 K: 4.4 Cl: 107 HCO3: 29 BUN: 13 Cr: 0.98 glucose: 102 Ca: nl Mag:
AST: nl ALT: nl TSH: nl A1c: 5.5
High-sensitivity troponin: 7 (delta of 0) CK: 244
BNP: 15 UA: Specific gravity 1.02, otherwise unremarkable.
VBG: pH 7.37 CO2: 54 COVID, RSV, influenza: negative
Imaging:

CXR: mild cephalization of pulmonary vasculature
Echocardiogram: normal EF, exaggerated ventricular septal motion with respiration differential including constrictive vs obstructive disease
CTA: No signs of PE. No pericardial effusion. No coronary or pericardial calcifications
PFT: FVC 44%, FEV1 43%, FEV/FVC 99. TLC 31% VC 37%. DLCO 52% (adjusted). Normal MIP/MEPs
Sniff test: R hemidiaphragmatic paralysis
MRI cervical spine: Some disc disease. No cord signal
B1, B6, B12, copper, zinc: normal. ANA, myositis, SPEP, HIV, Hep: normal
Improved with NIV. 10 days after discharge, noticed droopy eyelids, intermittent blurry vision, fatigue in chewing. AChR: 15.4. Started on tx
Dx: **myasthenia gravis**

Problem Representation:

60 YOM with history of HTN p/w dyspnea, orthopnea, and PFTs c/w restrictive pattern f/t/h myasthenia gravis.

Teaching Points:

Orthopnea

Diurnal or positional - two possibilities from a pathophys perspective:
venous return/preload >> diaphragmatic
Specific diseases: LV dysfunction/HFrEF, COPD/Asthma (vagal tone), Neuromuscular/Diaphragmatic weakness, OSA, Obesity Hypoventilation.
In the absence of extra-diaphragmatic weakness, paradoxical abdominal breathing (advanced disease!) and negative sniff test **neuromuscular** etiologies are even less likely!
Typical chronicity of LV dysfunction: Slow reduction in exercise capacity followed by a more acute phase of congestion, namely LE edema, pulmonary edema, ascites precox (an acute presentation would sway us towards hyperacute HF - ACS, tamponade, arrhythmia - or neuromuscular diaphragm disease)

Tension of severe, visible SOB and hypoxemia when lying flat

venous return/preload < diaphragmatic
Heart: the underlying LV dysfunction would have to be very severe to be not able to deal w/ a bolus of 500mL

In the absence of apparent LV dysfunction (echo, CT chest) and severe aggravation when lying flat a diaphragmatic disorder is most likely.
PFTs showing restriction: Think external vs internal disease. A nl DLCO supports the hypothesis of external disease, but ca low DLCO can still be in line with external disease causing atelectasis. → R hemidiaphragmatic paralysis. Symptomatic diaphragmatic is often bilateral - suspicion that BMI contributing to the severity of the presentation.

And then we get our extra-diaphragmatic muscle/NMJ issue...