A long story of evolving skin lesions

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Conflicts of interest

None

Our patient

- Currently a 73 year old female who has hypertension, vitiligo and hypothyroidism
- December 2019 she <u>developed a swelling over the spine</u> that was tender but more of a lump a "hotdog"
- Sept 2020 she developed <u>erythema and violaceous discoloration of</u> the dorsum of both feet
- Except for paresthesia in the feet, she has no other symptoms







Labs in 2021

- ESR 50 and CRP 15 mg/L
- ANA +1:320 with neg ENA
- SPEP normal
- ANCA, C3, C4, CK, Cryos, CCP, RF, Hep A, B and C, QuantiFERON, ACE
- CT CAP negative
- MRI of thoracic spine showed no bony abnormalities

Thoughts?

She is sent to our dermatology/rheumatology clinic

- 2 biopsies done at DH prior to her derm/rheum visit (6/23/2021) showed
 - Inflammatory/sclerosing disease with a dermal interstitial cellularity resembling an interstitial granulomatous process like GA or necrobiosis lipoidica
 - Seen in derm/rheum clinic in July 2021





Her exam also showed olecranon nodules



Thoughts now?

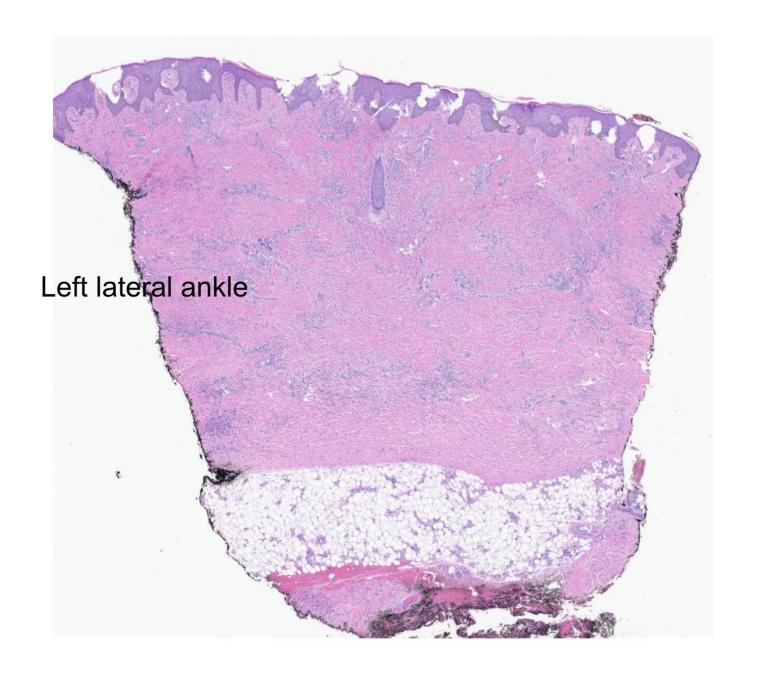
The derm/rheum thoughts

- Morphea or scleroderma
- Lipodermatosclerosis
- Fasciitis

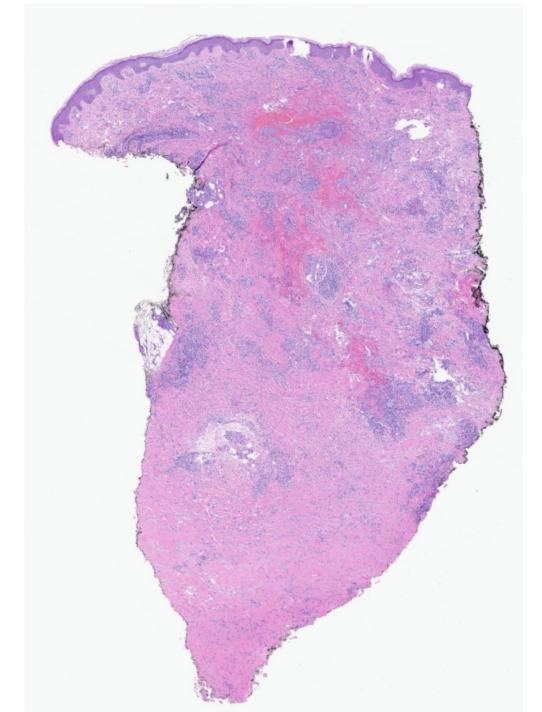
But what about the nodules?

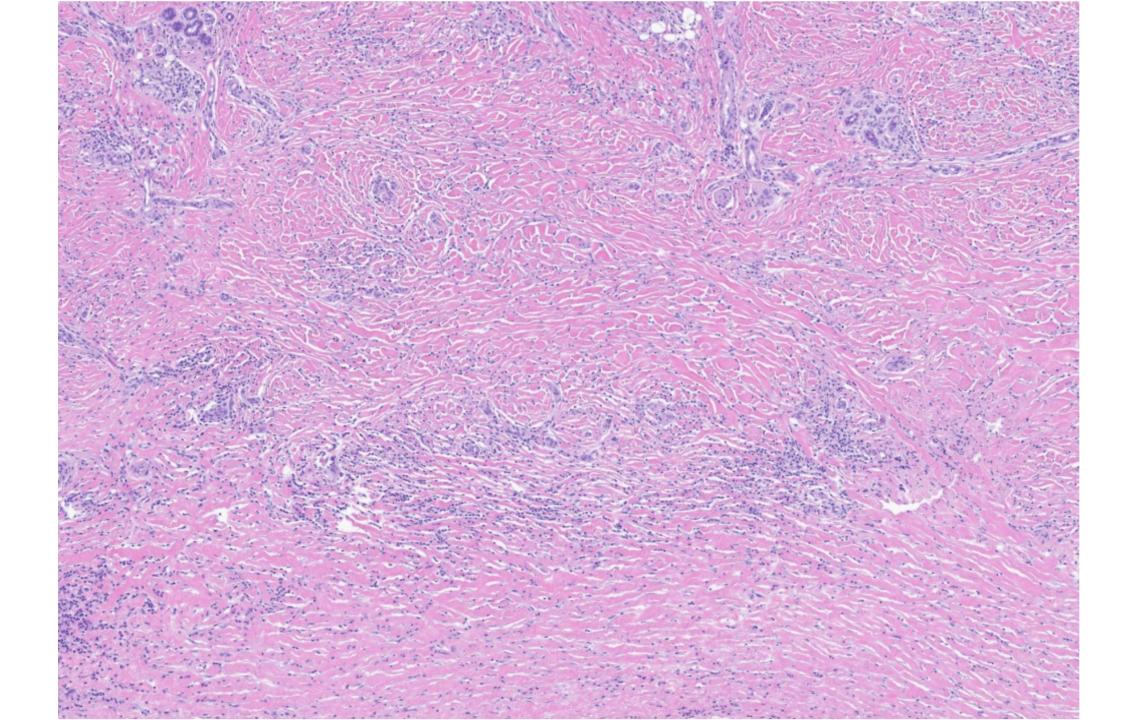
Ultimately had deep biopsy of the leg and of the nodules (11/2022!)

- No fasciitis
- Same inflammatory infiltrate and collagen changes as prior biopsy
- Both lesions same pathologic change



Right elbow





So a new diagnosis was given

- Diagnosis favored by pathology was <u>morphea profunda/nodular</u> <u>morphea</u> – so in July 2023 methotrexate recommended
- Patient thought about it and was seen again in 5/7/2024 where her symptoms were essentially unchanged

A diagnostic test was performed in 2024

A Lyme test

Acrodermatitis chronica atropicans (ACA)

- Treated with doxycycline for 28 days with improvement in the rash and in her nodules
- Second course of doxycycline resulted in further improvement
- No swelling
- Improved mobility
- Still paresthesia













ACA

- Late Lyme manifestation
- May be sole feature of Lyme disease or follow prior symptomatic infection
- Does not resolve spontaneously (like ECM)
- Occurs on extremities; nodules described infrequently (2-3%)
- Begins with edematous erythema and violaceous discoloration
- May progress to atrophy
- Typically not diagnosed for a year or more

• Often diagnosed as vascular insufficiency, venous insufficiency, superficial thrombophlebitis, hypostatic eczema, arterial obliterative disease, acrocyanosis, livedo reticularis, lymphedema, or chilblains

- Older women
- Felt to not occur in US, but there are scattered reports and Borrelia burgdorferi sensu stricto is found in European patients; B afzelli predominates
- Definition includes the right clinical appearance, +Lyme antibody test and biopsy showing compatible inflammatory changes*
- Spirochettes demonstrated in ACA in a higher number of cases than ECM

If at first you don't succeed try, try again!

Thank you

 And thanks to my dermatology colleague at DHMC, Dorothea Barton, MD, plus Rob LeBlanc, MD pathology, and Shaofeng Yan, MD pathology, as well as the patient

References

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