

A Rare Presentation of Cauda Equina Syndrome due to Cryptococcosis

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ABSTRACT

Introduction: Cauda Equina Syndrome (CES) is a frequently encountered neurological disorder characterized by dysfunction of the sacral nerve roots. Cryptococcosis is a fungal infection caused by exposure to fungi of genus cryptococcus. It most often presents in the pulmonary and central nervous systems. Spinal cryptococcosis is much rarer presentation.

Case Description: We present the case of a 51-year-old male with a history of heavy alcohol use presenting with a sudden onset of weakness in his lower extremities and fever after a vacation to Mexico. After undergoing bilateral decompressive laminectomy, durotomy, and partial resection, pathology showed evidence of cryptococcal infection in the cauda equina region. The patient was treated with induction therapy with Amphotericin B and Flucytosine with a good outcome.

Conclusion: *Cryptococcus neoformans gattii* infection caused a mass-like arachnoiditis, without meningoencephalitis, and is the first fungal infection presenting as CES to our knowledge.

CASE REPORT

INTRODUCTION

A 51-year-old male had an insidious onset of bilateral weakness in the lower extremities, which was progressively worsened over a period of 12 months. Additionally, he experienced bowel and urinary incontinence for 3 months. He was sent to the emergency department after an abnormal outpatient magnetic resonance imaging (MRI).

He also reported several mechanical falls, without significant injury, which required the use of a walker. Since symptom onset, patient had lost 20-30 lbs with an associated loss of appetite. The symptoms started after patient returned from Mexico where he vacationed on a ranch. He did not report interaction with the animals on the ranch.

He had a history of heavy alcohol use, with over 40 drinks weekly, though he stopped drinking 3 months before being seen in the clinic. There was no significant history of tobacco or drug use. Additionally, there was no history of cough, fever, close contact with a patient with tuberculosis, recent infection, or prolonged duration of receiving corticosteroids.

The neurological exam revealed a strength of 4/5 of bilaterally of the lower extremities. Sensation was diminished with presence of numbness and saddle anesthesia. Rectal tone was intact, however deep anal pressure was not present. Other systemic exam including respiratory system, cardiovascular and GI system were within normal limits.

The outpatient MRI showed clumping and thickening of the nerve roots within the thecal sac on T2 weighted images. After contrast administration, there was nodular enhancement, which was more pronounced caudally. Additionally, degenerative disc disease was noted at L5-S1 level.

DIAGNOSTIC PROCESS

The differential diagnoses at this stage radiologically included ependymoma, nerve sheath tumors or drop metastases. Labs were negative for normocytic anemia, hypomagnesemia, and HIV.

Patient underwent posterior bilateral L3-L5 decompressive laminectomy, durotomy and partial resection for initial suspected diagnosis of spinal intradural tumor. Initially, frozen specimens

were suggestive of chordoma. Final pathology was negative for malignancy and showed evidence of Cryptococcosis. Repeat MRI showed persistent leptomeningeal enhancement of the cauda equina, consistent with the known history of spinal Cryptococcus. Lumbar puncture was then performed and revealed a cryptococcal antigen titer of 1:2048 and CSF culture was positive *cryptococcus gattii*. The rate of positive fungal culture in the CSF is low and fungal infection cannot be ruled out based off CSF culture. Patient was started on Amphotericin B with Flucytosine. 2nd lumbar puncture was done 7 days after starting the treatment and the antigen titer was 1:1024. A 3rd lumbar puncture was done 5 days after the 2nd lumbar puncture and titer was 1:256. Follow-up lumbar puncture cultures were negative (Tables 1,2).

CASE RESOLUTION

The patient completed a total of 4 weeks of induction therapy with Amphotericin B and Flucytosine. Post induction, patient's symptoms improved, and he reported no fevers, headaches, back or neck stiffness. We noted improvement of his lower extremity weakness, and he was able to ambulate with walker. Patient was discharged home with a prescription of 800 mg of oral fluconazole. Patient was treated with antifungals for a total period of 5 months. During his follow up visit, his strength in bilateral lower extremity had improved and he had no signs of infection.

DISCUSSION

Patient's initial diagnosis was of a spinal intradural extramedullary tumor due to mass-like appearance in the Cauda Equina region on the MRI. After surgical decompression and laminectomy was conducted, aggregates of foamy histiocytes (supported by positive CD68 stain) which are associated with somewhat ovoid to irregular yeast-type organisms which are highlighted on PAS, GMS and mucicarmine stains were found leading to diagnosis of cryptococcosis.

Cryptococcosis is a fungal infection caused by exposure to fungi of the genus *cryptococcus*. There are 2 known strains of cryptococcosis, *Cryptococcus neoformans*, *Cryptococcus gattii* and *Cryptococcus grubii*. *C. neoformans* is found in environmental reservoirs around the world while *C. gattii* is found mainly in eucalyptus trees in tropical and sub-tropical climates. 95% of the cases of cryptococcal infection occur from the *C. neoformans* (serotype A) strain and the remaining 5% occurs from the *C. gattii* (serotype D) strain (1). *C. grubii* is also widespread but is not pathogenic.

Cryptococcosis most commonly involves the pulmonary and the central nervous system (CNS). Less common sites of involvement have been reported in the skin, prostate, eyes, bones, and joints. In the lungs, cryptococcus vary from asymptomatic colonization to pulmonary nodules on chest radiographs to life threatening pneumonia [1]. Skin lesions when present indicate disseminated cryptococcal infection (1). In the brain, symptoms

involve headache, fever and altered mental status. On MRI, the most common reported finding is leptomeningeal enhancement and perivascular space dilation in the brain (2). Oshima et al have reported similar findings on magnetic resonance imaging in both immunocompromised and immunocompetent patients [2].

Spinal cryptococcosis is a fungal infection associated with immunocompromised patients. The most common presentation of spinal cryptococcosis is cryptococcal meningitis and meningoencephalitis [3]. Another manifestation of cryptococcosis found in existing literature is intramedullary spinal cryptococcoma [4]. Our case highlights mass like arachnoiditis of the cauda equina as a cause of cryptococcosis without the presence of meningoencephalitis.

Cauda Equina Syndrome (CES) is a frequently encountered neurological disorder characterized by dysfunction of the sacral nerve roots S2 or below resulting in loss of feeling in lower extremities and urinary incontinence [5]. The most common cause of CES is disk herniation. Other causes of CES include spinal lesions, tumors, birth abnormalities and trauma. Infection such as tuberculosis and bacterial infection can also be a cause of CES. Fungal infection presenting as CES is rare. Other reports discuss cases of Aspergillosis and exposure to other pathogenic organisms [6]. There is no existing report to our knowledge of CES from cryptococcal infection (Figures 1-11).

Travel history is also important in recognizing exposure to rare pathogens as seen in our patient who was immunocompetent and had no recent history of fever, headache, and other inconsistencies. There are few reports that present cryptococcal infection because of short-term travel.

CONCLUSION

Cryptococcus neoformans var *gattii* is a rare cause of CES in an immunocompetent patient. We describe its clinical presentation, imaging, CSF and pathological findings. Infection can mimic tumor and must be considered in the differential diagnosis in a patient with international travel history.

TEACHING POINT

The most common cause of Cauda equina Syndrome is disc herniation or neoplastic process. *Cryptococcus neoformans* var *gattii* is a rare cause of CES in an immunocompetent patient.

AUTHORS' CONTRIBUTIONS

Author 1 is the principal investigator who designed the case report and supervised authors 2 and 3 in writing of the case report.

Author 2 collected most of the clinical data and provided significant contribution to the preparation of case report.

Author 3 was instrumental in the article research and preparing the manuscript.

CONSENT

Did the author obtain written informed consent from the patient for submission of this manuscript for publication? Yes a verbal consent was obtained.

QUESTIONS

Question 1: What is the most common cause of Cauda Equina Syndrome?

- Answer choice 1: Cryptococcosis
- Answer choice 2: Tuberculosis
- Answer choice 3: Spinal disc herniation
- Answer choice 4: Trauma
- Answer choice 5: Spinal cord tumor

Explanation

Answer choice: 3 The most common etiology of CES is herniated lumbar disc, which accounts for approximately 45% of cases (Korse NS., et al 2017)

Question 2

Question: Which of the following is the most suitable diagnostic modality for Cauda Equina Syndrome?

- Answer choice 1: CT Lumbar spine
- Answer choice 2: MRI Lumbar spine
- Answer choice 3: Surgical biopsy
- Answer choice 4: CT Myelogram
- Answer choice 5: EMG and Nerve conduction studies

Explanation:

Answer choice: 2. MRI plays a vital role in visualizing the spinal canal and identifying the underlying pathology responsible for CES, such as nerve root compression (Lavy et al., 2022).

Question 3

Question: Which of the following statement regarding treatment of cauda equina syndrome is true?

- Answer choice 1: Corticosteroid therapy
- Answer choice 2: Emergent surgery
- Answer choice 3: Surgical biopsy
- Answer choice 4: Antibiotics
- Answer choice 5: Antifungal treatment

Explanation:

Answer choice:2. Timely surgical intervention is crucial in preventing irreversible neurological deficits and improving patient outcomes (Lavy et al., 2022).

Question 5: Which of the following statement regarding the most common cause of MRI finding of spinal cryptococcosis is true?

- Answer choice 1: Nodular enhancement with cystic appearance
- Answer choice 2: No contrast enhancement
- Answer choice 3: Leptomeningeal enhancement is the most common finding.
- Answer choice 4: Homogenous enhancement
- Answer choice 5: Heterogenous enhancement

Explanation:

Answer choice: 3. On MRI, the most common reported finding is leptomeningeal enhancement and perivascular space dilation in the brain ⁽²⁾

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FIGURES



Figure 1: 51-year-old male with progressive weakness of bilateral lower extremities. MRI Lumbar spine, 1.5 tesla T1 pre-contrast, pretreatment. Demonstrates clumping of the nerve roots within the thecal sac, which appear thickened. The enhancement and nodularity are more pronounced caudally. The possibility of intradural adhesive change- arachnoiditis or possible neoplastic process suspected.



Figure 2: 51-year-old male with progressive weakness of bilateral lower extremities. MRI Lumbar spine, 1.5 tesla T1 post-contrast, pretreatment. Demonstrates clumping of the nerve roots within the thecal sac, which appear thickened. The enhancement and nodularity is more pronounced caudally.



Figure 3: 51-year-old male with progressive weakness of bilateral lower extremities. MRI Lumbar spine, 1.5 tesla T2 image, pretreatment.



Figure 4: 51-year-old male with progressive weakness of bilateral lower extremities. MRI Lumbar spine, 1.5 tesla T1 pre-contrast post treatment.

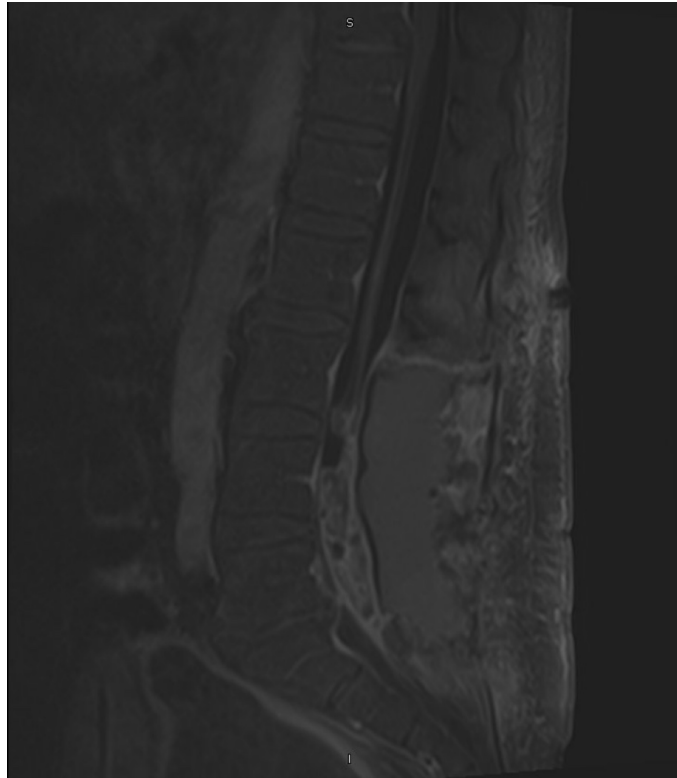


Figure 5: 51-year-old male with progressive weakness of bilateral lower extremities. MRI Lumbar spine, 1.5 tesla T1 post-contrast, post treatment. Demonstrates heterogeneous enhancing mass and multiple small cystic changes noted and appearance of split of the cauda equina.

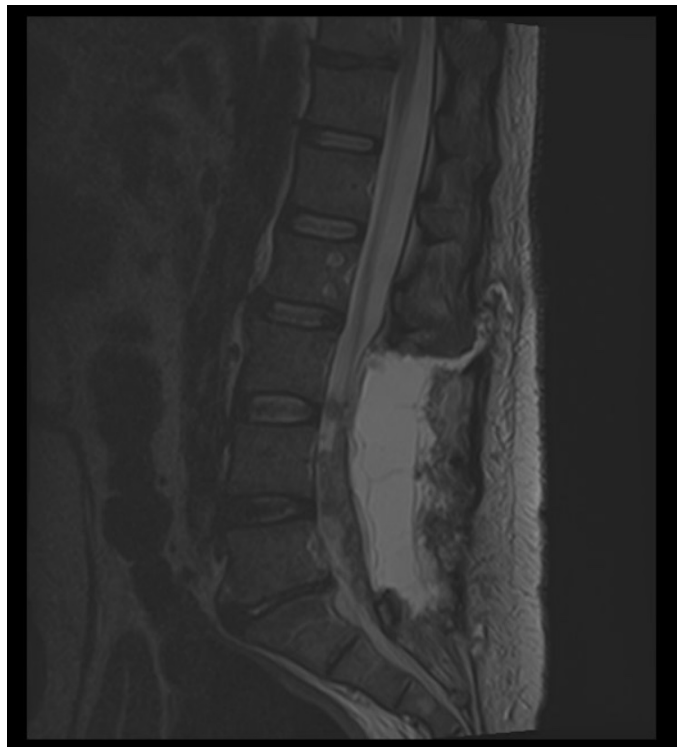
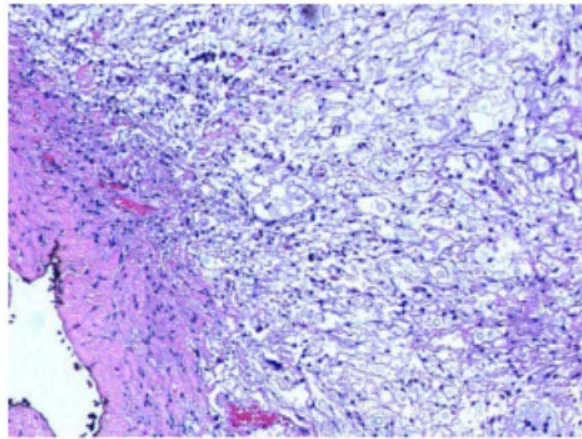


Figure 6: 51-year-old male with progressive weakness of bilateral lower extremities. MRI Lumbar spine, 1.5 tesla T2 post treatment. Demonstrates heterogeneous enhancement of the cauda equina centered at the L4-L5 level with peripherally enhancing lobulated configuration.

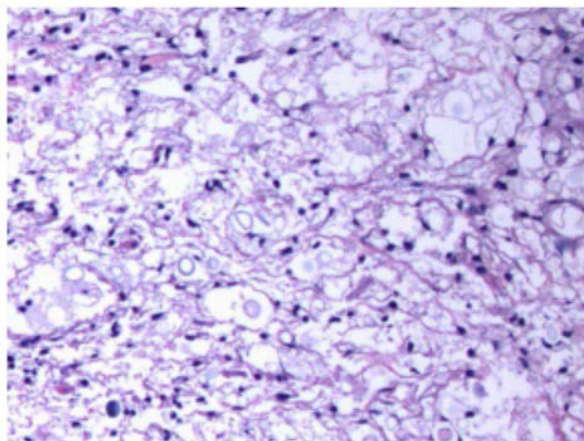
Frozen section of excision biopsy

- A. Chronically inflamed fibrous tissue with some adjacent histiocytic cells with associated GMS positive/PAS positive yeast-type organisms most suggestive of Cryptococcus organisms.
- B. No evidence of malignancy.



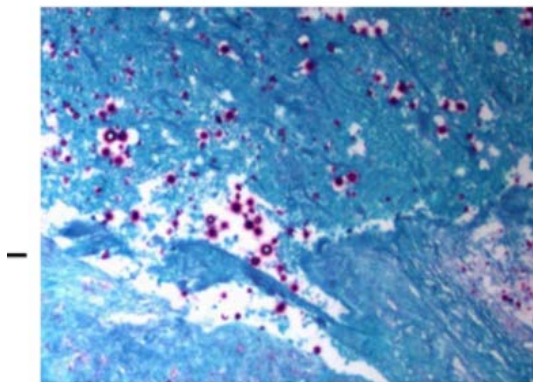
S22-54183-1: Intradural L3 spinal tumor:

Figure 7: Hematoxylin and Eosinophil Stain of L3 spinal tissue/tumor.

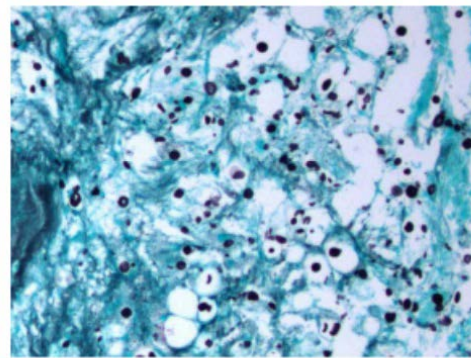


S22-54183-1: Intradural L3 spinal tumor:

Figure 8: CD 68 showing numerous foamy cells which are positive, consistent with histiocytes and macrophages.

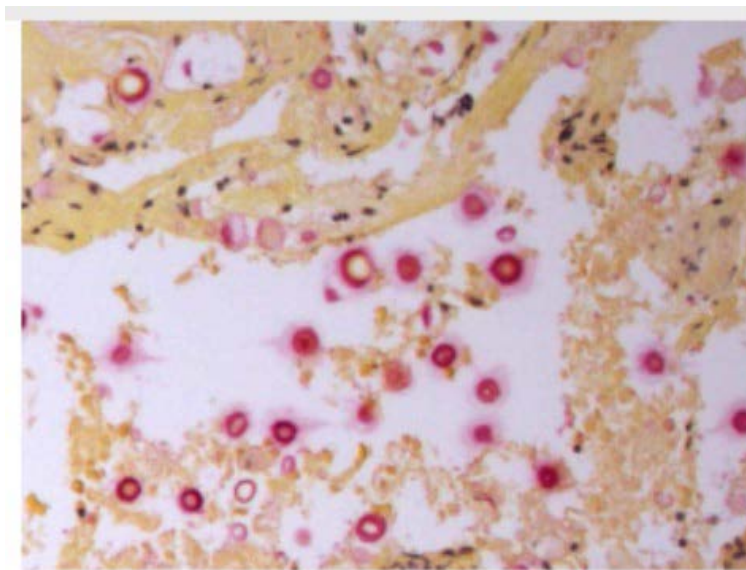


S22-54183-1: Intradural L3 spinal tumor: PAS



S22-54183-1: Intradural L3 spinal tumor: GMS

Figure 9 and 10: Grocott's methanamine silver stain (GMS) and periodic acid schiff stain (PAS): Chronically inflamed fibrous tissue with some histiocytic cells with GMS positive and PAS positive yeast type organisms most suggestive of cryptococcus organisms.



S22-54183-1: Intradural L3 spinal tumor: Mucicarmine

Figure 11: Mucicarmine stain: Foamy histiocytes associated with ovoid to irregular yeast- type organisms. Morphologically suggestive of cryptococcus.

There were prominent aggregates of foamy histiocytes (supported by positive CD68 stain) which are associated with somewhat ovoid to irregular yeast-type organisms which are highlighted on PAS, GMS and mucicarmine stains. Morphologically these organisms appear most suggestive of Cryptococcus. No malignancy is identified on permanent sections.

Table 1: 51-year-old male presented with progressive bilateral lower extremity weakness, the CSF analysis shows persistently increased protein and low glucose.

	Initial Lumbar Puncture	2nd Lumbar Puncture 4 weeks post treatment	3rd Lumbar Puncture 8 weeks post treatment
Color	Yellow	Colorless	Xanthochromic
Pressure (cm H2O)	8	11	5
WBC	57	84	76
Protein	386	349	439
Glucose	12	19	33
Culture	No growth	No growth	No growth

Table 2: 51 y/o male with progressive lower extremity weakness, the cryptococcal antigen titer were significantly elevated at presentation and show a decreasing trend with treatment.

	Initial	2nd	3rd
Cryptococcal antigen titers	1:2048	1:1024	1:256

KEYWORDS

Bilateral lower extremity weakness; Paraparesis; Cauda equina syndrome; CNS cryptococcosis; Spinal cryptococcosis

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