A Woman With Progressive Cutaneous Ulcers

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Disclosures

- No relevant conflicts of interest

Case Presentation

- An 88-year-old woman presents for the evaluation of progressive lower extremity ulcers.

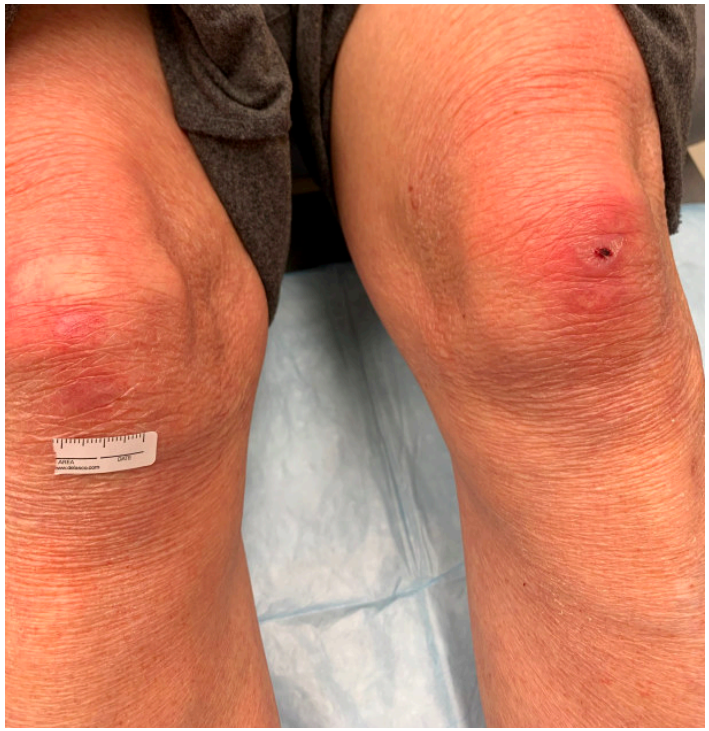
Initial Presentation

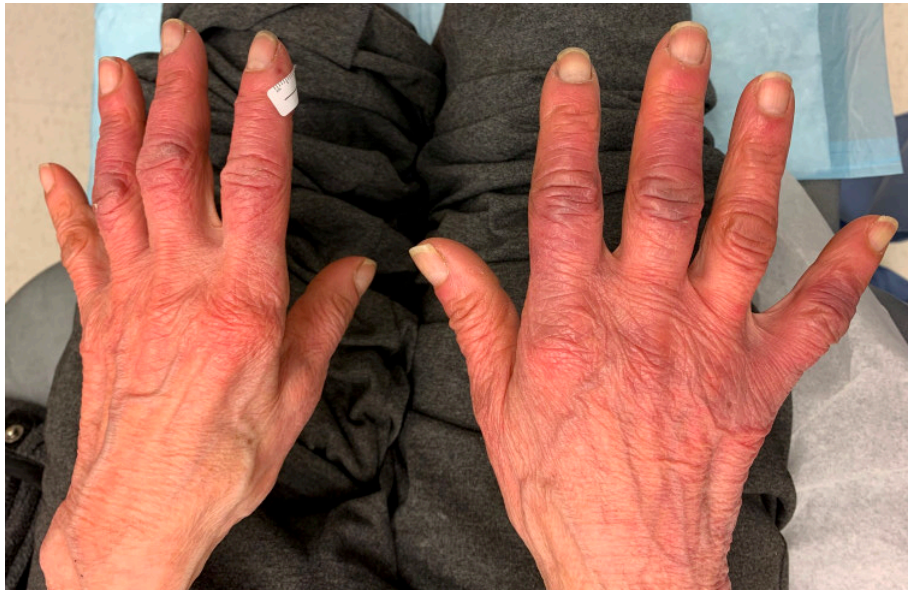
- **February 2019:** Erythema of the hands and feet
- **August 2019:** Dermatology evaluation
  - Well-demarcated bright, blanching erythema of the soles and dorsal hands.
  - Fingers unaffected, nailfold capillaries unremarkable.

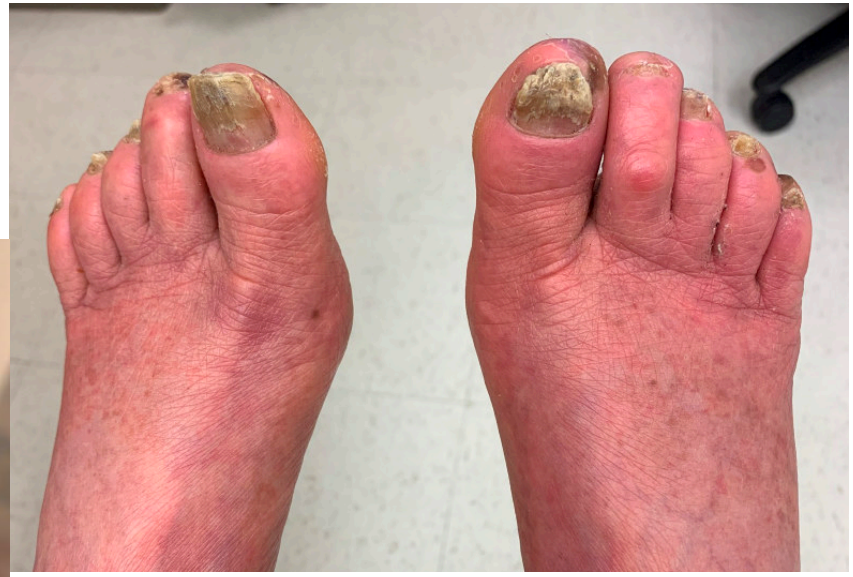
Initial Presentation

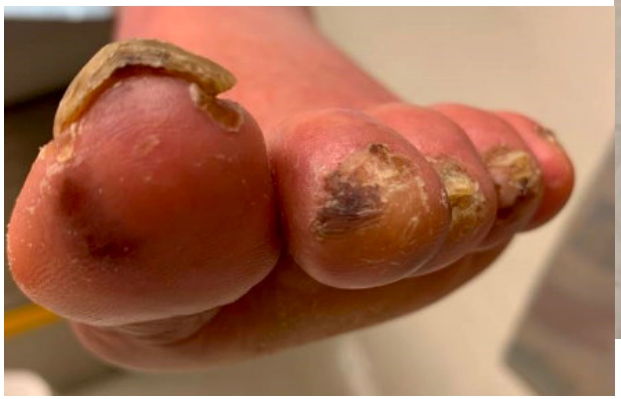
- **February 2020:**
  - Puffiness of the hands and feet
  - Purpuric lesions on fingers and toes
    - Exacerbated by cold
  - Bilateral knees: erythematous to violaceous patches anteriorly
    - Left knee: 5 mm ulcer

February 2020









Past Medical History

- Monoclonal gammopathy of undetermined significance (MGUS)
- Mild chronic ITP
- Osteoporosis
- Atrial fibrillation
- Hypertension
- GERD with Barrett's esophagus
- Glaucoma
- Leukocytoclastic vasculitis
  - 2016, resolved with topical triamcinolone x 1 month
  - ANA 1:160, homogenous; SSA/SSB negative

Medications

Apixaban

Calcium citrate

Cholecalciferol

Denosumab

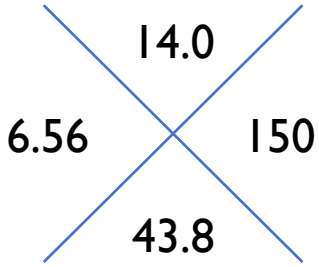
Docusate

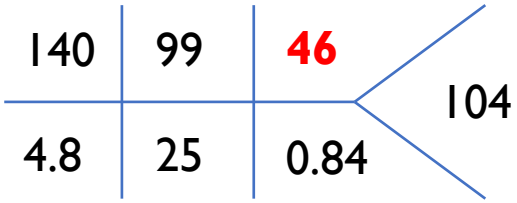
Latanoprost eye drops

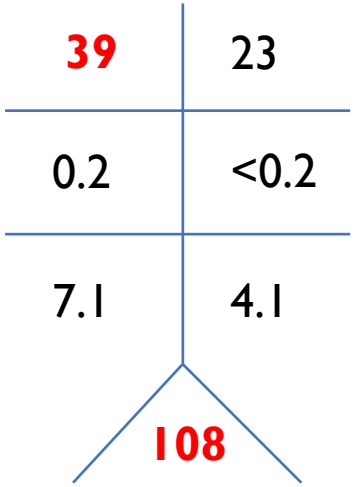
Lisinopril

Metoprolol tartrate

March 2020 Labs







LDH 225 U/L
ESR 11 mm/hr
CRP 1.5 mg/L

SPEP: IgG 1204, IgA 144, **IgM 47 mg/dL (↓)**
- **M-spikes: 0.05-0.5 g/dL IgG kappa**

Free light chains (mg/L): **Kappa 84.6 (↑)**, lambda 13.0
- **K/L ratio: 6.51**

ANA 1:2560
- **Homogenous, oligodot (0-3)**

dsDNA 1:320
ANCA, RF, cryoglobulins neg.
Beta-2 glycoprotein, cardiolipin neg.
Lupus anticoagulant neg.
C3, C4 WNL
UA: no proteinuria or hematuria

Rheumatology Evaluation

- **April 2020 rheumatology evaluation**
  - Longstanding dry eyes and dry mouth
  - Intermittent hand joint pain with activity
    - Denied morning stiffness
  - Denied Raynaud phenomenon, mucosal ulcers, dysphagia, alopecia, pleurisy, constitutional symptoms
  - No family history of autoimmune disease

Rheumatology Evaluation

May 2020 Labs

ANA >1:5120

- **Homogenous**

dsDNA 1:5120

Ro, La, Smith, U1RNP neg.

ANCA, RF neg.

Beta-2 glycoprotein, cardiolipin neg.

Lupus anticoagulant neg.

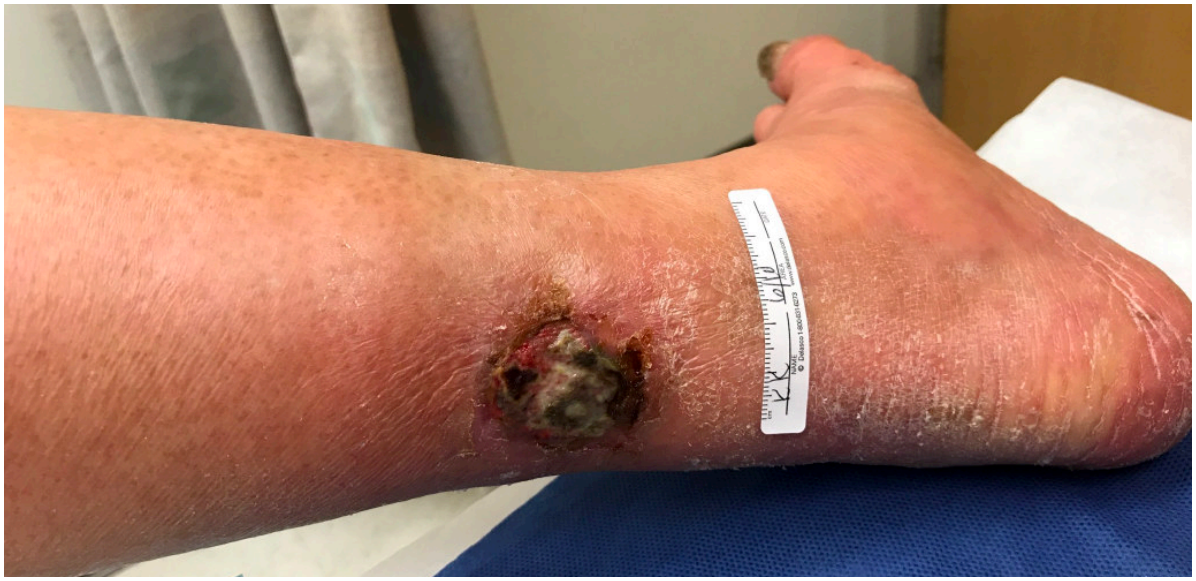
C3, C4 WNL

UA: no proteinuria or hematuria

Initial Presentation

- **June 2020**
  - Fingertip and toe lesions beginning to heal
  - Progressive left posterior calf ulceration, L>R lower extremity edema

June 2020



History of Present Illness

- **June 2020**
  - Punch biopsy of left posterior calf:

Spongiosis, serum-imbued parakeratosis, extensive dermal fibrosis and a lobular vascular proliferation with acute and chronic inflammation, extravasated erythrocytes, and hemosiderin deposition.

There is multifocal perivascular fibrin in the superficial dermis. Vasculitis is not seen. No micro-organisms are detected with special stains (Brown Hopps and PASD). Von Kossa stain does not show significant calcium deposition, and elastic tissue stain shows preservation of vascular elastic lamina.

Overall, the findings are most consistent with stasis dermatitis.

Initial Presentation

- **June 2020**
  - Left posterior calf lesion felt to be consistent with stasis dermatitis
    - Treated with compression, elevation, and wound care
  - All other lesions resolved spontaneously
- **August 2020**
  - dsDNA titer decreased to 1:320

History of Present Illness

- **September 2020**
  - Recurrent mild acral erythematous patches
  - Left lower extremity ulcer healing slowly
- **November 2020**
  - Recurrent puffiness of the hands and feet

History of Present Illness

- **December 2020**
  - Left lower extremity ulcer enlarging
  - New rapidly worsening nodules and ulcers throughout bilateral legs
  - Presented to the hospital for rapid worsening of symptoms

Physical Examination

Gen: Thin, no apparent distress

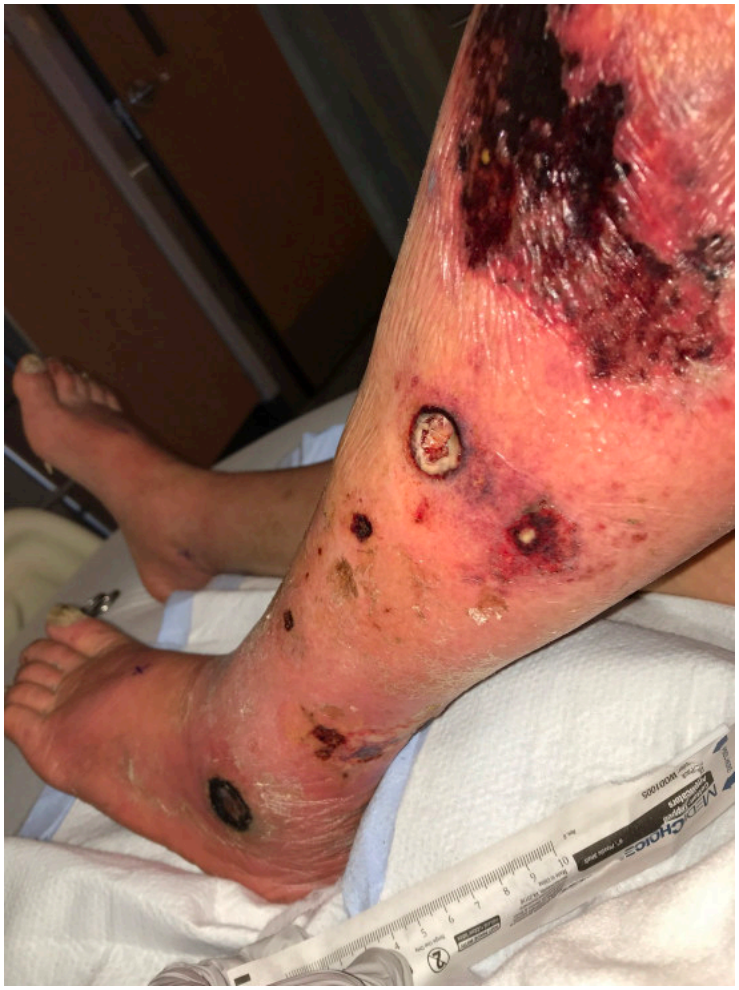
Ext: L>R 3+ pitting edema

MSK:

- Accentuated thoracic kyphosis
- Puffy fingers without synovitis

Skin:

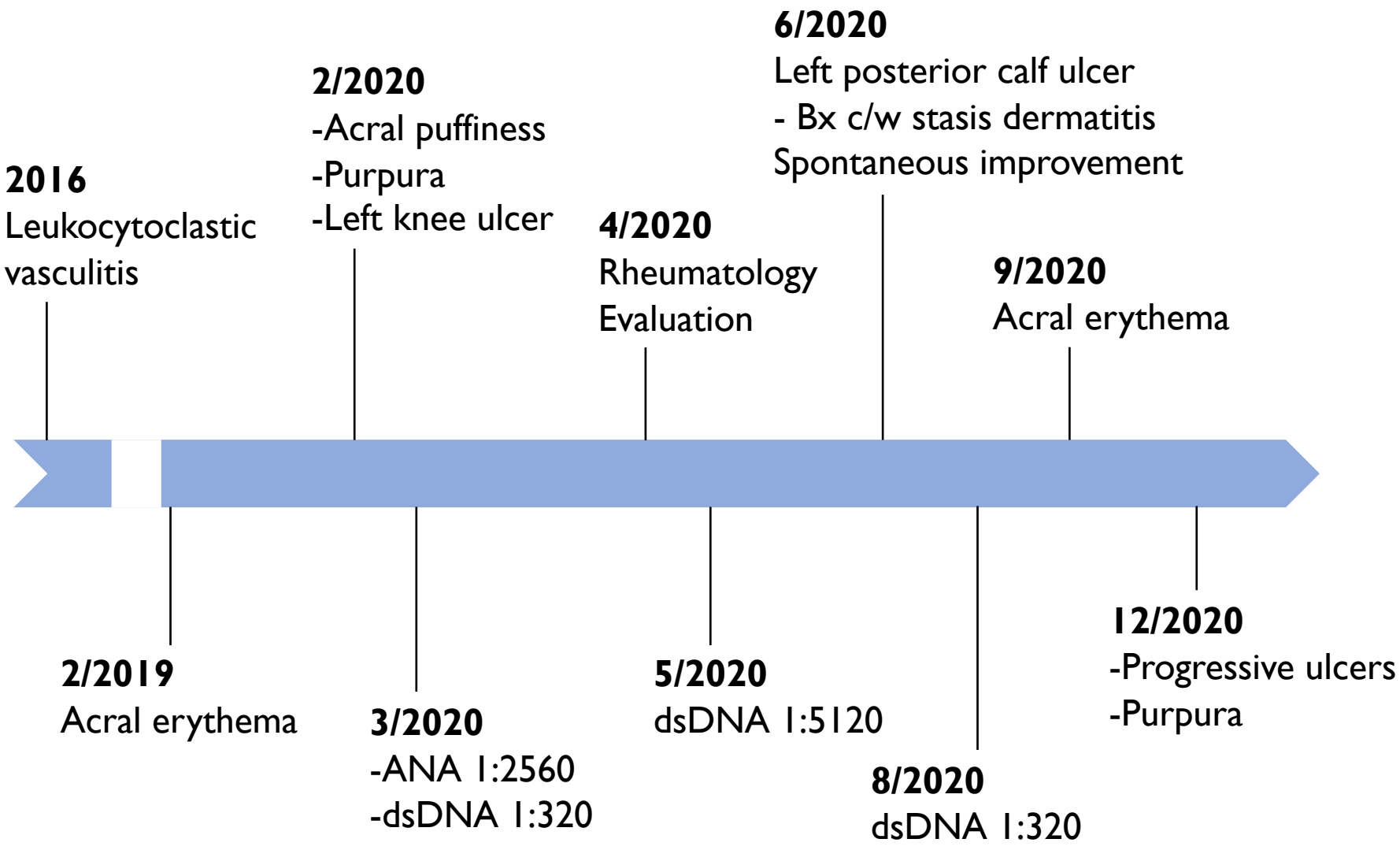
- Numerous tender erythematous/violaceous nodules and ulcers in lower extremities
- Erythematous/violaceous discoloration of fingers
- Purpura on multiple distal fingers and toes



Laboratory Evaluation

- **ANA 1:5120, homogenous and speckled**
- **dsDNA 1:320**
- **Platelets 126** (baseline: 110-150)
- IFN-gamma release assay negative
- Otherwise unchanged from prior

Timeline of Events



Skin Biopsy #2

Subepidermal split overlying thick-walled vessels with hyalinization/fibrin deposition, extravasated erythrocytes and a perivascular and interstitial lymphohistiocytic infiltrate with neutrophis associated with a mixed septal and lobular panniculitis.

The findings are non-specific, but **compatible with the clinical impression of a vasculitis or vasculopathy**.

No microorganisms are detected with a Brown Hopps, PASD, GMS, AFB, and Fite stain.

Summary

- 88-year-old woman presents with acral erythema, purpura, and lower extremity nodules and patches
- Initial spontaneous improvement followed by rapid development of lower extremity ulcers
- Labs notable for high-titer positive ANA and dsDNA
- Biopsy consistent with septal and lobular panniculitis and vasculitis or vasculopathy

Differential Diagnosis

Autoimmune

Lupus profundus

Polyarteritis nodosa

Cryoglobulinemia

Erythema nodosum

Infectious

Erythema induratum

Fungal, mycobacterial infection

Neoplastic

Cutaneous lymphoma

Monoclonal gammopathy-associated vasculopathy

Other

Lipodermatosclerosis

Hypercoagulable vasculopathy

Skin Biopsy #2, continued

Mycobacterial culture: ***Mycobacterium chelonae***

Course

- Started on azithromyin and imipenem then tobramycin
- Narrowed to azithromycin and doxycycline based on susceptibilities
- All cutaneous lesions resolved over the next few months
- Remains on azithromycin and doxycycline until 10/2021

Mycobacterium chelonae

- Nontuberculous mycobacterium (NTM)
- Ubiquitous in freshwater, soils, plant material, tap water
- Infection typically through cutaneous inoculation
- Typically presents with skin/soft tissue infection in area of inoculation
- Highly drug-resistant
- Frequently affects immunocompetent hosts

Yu E et al. *J Foot Ankle Surg*. 2020 Sep-Oct;59(5):1084-1091.

Erythema Induratum

- AKA nodular vasculitis, erythema induratum of Bazin
- Hypersensitivity reaction, usually to *Mycobacterium tuberculosis* infection
  - Has been reported with other mycobacteria, including *M. chelonae*, *Nocardia*, *Pseudomonas*, *Fusarium*, *HCV*
  - Also reported with
- Presents with recurrent erythematous/violaceous plaques or nodules, often with ulceration
- Skin biopsy: septolobular panniculitis, granulomatous inflammation, neutrophilic vasculitis, necrosis

Campbell SM et al. *J Clin Aesthet Dermatol*. 2013 May;6(5):38-40.

Gilchrist H, Patterson JW. *Dermatol Ther*. 2010 Jul-Aug;23(4):320-7.

Magalhães TS, et al. *J Dermatol*. 2018 May;45(5):628-629.

Erythema Induratum

- Diagnosis
  - Pulmonary or other evidence of systemic tuberculosis
  - PPD, IGRA not universally positive
  - Skin biopsy mycobacterial cultures often negative
  - Often requires empiric course of anti-TB regimen
- Treatment
  - Treat underlying cause
  - Usually 9-month anti-TB regimen

Campbell SM et al. *J Clin Aesthet Dermatol*. 2013 May;6(5):38-40.
Gilchrist H, Patterson JW. *Dermatol Ther*. 2010 Jul-Aug;23(4):320-7.
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Final Diagnosis

Erythema induratum secondary to *Mycobacterium chelonae*

Thank you

Questions?