9/22/23 Morning Report with @CPSolvers

"One life, so many dreams" ***Case Presenter****: Noah Nakajima (@Noah_Nakajima)* ***Case Discussants****: Rabih Geha (@rabihmgeha)*

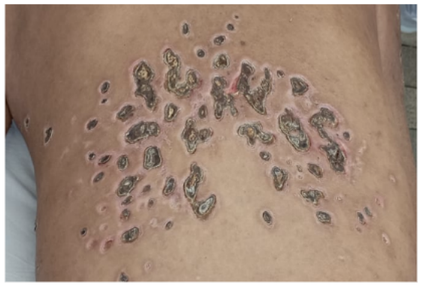
CC: 34M who presented with subjective fevers, violaceous facial infiltration, nausea, vomiting and arthralgias

HPI: 2y ago started having circular erythematous painful lesions in same skin distribution which with time became ulcerated. 1y ago began experiencing subjective fevers, fatigue and generalized arthralgias which where associated with a 15kg weight loss in the last 3m. Presented to outside hospital ED where he experienced a convulsive episode which prompted transfer.
Prescribed painkillers, discharged w/ close follow up after results came back.
On Follow up patient looked acutely ill. Ulcers increased in number and severity. This was associated with new onset Dyspnea and Hypoxemia.
Evolution: 1) Because of CTPE findings, started on empiric anticoagulation for PE. Dyspnea improved with IV diuretics. 2) Started on pulse steroid therapy w/ overall improvement but with worsening skin rash. H complicated by skin infections, biliary sludge. Started on lepromatous reaction treatment.

PMH: Syphilis 10y ago, treated
Meds: -

Health-Related Behaviors: Previous crack cocaine user (10y ago). Current cannabis smoker, Current Tobacco use (since 18y old)

Vitals: T: afebrile **HR:** 102 **BP:** 120/80 **RR:** normal on room air
Exam:
Gen: in pain/ **Neuro:** normal
Extremities/skin: ulcerative lesions in both arms, lateral face (deltoid area) and back (see picture)



Notable Labs & Imaging:
Hematology:
Hb 8.4 (12.4 previous), WBC 3.8, Platelets 256k
ALT: 152, AST: 43 GGT: 206
Blood cultures: normal
CSF: unremarkable
Skin biopsy: chronic dermatitis
PCR 12, LDH 694, Albumin 2.3, Ferritin: 3105, ESR 120, UA: Protein/RBC +, UPCR 0.94, Triglycerides 586, CK 1198, Peripheral blood Smear: schistocytes, C3/C4 low, Hepatitis serologies negative.
HIV -. , ANA: positive 1/1280
Anti- SSA +, anti- U1RNP +, anti- Smith +, Scl 70 -, Jo1 -, anti- DNA + (1/40), Lupus anticoagulant +.
EMG: myopathic pattern
Repeat skin biopsy: mixed inflammatory infiltrate w/ histiocytes, lymphocytes and neutrophils in dermis. Single vessel w/ luminal suboclusion in process of recanalization. Subc. with fibrosis and foamy histiocytes. No vasculitis, BAAR negative, no granulomas
Leprosy: serology positive, skin shaving DNA positive, + bacilli.
Imaging:
CXR: Cardiomegaly.
Echocardiogram: R ventricular dilation and dysfunction
CTPE: Interseptal thickening, increased pulmonary artery caliber, voluminous pericardial effusion and bilateral pleural effusions
Heart cath: PSAP 50, w/o response to NO

Dx: SLE + Leprosy

Problem Representation: 34M w/ 2y history of ulcerative skin lesions presents with fever and generalized arthralgias found to have positive SLE and Leprosy serologies and skin biopsies.

Teaching Points (Ayesha):
- Look for life threatening findings: Airway occlusion as a result of facial edema. If they're positioned with head up, most likely no airway occlusion.
- Is swelling unilateral (localized - infections) or bilateral (systemic - allergic, anaphylaxis), any evidence of plethora or venous engorgement - indicative of *SVC syndrome*.
- **Typical Ulcers:** Due to 1) Arterial insufficiency (Lateral - lack of perfusion), 2) Venous insufficiency (Medial - lack of drainage), 3) Excessive pressure on skin. → Present as: Lower extremities, and are slowly progressive
- **Atypical Ulcers:** Skin necrosis with no issue with healing or perfusion → usually due to inflammation, infection, or nonatherosclerotic vascular diseases.
- The longer a disease goes without catastrophic deterioration, the more likely it's autoimmune in nature.
- ***Commensal organisms*** (staph, strep - focal infections (penum, ecthyma (only endogenous organism that causes a widespread ulcer)) vs ***exogenous*** from outside of body that cause widespread symptoms (fatigue, myalgias)
- Widespread ulcer most likely isn't from a commensal organism -Usually from → (viral - but not for a long duration. Bacterial (acute life threatening - e.g. anthrax, plague), fungi, parasites.
- Visceral involvement with cardiopulmonary compromise: disease escalation is alarming. If there is pulmonary hypertension, Is it acute or subacute or chronic?
- The thicker the RV, the longer the RV has been adapting to the pressure overload → look at echo. Is there RV thickening? Heart cath can tell with a high degree of certainty.
- PAH: Left heart disease or intrinsic heart disease
- Young demographic: Pulmonary arterial hypertension class 1
- Pulmonary hypertension and ulcers: HIV (important here), cancers → externally occlude or intiterally block the artery. Microscopic malignancy - tumor thrombotic microangiopathy
- Rash around face → consider scleroderma and dermatomyositis (MDA5) → AI testing - get ANA, CK (because high chance of myositis - dermatomyositis), SPEP analysis would be helpful.
- acute anemia in weeks/days → blood loss/hemolysis (prioritized - if smear shows schistocytes → MAHA (could they have **Lupus with APLS *OR* Scleroderma with renal crisis**?
- CK - muscle involvement→ clues us towards inflammatory causes.
- involvement of small vessels and medium vessels in kidney - consider overlap of lupus with glomerulonephritis, scleroderma, dermatomyositis – careful with giving steroids (and cyclosporin) because they worsen scleroderma renal crisis → worsen renal blood flow that's already compromised, and increase the BP.
- Use ACEi to improve renal function for scleroderma renal crisis - more likely b/c of pericardial effusion
- Worsening rash after steroid tx despite overall improvement → Paraneoplastic process? Infection that's due to immunodeficiency? Consequence of a prior drug use from cocaine use?
- Leprosy - neuro and rash manifestations + foamy histiocytes.