5/5/22 Morning Report with @CPSolvers

Case Presenter: *Sonia SIlinsky Krupnikova* ***Case Discussants:*** Sharmin Shekarchian *(@Sharminzi) and Rabih Geha(@rabihmgeha)*

CC: skin rash

HPI: 30yo who presented in outpatient setting with a rash on eye, forehead and anterior chest. Never had this before. Rash came on suddenly, over days. No new exposures, detergents. Was on vacation when first noticed. Rash has gotten progressively worse. Went to urgent care, told it was reaction from vacation exposure, was prescribed topical steroids which helped a little. Rash is scaly and itchy, not painful.

PMH: none

Meds: none

Fam Hx: none

Soc Hx:
Recent vacation

Health-Related Behaviors: alcohol 1-2x per week. 1 drink each time.
No tobacco use

Allergies: none

Vitals: stable, afebrile
Exam: **Gen**: well
CV, pulm, joint: wnl
Extremities/Skin: red to purple rash on eyelid and forehead on R side of face, extending to cheek. Rash also on anterior chest: red w/ scaling, excoriations, and telangiectasia around rash. Red rash also present on top of back and outer parts of shoulder.

Notable Labs & Imaging:
Course of illness: Given steroids, told to come back if did not improve. Weeks later, not improving w/ steroids. Told PCP that developed increasing dyspnea over last few weeks w/ nonproductive cough, keeping her up at night. No fever, chills, sweats. Also reported new rash on fingers that is painful and "opening up" w/ no Raynauds. Sent to rheum
Rhem repeat exam: Vitals stable, reporting SOB and cough.
Pulm: crackles b/l base. GI: wnl
Skin: new rash on hands (fingertips and palmar side, red, .5cm raised papules, starting to ulcerate w/ erythema. Back of hand: wnl. Face: violaceous, not erythematous on R>L eyelid. Chest and back: less scaling and excoriation
Joint: strength wnl, joint w/ no arthritis
Labs & Imaging
CBC and CMP: wnl
CXR: wnl
CT chest: GGO b/l bases, no LAD
Infectious work-up: IFN-gamma neg, neg hepatitis, aspergillus, histo, blasto
Dermatomyositis antibody: positive for MDA-5
ANA: Jo-1 neg, Scl-70-neg, dsDNA: neg
Treat outpatient w/ high dose steroids and Rituximab: improved

Final diagnosis: Anti-MDA5-amyopathic dermatomyositis

Problem Representation: 30yo woman w/ no significant PMH p/w progressive rash on cheeks, forehead and anterior chest with progressive dyspnea and finger/palmar ulcerations

Teaching Points (Samy):

- Rash: tip of the iceberg (limited to the skin or caused by a systemic illness)? Consider the possibility of the rash being caused by excessive pruritus? -> DDx of pruritus
- Outside job (contact dermatitis, superficial infections, cutaneous mycoses) vs. intern job (signs of visceral, more systemic involvement, e.g. infections, autoimmune diseases, hematologic malignancies)
- Pruritic rash: dry skin, scabies, medication side effect, urticaria, atopic dermatitis, dermatitis herpetiformis duhring
- Purple orbits? Type 1 thinking: heliotrope rash seen in dermatomyositis, typical involvement of the nasolabial folds, gottron's papules, mechanic's hands, shawl sign, V-sign
- Dermatomyositis:affects the skin, muscle, lung (ILD) and vasculature (raynauds, pulmonary HT)
- How to differentiate mechanic's hands in dermatomyositis from trauma induced lesions on the fingers (true mechanics): involvement of the lateral sides of the fingers in DM
- ILD usually a subacute/chronic disease, but certain forms can also lead to rapid progressive pulmonary failure (e.g. anti-MDA5-dermatomyositis, usually associated with amyopathic dermatomyositis) Can have normal CK and aldolase! Less likely to be associated with cancer.
- Palm ulcerations violate rule of ulcers (usually affect lower extremities,, associated with vascular or neurologic disease) -> consider vasculitis, cutaneous cancer and endocarditis
- Lung+Skin: infectious (TB, endemic mycosis, nocardia), autoimmune (notoriously anti-Jo1 and MDA5-antibodies)
- Dermatomyositis usually is not itchy and is bilateral! Don´t exclude a diagnosis due to single findings that don´t fit well!