

INTRODUCTION

- ❖ Patients with coccidioidomycosis are often asymptomatic (60%). Symptoms manifest typically in the immunocompromised population
- ❖ Extrapulmonary coccidioidomycosis is rare and usually manifests after initial pulmonary involvement (5-20%)
- ❖ Up to 50% of central nervous system (CNS) involvement associated with disseminated disease develops hydrocephalus (HCP), with 12 fold increase risk in mortality
- ❖ Early diagnosis is to avoid catastrophic complications: hydrocephalus, vasculitis, and thrombosis
- ❖ Primary CNS presentation in an immunocompetent patient and no known prior disease is rare
- ❖ This is a case of an immunocompetent individual presenting with isolated coccidioidal meningitis and HCP without initial systemic manifestations
- ❖ We discuss the diagnostic challenges in cerebrospinal fluid (CSF) evaluation and therapeutic measures given intraventricular involvement

CLINICAL COURSE

- ❖ 39 year old immunocompetent man presenting with progressive headaches with associated visual deficits for 4 months, and imbalance, intermittent vision loss, confusion, 1 month prior to admission
- ❖ Exam: Encephalopathic, right afferent pupillary defect, bilateral lateral gaze palsy, gait imbalance
- ❖ External ventricular drain (EVD) placement for communicating HCP
- ❖ Repeated CSF studies from EVD unremarkable
- ❖ Prophylactic antibiotics + fluconazole → improvement in cognition
- ❖ Ventriculoperitoneal shunt placed, dural biopsy performed: acute and chronic inflammation with no fungal elements identified
- ❖ Lumbar puncture: lymphocytic leukocytosis and significantly elevated protein (WBC 224, Lymph 84%, Protein 410, Glucose 39)
- ❖ Lumbar CSF coccidioides antibody elevated at >1:1024 titer
- ❖ Repeat serial lumbar punctures demonstrated decrease in CSF protein, lymphocytosis

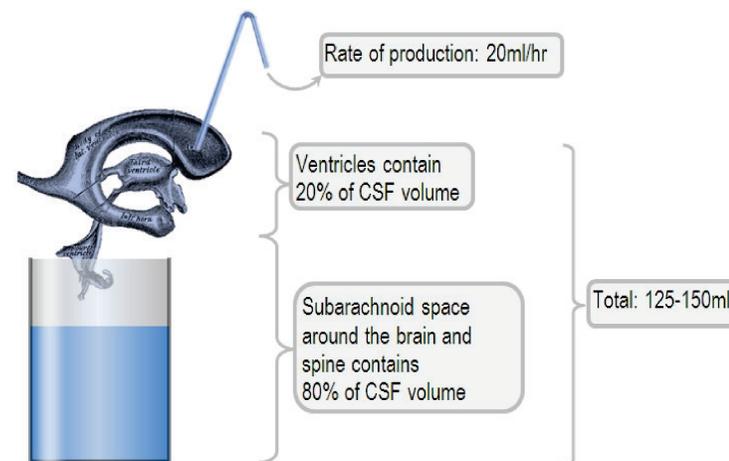
DIAGNOSTIC HURDLES

- ❖ Initial CSF studies from EVD unremarkable and non-diagnostic → unknown choice and course of therapy
- ❖ Biochemical and immunological composition of CSF varies along the cerebrospinal axis, potentially delaying confirmation of diagnosis
- ❖ No systemic involvement despite thorough investigation with imaging and serum studies

THERAPEUTIC MEASURES

- ❖ Early initiation anti-fungal agents in chronic meningitis
- ❖ HCP may develop despite appropriate therapy; need not require alternative therapy
- ❖ 75% relapse rate in patients with HCP
- ❖ Severe presentation mandates life-long fluconazole

AREAS OF CSF SAMPLING



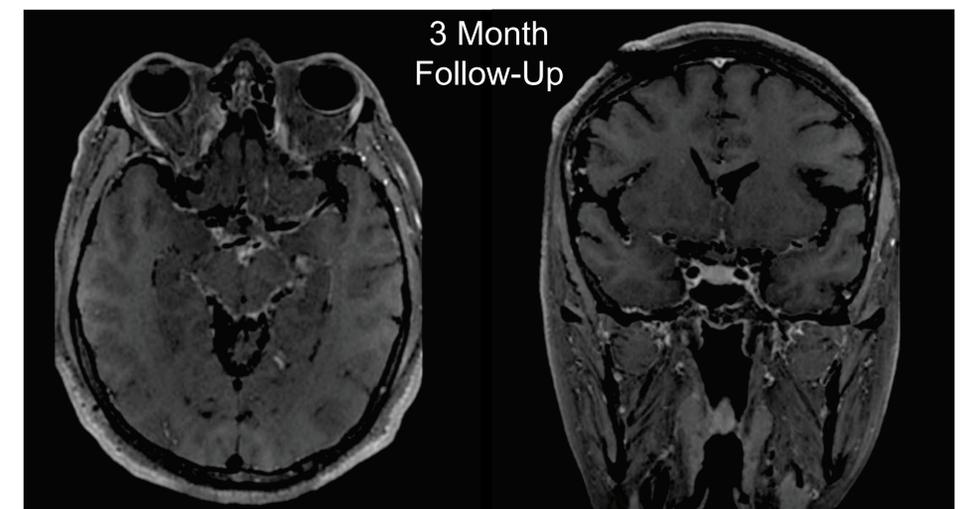
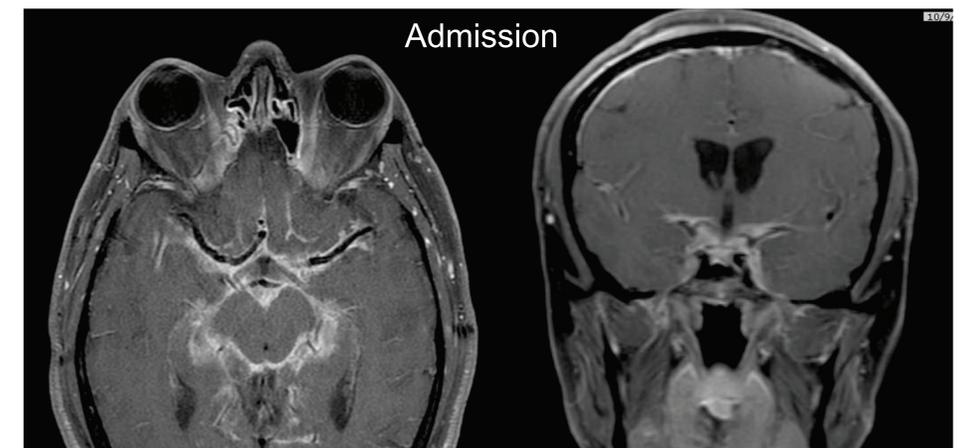
CONCLUSIONS

- ❖ Severe CNS manifestations of fungal disease can present even with immunocompetence
- ❖ Subarachnoid space as an immunologically active substratum
- ❖ Low threshold for initiating anti-fungals in case of chronic meningitis
- ❖ Intact immune system and early therapy contributed to remarkable clinical recovery



DISEASE PROGRESSION

Enhancement surrounding bilateral optic nerve sheaths and optic chiasm. Leptomeningeal enhancement throughout the visualized sulci, Sylvian fissures, basilar cisterns, cerebellar folia, and abnormal enhancement about the bilateral CN VII/VIII complexes and Meckel's cave are also noted.



Decreased thickening and leptomeningeal enhancement of the basilar cisterns

SELECTED REFERENCES

- Hardesty, Douglas A., et al. "Patient outcomes and surgical complications in coccidioidomycosis-related hydrocephalus: An institutional review: Clinical article." *Journal of neurosurgery* 121.4 (2014): 785-789.
- Galgiani, John N., et al. "2016 Infectious Diseases Society of America (IDSA) Clinical Practice Guideline for the Treatment of Coccidioidomycosis." *Clinical Infectious Diseases* (2016): ciw360.
- Wong, F.W. "Cerebrospinal fluid collection: A comparison of different collection sites on the external ventricular drain." *Dynamics* 22.3 (2011): 19-24.
- Drake, Kendra W., and Rodney D. Adam. "Coccidioidal meningitis and brain abscesses Analysis of 71 cases at a referral center." *Neurology* 73.21 (2009): 1780-1786.
- Erly, William K., et al. "MR imaging of acute coccidioidal meningitis." *American journal of neuroradiology* 20.3 (1999): 509-514.