9/11/23 Rafael Medina Subspecialty VMR with @CPSolvers

"One life, so many dreams" Case Presenter: Dr. Dimple Shah Case Discussants: Dr. Mangala Narasimhan (@mnarasimhan)

CC: 34 year old female 16 weeks pregnant p/w SOB

HPI: Progressive dyspnea on exertion for about 3 months
Small activities such as pushing a stroller is causing difficulty breathing
Turning point: just walking caused severe SOB, something that did not happen a few days priors
Urgent care: swabbed for covid, negative.
Oxygen was on the lower side, sent to ED.
First pregnancy 2 years ago: vaginal delivery, hypoxic postpartum, received diuretics and went home.

PMH:
Latent TB - treated at age 4

Meds:
Prenatal vitamins

Allergies:
none

Fam Hx:
No lung dz, no Ca hx
Father w/ HTN
1st baby born healthy
No prior miscarriages

Soc Hx:
not working currently, born in NYC, no recent travel

Health-Related Behaviors:
no smoking, drinks socially, no pets, no known exposures,

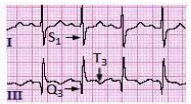
Vitals: T: afebrile **HR:** 100 **BP:** 123/86 **RR:** 24 **SpO2:** 95% ra **BMI** 32
Exam:
Gen: well-appearing, speaking comfortably, no icterus, no LAD
CV: RR **Pulm:** Crackles bilateral bases
Abd: Palpable uterus below umbilicus
Neuro: AOx3, no focal deficits
Extremities/skin: No edema, no obvious rash

Notable Labs & Imaging:
Hematology:
WBC: 9.2 Hgb: 11.3 Plt: 227
Chemistry:
Na: 137 K: 3.8 Cl: 107 CO2: 19 BUN: 5 Cr: 0.46 Ca: 8.8 Phos: 3.7
Mag: 2.1 AST: 32 ALT: 15 T. Bili: 1 Albumin: 4.0
UA: Unremarkable HIV: Negative
Imaging:
EKG: sinus tachy, no signs of PE (s1q3t3 - cor pulmonale; right heart strain)
CXR: Largely unremarkable
POCUS: A line pattern bilaterally, no effusion.
Echo: Normal LV and RV function, chamber sizes, no evidence of PH nor valvular disease. Bubble study is negative.
Walking 100 feet ➡ saturation dropped to 70%
CT: uniform thin walled cysts all evenly throughout the lung fields
VEGF-D inconclusive range
Renal US: normal
FEV1 < 70%

Final dx: Lymphangioleiomyomatosis (LAM)
Referred to outside institution. By week 21 condition worsened, 5 liters on NC.

Problem Representation: 34 yo F, pregnant at 16 weeks, presenting with progressive SOB over the past 12 weeks, found to be hypoxemic. CT scan with diffuse cysts.

Teaching Points (Debora):
Pregnancy: autoimmune diseases, cardiomyopathy, pulmonary edema, hormone receptor gets positive in pregnancy.
- BP and UA can suggest preeclampsia.
- During pregnancy WBCs (esp. PMNLs) & Hct increase, but plasma volume expands more so there is physiologic anemia
Ddx: Postpartum cardiomyopathy, PE, DVT, interstitial lung disease (she is stress and can manifest now), progressive lung cancer, congenital heart disease, Pulmonary hypertension. Infection vs Malignancy vs Structural
TB can come back? We should expect some cough, or symptoms before the pregnancy. And there is no manifestation on CXR and Pocus.
During the pregnancy: Physiologic dyspnea vs. not physiologic (hypoxemia is not normal)
Pregnancy + Pulmonary Hypertension: Autoimmune diseases can cause pulmonary hypertension, increase DVT causing Pulmonary hypertension. Can happen for unclear reasons, and present more during pregnancy.
PE can present on EKG with the pattern S1Q3T3



The $S_1Q_3T_3$ pattern

Sat drop 70% walking: Ddx ILD, PCP Pneumonia.
LAM: Not common disease. Dyspnea is the common presentation, besides spontaneous pneumothorax, pleural effusion, chylothorax. Cystic lung disease bilateral, age (30s), get worse in pregnancy with estrogen.
Cystic pattern: Cystic on the top to the bottom, different sizes, everywhere in the lung. Look the kidney too and order the marker VEGF-D.
Treatment: Sirolimus (not in pregnancy!!) Stop the progression do not stop the disease.