

Diagnosis and Treatment of a Patient with Refractory Scleromyositis and Eosinophilic Granulomatosis with Polyangiitis

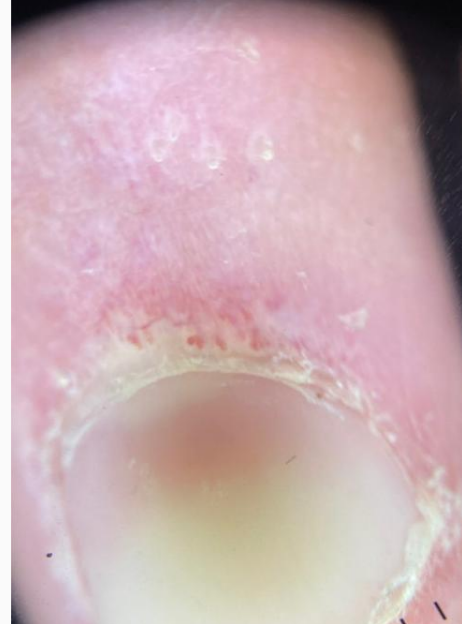
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Our Patient

32 year old male hx of asthma, eczema, and chronic rhinosinusitis with nasal polyps who presented with new onset rash along with fatigue, arthralgias, and muscle weakness



Diagnostic Workup

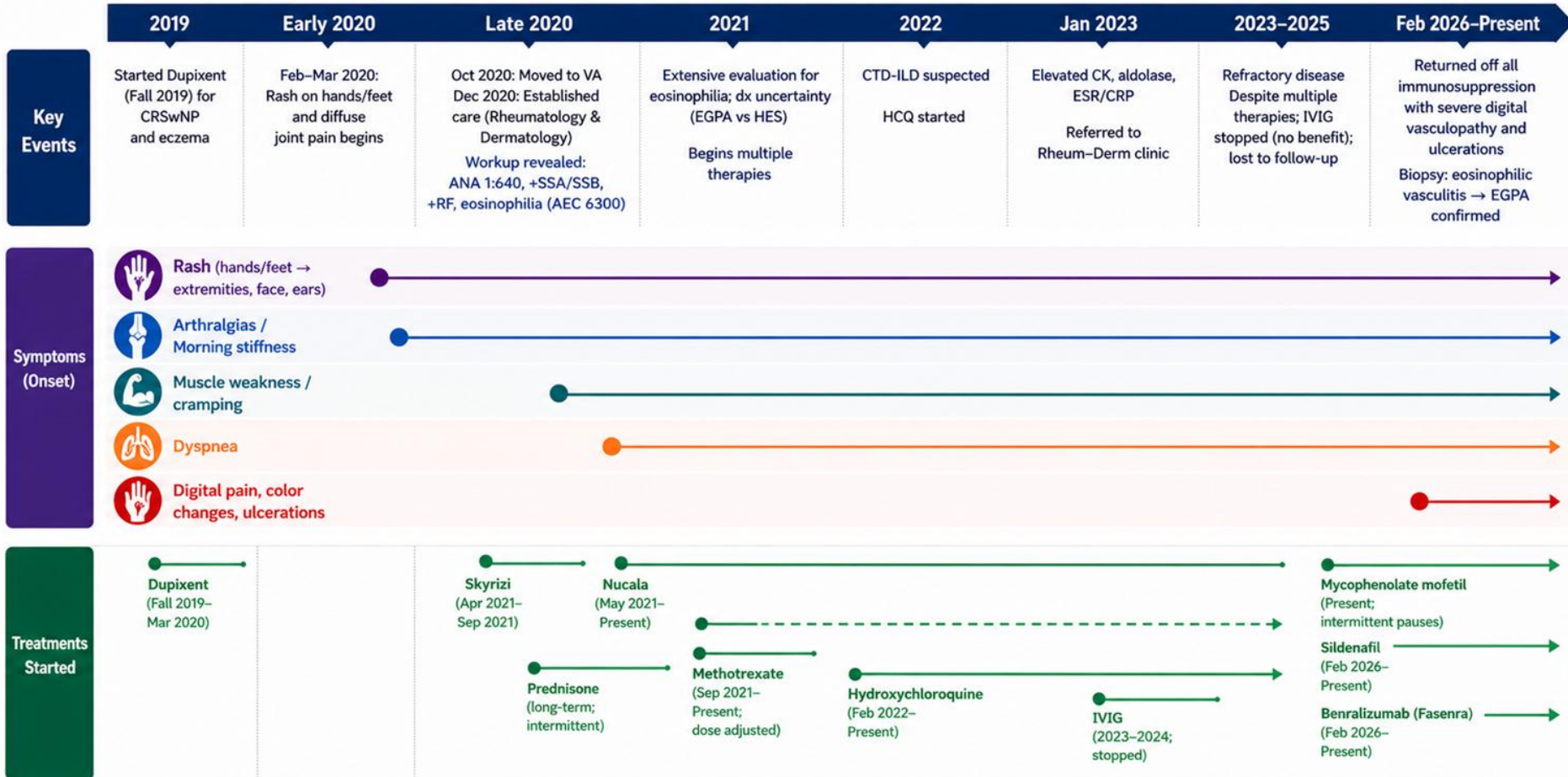
- Labs:
 - ANA 1:640, +SSA, +SSB, +RF, +PM/Scl, +Ro60
 - Marked peripheral eosinophilia 6300
- Imaging:
 - MRI thighs: extensive bilateral fasciitis
 - CT chest: ground glass opacities concerning for possible CTD-ILD
- Biopsies:
 - Fascial biopsy: chronic inflammatory fasciitis consistent with dermatomyositis
 - Skin biopsy: mixed pattern dermatitis with psoriasiform and interface changes
 - Bone marrow: benign
 - Salivary gland: benign

Initial diagnostic workup consistent with scleromyositis and questionable Eosinophilic Granulomatosis with Polyangiitis vs Hypereosinophilic Syndrome

Physical Exam



Clinical Course: Symptom Onset & Treatments Over Time



Treatment History and Current Therapy

- Multiple therapies tried with incomplete response including prednisone, methotrexate, Mepolizumab, hydroxychloroquine, mycophenolate, IVIG
- Patient re-presented to clinic after ~2 years; started on prednisone, mycophenolate, and sildenafil with mild improvement
- Patient currently on prednisone, mycophenolate, sildenafil, and recently started Benralizumab (IL-5 receptor inhibitor)

Most Recent Follow Up

- Patient currently improving on mycophenolate, Benralizumab, prednisone 10 mg, and sildenafil
- Notably, Botox injection at the base of ulcerated digits expedited healing and offered significant pain relief

Take Home Points

- It's important to consider overlapping diagnoses when clinical features do not fit a single diagnosis
- Treat patients holistically. An important part of our job as a physician is empowering patients to navigate the healthcare system
- It can take months to years to achieve a final diagnosis, it's important to follow these patients closely and earn their trust

Questions?

References

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