LEARNING OBJECTIVES:
- To maintain a high clinical index of suspicion for diagnosing disseminated coccidioidomycosis, as prompt treatment may delay disease progression and prevent further complications such as cerebrovascular accident
- To understand the role and duration of steroid therapy treatment in cerebral vasculitis remains unknown, as this is an extremely rare complication of disseminated coccidioidomycosis

CASE PRESENTATION:
History of present illness: A 70-year-old Caucasian female with no significant past medical history, admitted for further work-up following a one-month history of new onset headache, two weeks of intermittent subjective fever, fatigue, and a twenty pound weight loss. She recently traveled to Arizona, and had no other significant exposures. History revealed a recent sinus infection and pneumonia with a course of macrolides.

Past medical history: Medications prior to admission:
- Dyslipidemia
- Aspirin 81mg
- Atorvastatin 20 mg
- Tylenol prn
- Social and family history: non-contributory

Physical exam: Significant only for submandibular lymphadenopathy, but was otherwise unremarkable.

Laboratory Studies:
- Basic metabolic panel: hyponatremia to 128
- CBC, LFT’s
- Hematologic:
  - Baseline Hematocrit: 42%
  - White blood cell count: 12.5 K/mm³
  - Platelet: 261 K/mm³
- Fungal serology
- Basic metabolic panel: hyponatremia to 128
- CBC, LFT’s
- Hematologic:
  - Baseline Hematocrit: 42%
  - White blood cell count: 12.5 K/mm³
  - Platelet: 261 K/mm³

Imaging:
- CT head: hypodensity within the right basal ganglia and hypodensity in the medial right temporal lobe concerning for infarct
- Brain MRI: area of infarct in the right basal ganglia
- Brain MRA: Perivascular enhancement of the right middle cerebral artery and adjacent infarct is concerning for vasculitic involvement
- Brain MRI: cannot visualized large vessels vasculitis

Pathology: Figure 3
- A: Granuloma on biopsy
- B: Multiple spherules
- C: Coccidioides immitis spherule contains multiple endospores
- D: GMS stain with positive cultures for Coccidioides

DISCUSSION:
Coccidioidomycosis is endemic in the southwestern United States and northwestern Mexico region. 150,000 patients are diagnosed yearly with primary pulmonary coccidioidomycosis. Most patients respond well to therapy, with only 1-5% progressing to disseminated disease. The most feared extrapulmonary complication is CNS involvement.

- Coccidioidal meningitis (CM)
- Found in 1% of individuals with disseminated disease
- Presents with non-specific symptoms: headache, altered mental status, with or without fever, personality changes, nausea, vomiting, and focal neurological deficits
- Diagnosis: CSF studies - lymphocytic pleocytosis, elevated CSF protein and depressed glucose, histopathological staining showing endo- and evaginulating spherules with positive cultures for Coccidioides
- Complications include: hydrocephalus, vasculocerebral infarction, venous and dural thrombosis, hyponatremia

- Vasculitic infarction
- Extremely rare complication, difficult and controversial to manage
- Largest 10 case series of immunocompetent patient diagnosed with CM with associated vasculitis
- Primarily presented with non-specific pulmonary symptoms
- All had focal neurological findings at some time during their illness including hemiparesis, aphasia, somnolous menaprosysis, memory deficits
- Imaging consistent with infarction in 8/9 patients
- 7/10 died (mean of 9.6 months)
- Therefore, even in the era of triazoles and liposomal amphotericin B formulations, mortality in those with coccidioidal vasculitis approach that of 70% in nine month

REFERENCES
1. Harrison’s Principles of Internal Medicine
5. Vascular Principles of Internal Medicine

CONCLUSION
- Treatment for coccidioidomycosis center around triazoles and amphotericin B
- Disseminated disease: liposomal amphotericin B
- Meningeal disease: liposomal amphotericin B + low vs high dose triazoles (fluconazole)
- Vasculitic: liposomal amphotericin B + high dose fluconazole + high dose steroids
- Steroid duration and tapering remains controversial
- Unclear exact mortality benefit
- No standard therapy guidelines
- High mortality regardless of therapy

Pathology slide courtesy by Aimie Millstein, MD