Think Horses: an SLE patient with Necrotizing Lymphadenitis

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Discolsure

- Nothing to disclose

Chief Complaint and HPI

- 24 Year old previously healthy woman presenting from OSH
- Fatigue
- Weakness
- Neck swelling
- Unintentional weight loss of 20 pounds
- Hand sores
- Gradual over 5 weeks

HPI continued

- PMH: TMJ dysfunction
- PSH: none
- No known exposures
- Family HX: no cancers or autoimmune disorders
- Meds: NSAIDS prn
- Social history
  - House keeper at local hotel
  - Denied ETOH, tobacco or ilicit drug use
  - Sexually active with boyfriend, used condoms

Brief OSH Course

- Transferred to academic medical center after 1 week
- Cervical lymph node biopsy
  - CD15+, CD30- cells
  - Necrotizing lymphadenitis
- Corticosteroid treatment initiated
  - D/C due to lack of clear etiology

Vitals

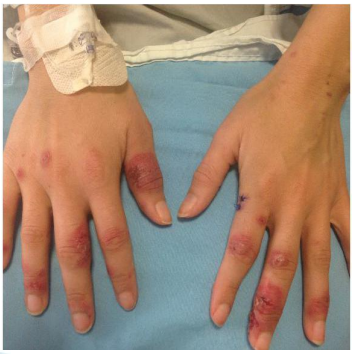
- BP: 115/64 mmHg
- Temperature: 100 F (spiking to 103 F)
- Pulse: 68 bpm
- RR 18 bpm
- SpO2 96%

Pertinent Physical Exam Findings

- General: uncomfortable but no acute distress
- Lymph: posterior cervical lymphadenopathy, parotid swelling
- GI: absent hepatosplenomegaly
- Skin: numerous erythematous, non-painful/tender plaques and pustules on hands

Cervical Lymphadenopathy

Hand Lesions



Differential Diagnosis?

Differential Diagnosis?

- Kikuchi Disease
- Sarcoidosis
- Sjögren's syndrome
- CREST Syndrome
- Mixed connective tissue disease
- Tuberculosis
- Lymphoma
- Hepatitis C
- Scleroderma

Labs: hematology and infectious

- Hb: 8.5 g/dl
- WBC: 2.5 x 10^3/mm^3
  - ANC: 1.62 x 10^3/mm^3 (L)
  - ALC: 0.63 x 10^3/mm^3 (L)
- PLT: 266
- BCX, HIV, mumps, EBV, CMV: negative
- HCV AB: positive

Labs: chemistry and immunological

- K: 3.3 mmol/l
- ALB: 3.2 g/dl
- AST: 41 U/L
- ALT: 35 U/L
- ANA: positive (index 2.28)
- SCL 70: positive
- Anti dsDNA AB: negative
- ESR 108
- Normal complement levels

Imaging

- CT of neck: hyperplasia of bilateral parotid glands and bilateral LAD cervical nodes
- CT of Abdomen: benign, no HSM

Diagnostic studies

- Punch biopsy of skin lesions
  - Immune complex deposition
- Bone marrow biopsy
  - Decreased hematopoiesis
  - No significant dysplasia
- Review of outside slides
  - Negative for infectious etiology

Why we thought she had a "zebra"

- Necrotizing lymphadenitis
  - Rare in SLE (Shrestha et al)
  - But not impossible (Wallace et al)
- The negative dsDNA antibodies
  - Specific, not sensitive for SLE (Isenberg et al)
- Status of HCV
  - Infection vs. inflammatory "red herring"
- CD15+ cells
  - Malignancy concern

Kikuchi Disease (Khanna et al)

- Rare, benign necrotizing lymphadenitis
- Idiopathic
  - Speculation autoimmune
- Young, Asian women
- Symptoms overlap with SLE
  - Fever
  - Weight loss
- Generally self resolving

Why She had SLE

- Results of skin biopsy
  - Classic for Type III autoimmune disease
- Clinical diagnosis
  - Nasal ulcers
  - ANA positive
  - Evidence of chronic inflammatory state
  - Rash

Hospital Course

- Diagnosed with SLE
- Initiated on corticosteroids and hydroxychlorquine
  - Improvement of hand lesions
  - Subjective improvement in symptoms
- Following with rheumatology clinic

SLE diagnostic criteria

- Serositis
- Oral (nasal) ulcers
- Arthritis
- Photosensitive rash
- Blood dyscrasia
- Renal abnormalities
- ANA positivity
- Immunological phenomenon
- Neurological symptoms
- Malar rash
- Discoid lesions
- American College of Rheumatology
  - 4 of 11 required
  - "SOAP BRAIN MD"

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