02/07/24 Morning Report with @CPSolvers

"One life, so many dreams" Case Presenter: Vijay Balaji (@vijaybramhan) Case Discussants:Steph (@StephVSherman), Zaven (@sargsyanz) and Hans

CC: Complete or incomplete (Clue):
24 M presents with low grade fevers, SOB since one month, multiple joint pains since 1 month, significant weight loss since 2 months

HPI: Low grade fevers, no chills or rigors, no significant improvement with OTC antipyretics. Progressive SOB, able to walk only 2 flight of stairs. Associated with episodes of dry cough since 15 days. Joint pains - predominantly small joints - wrist, hands, lower limbs of feet ; no swelling or redness, no stiffness / early morning stiffness. Had 2 episodes of streaky hemoptysis. Denies ulcers, joint deformities, photosensitivity. No h/o rashes or livedo.

PMH: Recently treated for unprovoked DVT with submissive PE 2 months back (Echo: Lesion in RV - mobile structure)

Meds: Rivaroxaban

Fam Hx: NA

Soc Hx: Denies smoking, not sexually active

Health-Related Behaviors:

Allergies: NA

Vitals: **T:** º **HR:** 78 /min **BP:** 120/ 70 mmHg **RR:** 27/min ; SpO2- 97% on RA

Exam:

Gen:

HEENT: Oral ulcers ; no other lesions

CV: Systolic murmur in the left lower sternal border.

Pulm: wnl

Abd: wnl

Neuro: No focal neurological deficits, Rest of exam wnl

Extremities/skin: wnl

Notable Labs & Imaging:

Hematology:
WBC: 5400 Hgb: 12 ; MCV - 78 Plt: 286; normal differential

Chemistry:
Na: wnl K: wnl Cl: wnl HCO3: wnl BUN: wnl Cr: wnl glucose: wnl Ca: wnl Mag: wnl ;
AST: wnl ALT: wnl Alk-P: wnl Albumin: wnl
PT- 15.9 ; INR-1.2 ; PTT - wnl ; TSH - wnl
ANA, ANCA, Antithrombin, Protein C, Protein S, Beta 2 glycoprotein, cardiolipin, lupus anticoagulant, Factor V Leiden: wnl

Imaging:
EKG: Sinus rhythm
Echocardiogram: Echogenic content attached to RV wall, RVSP - 35 mm Hg, no significant RV/ LV dysfunction
US Lower limb - Thrombosis in the femoral veins
CT: P/A >1. Acute segmental thrombus in B/L LL. Mildly dilated Right bronchial artery. Multiple GGO in BL lower lobes.
Blood cultures - negative; HIV, HBV, HCV - non reactive
HLA B51 - Positive
Cardiac MRI - small organized thrombus in RV ; T2 hyperintense areas in perihilar locations and superior segment of RLL of lung
CT: Multiple pulmonary aneurysms in RLL and segmental branches of RML and LUL
Pathergy - negative

Dx: Hugh Stovin Syndrome (variant of Behcet - Clue: Complete/Incomplete)

Problem Representation: 24 M with PMH of recurrent thrombotic phenomena (DVT, PE) presented with low grade fevers, joint pains, SOB and hemoptysis since 1 month.

Teaching Points (Kuchal):

1. The approach to understand this case would be first understand SOB/DOE and then tie it up with Fever, joint pains and weight loss.
2. Hemoptysis: ? due to PE. **Patient was on anticoagulants at the time of the episodes, broadens the cause of it.** This calls for r/o **LUPUS/ APS: Rx of choice is Warfarin. It could also be because of VTE (a recent h/o),** PE can cause DAH.
3. Hypercoagulable states are usually reflected in the hematologic footprint: MAHA, Myelofibrosis, leukemia etc. or other cancers.
4. Unprovoked DVT: could be due to rheumatologic disease, needs to be ruled out.
5. Oral ulcers (painless) Behcet', malignancy,
6. Mobile RV: a) Mobile clot. b) more common actually in the RA. c) Consequence of Eosinophilic Myocarditis, d)
7. Behcet's DVT: Stable thrombus in the femoral vein. But so far the patient hasn't got recurrent ulcers.(which are present in 70%) ?aneurysms in the Pulmonary vessels (rare< 5%) Associated with HLA B5, HLA B51.
8. Other causes to consider are Estovan syndrome, PNH.Review the Recurrent Thrombus schema.
9. Hugh Stovin Syndrome: Rare autoimmune disease, a variant of Behcet's. Usually presents with multiple venous thrombi, thrombophlebitis, pulmonary and bronchial aneurysms.Death is usually due to Massive pulmonary hemorrhage, if not diagnosed early and treated properly. Treated with immunosuppressant(Cyclophosphamide) and steroids.