To Swell and to Stay

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Case Presentation

This is a 27-year-old Hispanic male with PMH overlap seropositive rheumatoid arthritis and systemic sclerosis who presented to the hospital with a worsening left inguinal mass since Sept 2022 and fever, now status post I&D. Rheumatology is consulted for the management of immunosuppression in the setting of infection.

Current Immunosuppressant regimen on EMR: Prednisone 5mg, Leflunomide 20mg daily

History of Rheumatic Disease:

- Onset March 2021, Diagnosis Nov 2021 when he moved to US from Guatemala
- Rheumatoid Arthritis: (+) RF 11, (-) CCP
- Systemic Sclerosis: ANA 1:1280, nucleolar, Scl70 neg, PM/Scl75 neg, RNApol3 neg
- Manifestation: inflammatory arthritis (left knee and left ankle), diffuse cutaneous systemic sclerosis, Raynaud's with ulceration, tendon friction rubs, abnormal nail fold capillaries

History Continued

Patient: "I stopped the prednisone and Leflunomide three weeks ago. I went to urgent care for the mass and they asked me to stop. It wasn't helping my joints anyway; my knee always stayed swollen. The steroid injection in the knee also didn't help."

- Medication History:
  - Methotrexate- stopped d/t alopecia, transaminitis
  - Tocilizumab- stopped d/t inefficacy
  - Leflunomide- Stopped d/t suspected groin mass/infection

Physical Exam

- Gen: NAD
- HEENT: Sclera anicteric, no conjunctival injection, MMM, no oral ulcers
- Cardiac: RRR, no murmurs auscultated
- Pulm: CTAB, speaks in full sentences, normal respiratory effort
- Integumentary**: (+)Diffuse thickening of extremities distal to elbows (forearms, dorsum of hands, and fingers), distal thighs and below knee. (+) Digital tip pit- left third** No active raynaud's. **(+)Telangiectasias over face, along with patchy hypo and hyperpigmented rash; (+) Left groin scar post I+D: C/D/I. (+) area of superficial induration posterior thigh (different than skin thickening)**
- Neuro: Alert and oriented, moves all 4 extremities
- MSK: **(+) Left knee with joint swelling, no warmth or overlying erythema.** No ankle swelling restriction in ROM. Remaining joints without signs of synovitis (tenderness/swelling), deformity, or restriction in ROM

Objective History

- Vitals:
  - BP 110/77, HR 129, RR 24, Temp 101.1, SpO2 100%
- Labs:
  - ESR 118, CRP 11.7
  - CBC: WBC: 10.7, Hgb 11.9, Plt 361
  - BMP: Electrolytes wnl, Cr. .6
  - HIV, RPR, Urine Gonorrhea/Chlamydia neg
- XR Left Knee: No acute fracture or dislocation. Maintained joint spaces and alignment. Small effusion. Soft tissue appears unremarkable.

Imaging

- CXR: wnl
- CT Left Hip w/ contrast:

1. Inflammatory process extends from the caudal aspect of the left psoas muscle through the left iliacus muscle and into the soft tissues along the ventral aspect of the left thigh, contiguous with and possibly involving the proximal left quadriceps. There are multiple associated irregular-shaped pockets of encapsulated fluid along the left inguinal soft tissues concerning for multiple small abscess collections.
2. Asymmetrically prominent enlarged left inguinal lymph nodes with lymphadenopathy extending along the left pelvic sidewall.
3. Fluid attenuation encapsulated focus along the proximal left sigmoid colon measuring up to 2.1 x 8.9 cm in size concern for small abscess. This may be secondary to focal diverticulitis versus a bystander effect from the contiguous inflamed lymphadenopathy.

I+D Left Groin Mass

- Final Diagnosis
  "Left groin" (Specimens #1-2); excisions with intraoperative consultation:
  - Caseating and non-caseating granulomatous inflammation.
  - Permanent sections and histochemical stains reveal forms suggestive of degenerating fungal forms.

- Inguinal flow cytometry:
  Lymph nodes, left inguinal, flow cytometric analysis:
  - No evidence of increased blasts or clonal T or B cell populations (see Comment).

Next steps for primary team

- Caseating granulomas??
  - ID Consult: CXR, TB Quant, AFB x3 Neg
- Malignancy??
  - CT A/P:

  1. Redemonstrated left inguinal predominant multiloculated fluid collection with rim enhancement and adjacent fat stranding extending from the top of the ileum to the left lower extremity. No drainable fluid collection. Ongoing bilateral left external iliac adenopathy, centered around the left external vessels which shows some overall decrease in size from imaging dated 5 April 2023.
  2. No definite evidence of intraperitoneal active process.
  3. Significant pericardial effusion without evidence of tamponade physiology. Consider echocardiography.
- Now there's a pericardial effusion??
  - Mild tachycardia, other wise asymptomatic→ ECHO→ Pericardiocentesis

What additional history/work-up would you like?

- Micro:
  - Blood culture: No growth
  - Left groin biopsy: Coccidioidomycosis
  - Pericardial fluid: No growth

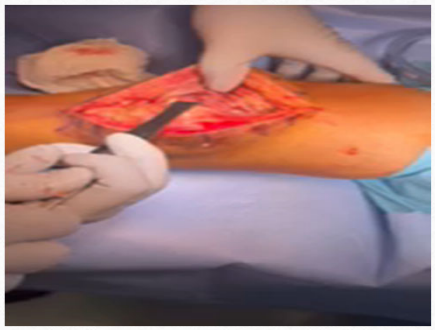
The knee is still swollen…

- We're rheumatologists, we like to tap joints:
  - Left knee synovial fluid analysis: 4583 WBC, 11200 RBCs, no crystals. Culture negative.
- Thoughts on the differential? What's the next step in the work-up?

Things are not adding up…

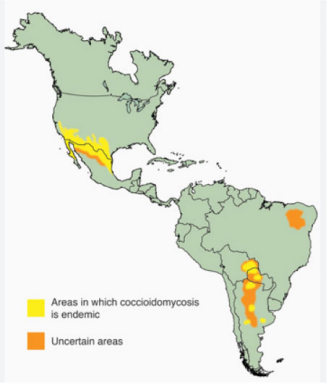
- Patient with history of seropositive RA, however RF low positive at 11
- Two years of persistent knee swelling refractory to multiple DMARDs
- What would you ask for next in the work-up? Why?

Left Knee Arthroscopy



Learning Points

- Disseminated coccidioidomycosis can masquerade as an inflammatory arthritis "Desert rheumatism"
- Coccidioidomycosis can present as an indolent infection; it is important to take a complete travel history
- Clinicians should be cautious of association bias when diagnosing a patient with an established autoimmune disease



References

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