4/11/23 Morning Report with @CPSolvers

Case Presenter: *Debora (@deboracloureiro)* ***Case Discussants***: *Ravi (@rav7ks) and Ann Marie (@annkumfer)*

CC: Muscle weakness

HPI: 27 years old female presenting with facial edema that extended to the neck, erythematous plaques on her shoulders and muscle weakness.

Patient started having dysphagia, voice changes and dyspnea. Underwent aditional testing with EMG and PFT.

Improved after pulse therapy with methylprednisolone.

PMH: none

Meds: none

Fam Hx: none

Soc Hx: no alcohol, no smoking, no drugs

Allergies: none

Vitals: T: nl **HR:** nl **BP:** nl **RR:** nl **SpO2** nl
Exam:
Gen: nl
HEENT: nl
CV: RRR, no murmur
Pulm: CTAB
Abd: Soft, non-tender, non-distended
Neuro: Weakness of the proximal region of superior and inferior extremities. Pain in the shoulders and pelvis predominantly.
Extremities/skin: Gottron's sign.
Erythematous plaques lesions in the periorbital, peribucal, thorax and back.

Notable Labs & Imaging:
Hematology:
WBC: nl Hgb: nl Plt: nl

Chemistry:
Na: nl K: nl Cl: nl CO2: nl BUN: nl Cr: nl glucose: nl Ca: nl
Phos: nl Mag: nl
AST: 227 ALT: 114 CPK: 2136

PFT: Vital capacity, peak flow, mouth pressure were low for her.

Anti-RO +
Anti-JO1 -
EMG: Inflammatory myopathy

Final diagnosis: Dermatomyositis.

Problem Representation: 27 year old female with no known PMH presents with proximal muscle weakness, facial edema and Gottron's papules, found to have increased serum muscle enzymes (AST, ALT and CPK).

Teaching Points (Yaz):

Approach to weakness

Locate the weakness: Muscle vs. Nerve; Bilateral vs. Unilateral, Proximal vs. Distal and the time course.
Where is the issue? Think along the chain of the **Muscle contraction:** Brain sends a signal → nerve → neuromuscular junction → neurotransmitters + electrolyte needs for the depolarization/repolarization, as well as hormones.

Approach to edema

Facial edema: Think of the skin layers and vasculature, could this be a primary issue of the skin? Consider localized vs. systemic (angioedema vs. anasarca)
Remember that edema in endocrine disorders are accompanied by other S&S (Skin fragility, "Buffalo hump", etc.)
Plaques: Could be caused by photosensitivity, is there eye involvement? Where is it located? → dermatomyositis, SLE
Pain in shoulders and pelvic girdle In young px: First determine is it in muscles or nerves?, is it inflammatory pain?(morning stiffness and worse pain) Is there swelling?

- Antisynthetase Sx.→ Search for Lung crackles, diaphragmatic involvement (Pulmonary fibrosis is very frequent in these patients)
- Gottron Sign → Pathognomonic of Dermatomyositis.
- Dermatomyositis etiology → Idiopathic vs. Paraneoplastic, antibody-mediated vasculopathy, associated with malignancies such as Ovary CA, Lymphoma. Lung CA and Stomach CA
- ALT is more specific for the Liver, but is still expressed in muscle, so if you get a high muscle breakdown, you can get a high ALT:AST ratio

INFLAMMATORY MYOSITIS can have Lung involvement, which start with subtle manifestations, although may evolve to FIBROSIS → CT Scan → Look for signs of ILD (Initial screening and throughout PE is important i.e. assess the patient walking) **30% of px w/anti synthetase sx has a classic triad of ILD, myositis and arthritis (19% with full triad at dx.) ALSO see mechanics of the hands.**

Esophageal involvement → Upper ⅓ of the esophagus are skeletal muscle ∴ there can be dysphagia due to Antibodies against muscle (as well as the cricopharyngeal muscle) → the MOST COMMON manifestation of DERMATOMYOSITIS.