7/18/25 Morning Report with @CPSolvers

"One life, so many dreams" **Case Presenter**: Youssef (@saklawiMD) **Case Discussants**: Rabih (@rabihmgeha) and Reza

Scribing (Vale)
CC: Progressive somnolence

HPI: 35yo M p/w progressive somnolence. In the ED very somnolent barely arousable. VBG pH 7.04 pCO2 117. Placed on BIPAP.

On previous admission was placed on 4L NC oxygen and nocturnal BIPAP wo improvement.

Initially presented w/ 2 weeks of worsening dyspnea, productive cough, nausea, vomiting, diarrhea and chills.

Patient was diuresed, developed hypotension. Was admitted to the ICU started on dobutamine and continued diuretics.

PMH: HFmrEF (45-50%). Severe RV dysfunction.

Meds: Only inhalers at home.

Fam Hx: Has 3 kids.

Social Hx: Supermarket manager.

Health-Related Behaviors: No EtOH, no tobacco.

Allergies:

Vitals: T: BP: HR: RR: Sat: BMI: 17
Exam: Gen: Cachexia
HEENT:
CV: Loud P2.
Neuro: Upper proximal extremity weakness (deltoids, triceps). Distal LE weakness (L>R dorsiflexion) unable to toe or heel walk. ROS decreased.
MSK: No edema

Notable Labs & Imaging:
Hematology:
WBC: Hgb: 11.5 Plt: MCV:95

Chemistry
Cr: nl **HCO3:**>45 ; **Am cortisol:** nl.
SPEP/UPEP: Normal. **ABG:** ph 7.24 pCO3 138. **CPK:** nl.

Imaging:
CTPE protocol on initial admission: bilateral subsegmental PEs, large pleural effusions (R>L) and severe right heart enlargement.
Echo: EF 40% Severe R heart disease
RV Cath: RA 22/21, RV 65/10, PA 63/40, PCWP 28/24, PVR 10.47, SVR 2413 PAPI 1.21, RA/PCWP ratio: 0.76.
Cardiac MRI: Findings consistent with pulm HTN as evidenced by the severely dilated R chambers w/ moderate RV dysfunction + systolic septal flattening and diastolic septal bowing. Enhancement along the entire septum and extending to the anterior wall of the RV which is too extensive to be only 2ry to pulm HTN.

EMG: Myopathic process in the humeroperoneal distribution.

Gene panel: Nemaline myopathy type 1 (NEM1). Heterozygous mutation in TPM3.
Dx : Nemaline myopathy type 1 (NEM1).

Problem Representation: 35yoM p/w somnolence, dyspnea and hypercarbia. Exam showed proximal and distal weakness. Exams suggested Pulm HTN. EMG revealed myopathy and a gene panel confirmed the NEM1 dx.

Teaching Points (Lera):

Confusion <-> hypercarbia:
- Is hypercarbia ***acute / chronic / acute on chronic?*** Look at HCO3
- Lung disease causing confusion: ***chronic >> acute.*** Acute -> think about other cause -> *primary CNS* (test of treating).
- ***Pleural effusion*** can contribute to hypoventilation.

Why overnight O2?
- Look at the ***weight and age*** (COPD vs. OSA).
- Increased O2 ***decreases pulmonary hypertension.***

RV failure etiology: MCC – ***LV dysfunction.***
- *+ mildly reduced EF* -> ***focus on RV.*** Is the lesion *upstream* (LV -> pulmonary vessels) or *at the RV itself?* RH cath can help.
- ***LV + RV:*** *LV -> RV* failure // *RV -> LV* failure (in long standing pulmonary HTN) // *simultaneous* problem (myocarditis).
- In patient with vulnerable RV ***any process*** in the lungs can cause RV failure exacerbation.

Reading the RH cath:
- ***Is this pulmonary HTN?*** -> MAP. ***Is the LH responsible?*** -> PCWP.
- ***RA / PCWP*** -> precapillary > 0.8 > postcapillary.

Hypotension with diuresis:
- ***Severe HF.*** *Not enough diuretics* -> cardiogenic shock + inotropes
- ***Preload dependent*** HF (*obstruction* -> some degree of hypervolemia needed to bypass the block) // ***not HF*** at all.

Cardiac enhancement of MRI: Pattern matters -> *along the coronaries* (ischemia) vs. *random* (infiltration).

Weakness DDx:
- [Hyporeflexia -> ***PNS***] + proximal >> distal -> ***muscles*** = *NMJ*
- ***Skeletal muscle + cardiac muscle involvement*** -> hereditary muscular disease (eg myotonic muscular dystrophy).
- Nl CK or hyporeflexia ***≠ not muscular disease.***