06/19/24 Morning Report with @CPSolvers

***"One life, so many dreams" Case Presenter**: Sarah Haeger and Evan Zehr(@) **Case Discussants**: Dr. Melissa Griffith (@) and Dr. Geoffrey Connors (@)*

CC: 50 y/o male feel tired and muscle aches

HPI: He was in his general state of health since last month he feels weak and needs to lay in bed all the time, he had an elk hunting trip last month in southern Colorado, he developed fatigue/weakness and migratory arthralgias,in shoulder knees wrist hands and ankles. He is not able to do his normal task, this limits his ability to walks. He reports chills and sweats no hemoptysis. few days after his symptoms onset, he developed purplish discoloration around his eyes and knuckles along with dry cough and exertional dyspnea, limiting his ability to walk multiple blocks.

PMH: Gallstone pancreatitis s/p cholecystectomy

Meds: No

Fam Hx: No autoimmune diseases, cancer, clots

Soc Hx:

Health-Related Behaviors: lives in rural Colorado, works in agriculture. Exposure to Elk, Pigs, ducks, goats. incarcerated 30 yo. Marijuana occasionally, no alcohol/ tobacco

Allergies: none

Vitals: T: 37.7 **HR:** 68 **BP:**104/63 **RR:**22 **Sat** 97% RA

Exam:

Gen: Calm appearing, fatigue, no acute distress

HEENT: PERRL, violaceous rash periorbital, no LAD, moist membrane

CV: wnl **Pulm:** faint bibasilar crackles

Abd: unremarkable

Neuro:, AOx3, moving all extremities, 2+ patellar reflexes, sensation intact

Extremities/skin: no synovitis, no dactylitis, Motor exam limited due to pain. Violaceous discoloration over MCP knuckles on dorsum of hands, minimal on bl wrist, cracking and fissuring first 3 fingers, healed ulcers involving palmar surface of thumb bl, left index and middle pIP on palmar side. ⅘ strength bl upper and lower

Notable Labs & Imaging:

Hematology:

WBC: 10.4 PMN 72%, lymph 17%, eso 1% Hgb: 14.1 Hct 42.7 Plt: 194

Chemistry:

Na:136 K:3.5 Cl: 107 BUN:13 Cr: 0.96 glucose: 99 Ca:8.7 AST: 51 ALT: 45 Alk-P: 106

Albumin: 3.1 total bilirubin 0.9 total protein: 6

CK nl 38 ALdolase 4.5

infectious w/u: BC no growth, RVP neg, cocci neg, Q fever neg, hantavirus neg, bartonella Quintana and Henselae neg, quantiferon neg

Bronchoscopy: PMN 8%, lym 32%, Mono 39% , bac neg, fungal neg, AFB culture neg.

Autoimmune: ANA pos no titer, C3 109 (NI) , C4 43, RF neg, SS-B neg DS-DNA neg RNP neg, Chromatin neg , Centromere neg

Myositis panel: MDA5 Ab highly positive, SS-A positive, SSA-52 positive, rest of panel negative.

Imaging:

CXR: coarse bibasilar suggesting ILD pneumonia/ pneumonitis/ coarse in left midline zone opacities suggesting scaring.

Chest CT: Left upper lobe and basilar predominant peribronchovascular irregular consolidative and linear opacities with mild associated architectural distortion .

Dx: **MDA5 Myositis**

Problem Representation: 50y/o M from rural area presenting w/ fatigue, migratory arthralgia, periorbital rash, dyspnea. Found violaceous periorbital rash bibasilar crackles, violaceous discoloration of MCP joint, ANA, MDA5 Ab and SS52 pos.

Teaching Points (Elena):

1. **Fatigue vs. weakness**: Define those symptoms and objectify them
2. Think about **inflammation** in the context: Exposure (pets, travel, occupation), host characteristics
3. The lung is a "filter"!
4. Most **autoimmune** disease are **subacute**/chronic
5. **Myositis** = Inflammation of the muscle
   - Myopathy (esp. genetic) **vs** Myositis (inflammatory/autoimmune, infectious, neoplastic)
   - Bimodal age distribution: Juvenile/early adult **+** 50/60s
   - Useful questions: Are you having trouble showering or sitting up?
   - Skin manifestations: Gottron, heliotrope, V-sign
   - Exam: proximal>distal weakness
   - Definitions of myositis are moving towards an antibody base classification system, whereas early definitions focused more on organ involvement ("dermatomyositis")
   - ANA positive or negative
   - SSA can point towards increased ILD, especially 52
6. **Inflammatory arthritis/synovitis**: Morning stiffness, redness, swelling, multi-system involvement
7. **Dermatomyositis**: Violaceous rash, cracking (subset: antisynthetase syndrome → think of ILD: parenchymal lung involvement in up to 85%)
   - **Diagnostic Path**: Exam, history, lab, antibody testing (statin associated AB are very specific), EMG, **MRI**, muscle biopsy
8. **Ulcers**: MDA5 Dermatomyositis or malignancy associated dermatomyositis
9. **Diffuse ILD**: NSIP, UIP; architectural distortion (timeline at least weeks → inflammatory myopathy more likely)
10. **Normal CK**: Keep looking (myositis panel takes up to 2 weeks) → consider amyopathic myositis (ulcers: MDA5 - initial presentation is on a spectrum from outpatient to initial ICU care requirement)
11. **MDA5 myositis**: cutaneous findings (mechanics hands, ulcerations), often rapidly progressive ILD (RP-ILD) and respiratory failure; risk stratification predicts mortality in theses patients

Take-aways from Dr. Connors and Dr. Griffith:

- Patients with muscle and lung disease should be watched carefully!
- Infectious etiologies of myositis has to be rule out, but don't delay immunosuppressive therapy
- RP-ILD patients often end up needing lung transplantation, sometimes ECMO will be needed as a bridging therapy.