Cryptic Presentation of Disseminated Cryptococcosis

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ABSTRACT

Cryptococcosis is a global invasive mycosis, commonly encountered in patients with HIV/AIDS with low CD4 counts, diabetics, organ and stem-cell transplant recipients, malignancies, and other patients with immunosuppression. The presentation depends on which organ is usually involved although multi-organ involvement may be present. Here, we describe a young female with an enlarging flank mass, found to have disseminated cryptococcosis in the setting of immunosuppression.

KEYWORDS: Cryptococcus, cryptococcus neoformans, immunocompromise, disseminated cryptococcosis, abscess

CASE PRESENTATION

A 47-year-old female with history of relapsing-remitting multiple sclerosis, on fingolimod, presented with enlarging right flank mass that has been present for one year. The mass reportedly fluctuated in size without intervention or drainage, and it would typically worsen and improve over the course of a few weeks. Physical exam was remarkable for a large, non-fluctuant right flank mass with mild tenderness and erythema without drainage or any other skin lesions. CT scan showed complex, right posterior lateral chest wall intramuscular fluid collection measuring 10 x 6.4 x 9 cm with extension into the right eighth intercostal space with cortical irregularity of the adjacent 10th rib **(Figure 1)**. Aerobic, anaerobic, and fungal cultures grew cryptococcus neoformans **(Figure 2)**. Serum cryptococcus antigen was positive at a titer of 1:256.

MRI of the brain demonstrated innumerable supra and infratentorial enhancing lesions with several lesions demonstrating vasogenic edema. There was relative sparing of the basal ganglia, which is typically noted in cryptococcal disease. A lumbar puncture (LP) showed a mildly elevated opening pressure of 31cm H20, although this was with the patient sitting up, and cryptococcal antigen titer of 1:64 in CSF. A repeat LP revealed an opening pressure of 14.5 cm H20.

Infectious disease consultants recommended an induction regimen of amphotericin and flucytosine for a 6-week course; however, the patient did not receive flucytosine due to insurance issues. She was started on fluconazole 800 mg daily in addition to the amphotericin and was asked to stop taking fingolimod at the time of discharge from the hospital. Repeat LP performed after 6 weeks showed no yeast or fungal elements in the CSF. The patient stated she was feeling better and was continued on fluconazole as part of consolidation regimen for 8 more weeks.

Figure 1. CT scan of abdomen pelvis with IV contrast showing fluid collection measuring 10 x 6.4 x 9 cm with extension into the right eighth intercostal space.

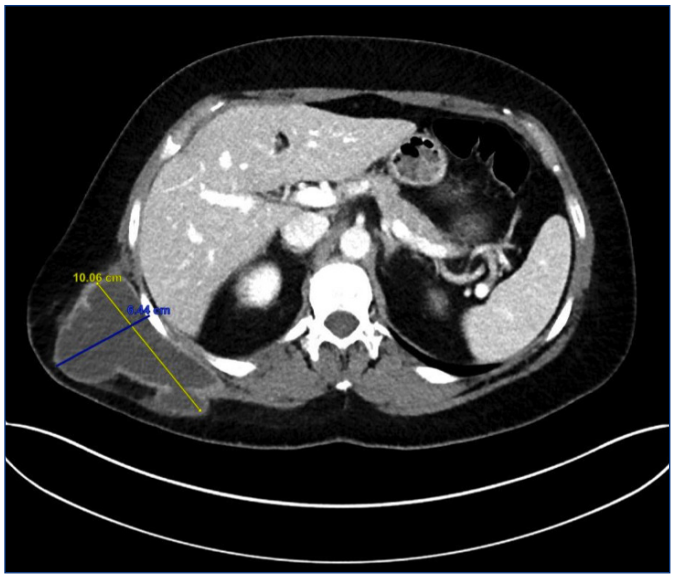
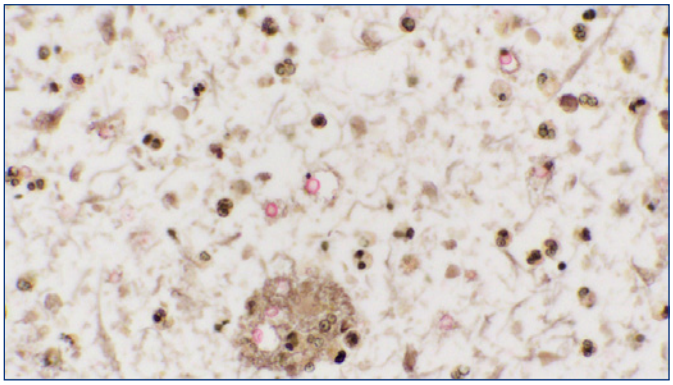


Figure 2. The photomicrograph of mucin-stained sections of the fluid at magnification x600. Cryptococcus neoformans demonstrated as mucin positive spores.



DISCUSSION

Cryptococcosis is caused by an encapsuled opportunistic fungus and is usually due to cryptococcus neoformans or cryptococcus gattii. Cryptococcus neoformans can affect any organ or tissue, but is mostly acquired through inhalation to the lungs and can then disseminate to other sites including the central nervous system (CNS), bone and skin. The clinical presentation of cryptococcosis varies between immunocompromised and immunocompetent patients. Meningoencephalitis is common in immunocompromised individuals whereas in immunocompetent individuals, patients generally present with pulmonary or CNS space-occupying lesion.[1]

Infectious Diseases Society of America guidelines recommend amphotericin B in conjunction with flucytosine as primary induction therapy for disseminated cryptococcosis, followed by fluconazole for consolidation therapy. Increased intracranial pressure and presence of cryptococcomas require special strategies for management. CSF pressure should be measured at baseline. If the CSF pressure is ≥25 cm H_20 and there are symptoms of increased intracranial pressure during induction therapy, relief of opening pressure by 50% through CSF drainage is recommended. Repeat LP after induction regimen is necessary to ensure response to therapy and clearance.[2]

Disseminated cryptococcosis is associated with significant morbidity and mortality. Delay in diagnosis leading to delay in appropriate anti-fungal treatment is related to increased mortality in non-HIV patients compared to HIV patients.[3-5] This case demonstrates that diagnosing cryptococcosis can sometimes be challenging, especially in patients with atypical presentations. Clinicians must maintain a high index of suspicion in immunosuppressed patients for timely diagnosis of disseminated cryptococcosis to facilitate prompt anti-fungal treatment.

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