When excluding all other diagnoses leaves you with three

A Case of Sarcoidosis-Lymphoma Syndrome with Idiopathic Thrombocytopenic Purpura

Clark Cutrer, M.D.
Introduction: Sarcoidosis

- Noncaseating granulomas
- Other characteristic findings: erythema nodosum, lymphopenia, hypergammaglobulinemia, hypercalcemia, and elevated ACE
- Highly variable clinical course
- Diagnosis of exclusion
- Mainstay of therapy: steroids
Idiopathic Thrombocytopenic Purpura

- Autoantibodies against platelet antigens
- Diagnosis of exclusion
- Bone marrow:
  - Normal cellularity w/ megakaryocyte hyperplasia
- Association between autoimmune disorders and lymphoproliferative disease
Hodgkin’s Lymphoma

- Fevers, chills, night sweats, weight loss
- Mediastinal lymphadenopathy common
- Reed-Sternberg cells
- CD30, CD 15 +
Timeline of events

- **October 2015**: Onset of symptoms
- **February 2016**: 1st LN biopsy negative
- **March 2016**: Onset of B symptoms: dx: ITP, sarcoidosis; 2nd LN biopsy/BMB neg
- **May 2016**: Worse B symptoms, Rheum consulted; long-term steroids
- **May-July 2016**: Rheum, Heme, Pulm, GI f/u; steroid taper; ongoing concern for lymphoma
- **July 2016**: B symptoms + N/V/D; C diff + (x2); CT CAP: worse LAD; final LN biopsy
Our Case: 1st Encounter

- 23 year-old African American male presented with groin/abdominal pain and nausea/vomiting for 5 months
  - Also c/o fevers, chills, night sweats, 14lb weight loss, and decreased appetite for 1 month
- Physical exam:
  - Shivering, thin young male
  - Tender in RLQ and right inguinal area
  - Right inguinal fullness but no distinct/well-defined lymph node
Initial Work-up

- Severe thrombocytopenia and diffuse LAD
- Lymph node biopsy: noncaseating granulomas
- Bone marrow biopsy: megakaryocytic hyperplasia
What did we do for him?

- **Diagnosis:** sarcoidosis and ITP
- **Treatment:** oral steroids, IVIG, and romiplostim with improvement in symptoms and thrombocytopenia
Timeline of events

October 2015
- Onset of symptoms

February 2016
- 1st LN biopsy negative

March 2016
- Onset of B symptoms: dx: ITP, sarcoidosis; 2nd LN biopsy/BMB neg
- May 2016
- Worse B symptoms, Rheum consulted; long-term steroids

May-July 2016
- May 2016
- May-July 2016

July 2016
- Early
- July 2016
- Late
- July 2016
- CT CAP: worse LAD; final LN biopsy
- B symptoms + N/V/D; C diff + (x2)
- Rheum, Heme, Pulm, GI f/u; steroid taper; ongoing concern for lymphoma
Our Case: 2\textsuperscript{nd} Encounter

- Interim: Follow-up with Rheumatology, Pulmonology; started on long-term steroids; negative LN biopsy x2
- Presented 3 months later with dyspnea on exertion, nausea, vomiting, and diarrhea
- Also noted 30lb weight loss and worsening fevers, chills, and night sweats
Additional Work-up and Management

- **Remarkable Lab Work:**
  - WBC 7.1, H/H 9.6/30.4, MCV 80, Platelets 380
  - AG 19, Albumin 3
  - LDH 164, Procalcitonin 0.11, Ferritin 545
  - - HIV/CMV/Arbovirus/Histoplasma
  - + C. diff PCR; + FOBT

- **Consults:** Hematology, Rheumatology

- **Diagnosis:** C. difficile colitis

- **Treatment:** PO Vancomycin and Metronidazole; restarted home Prednisone
Timeline of events

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May-July 2016: Rheum, Heme, Pulm, GI f/u; steroid taper; ongoing concern for lymphoma

July 2016 (early): B symptoms + N/V/D; C diff + (x2)

July 2016 (late): CT CAP: worse LAD; final LN biopsy
Our Case: 3rd Encounter

• Later that month re-presented with same symptoms
• Work-up:
  - Labs: LDH 276, ESR 58, negative C. diff PCR
  - CT C/A/P with worsening diffuse lymphadenopathy
  - Mediastinal lymph node biopsy:
Diagnosis: Classical Hodgkin's Lymphoma

Lymphocytic Infiltrate

Reed Sternberg Cells
So what happened next...

- **Treatment**: 8 cycles of chemotherapy (BEACOPP)
  - After 6 cycles, PET showed resolution of disease
  - Received Brentuximab instead of Procarbazine or Bleomycin
Quick recap:

- **Sarcoidosis-lymphoma syndrome**
  - Rare but well-documented (first in 1960s)
  - Patients with sarcoidosis have 1.5-9.5x higher risk of developing lymphoma (Hodgkins and non-Hodgkins).
  - 3 important factors (Bichel and Brinker)

- **ITP associated with lymphoma**
  - Hodgkins most common
  - Severity of ITP correlates to severity of lymphoma
  - ITP may be first presenting symptom of lymphoma
First Case Ever?

- Well described association between sarcoidosis and lymphoma; lymphoma and ITP
- No other literature reports ITP in the setting of sarcoid lymphoma syndrome.
Take home points

» Well described association between autoimmune disorders and lymphoma (Hodgkins most common)
» Sarcoidosis almost always precedes lymphoma
» Negative lymph node biopsy does not rule out lymphoma
Lesson Learned: When it doesn’t fit the script, the search you cannot quit!
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Questions????


