



Case Report

Cryptococcal meningitis presenting as anterior spinal cord syndrome with accessory nerve palsy in immunocompetent patient: A case report

J Javier Cuellar-Hernandez¹, Carlos Seañez², Ramon Olivas-Campos², Rodrigo Chavez², Paulo M. Tabera-Tarello², B. Manuel Serna-Roman²

¹Department of Neurosurgery, Swiss Hospital, ²Department of Neurosurgery, Northeast National Medical Center, Monterrey, Mexico.

E-mail: *J Javier Cuellar-Hernandez - javiercueher@gmail.com; Carlos - dr.seanezumae25@hotmail.com; Ramon Olivas-Campos - ramon_olivasc@hotmail.com; Rodrigo Chavez - drchavezneuro@gmail.com; Paulo M. Tabera-Tarello - paulo.taberam@gmail.com; B. Manuel Serna-Roman - manu.serna69@hotmail.com



*Corresponding author:

J Javier Cuellar-Hernandez,
Department of Neurosurgery,
Swiss Hospital, Monterrey,
Mexico.

javiercueher@gmail.com

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ABSTRACT

Background: *Cryptococcus* has a tropism for the nervous system with a higher prevalence of infection in immunosuppressed patients; it remains a major cause of human immunodeficiency virus (HIV)-related mortality worldwide. Neurological compromise caused by this microorganism mainly debuts as a meningeal syndrome, spinal involvement has been reported in literature, neuropathological assessments have found *Cryptococci* in spinal roots and meninges, with perineuritic adhesions probably explaining compromise lower cranial nerves and even spinal nerve roots.

Case Description: 39-year-old male seronegative for HIV, with a surgical history of hydrocephalus treated with ventriculoperitoneal shut 1 year before, he presented with progressive weakness in the four extremities evolving to be disabling with bilateral accessory nerve palsy and loss of sensation below his neck. The MR imaging showed diffuse leptomeningeal thickening both supra and infratentorial and over the spinal canal up to C5 with a cystic formation shown in the craniocervical union causing compression of the medullary bulb. The patient underwent a medial suboccipital craniectomy with resection of the posterior arch of c1 for sampling and decompression, pathologically appears numerous spherical organisms that have a thick clear capsule and are surrounded by histiocytes forming a granuloma compatible with *Cryptococcus*. Postoperatively, the patient's prior neurological deficits resolved.

Conclusion: It is an infrequently suspected pathology in immunocompetent patients, usually requiring only antifungal treatment with adjustment of immunosuppressive or antiretroviral management. In special and rare situations like our case as presenting with lower cranial nerve and spinal involvement, surgical treatment is a priority for the resolution of the pathology and improves disabling neurological deficit.

Keywords: Anterior spinalcord syndrome, *Cryptococcus*, Meningitis, Suboccipital craniectomy

INTRODUCTION

Cryptococcus has a tropism for the nervous system with a higher prevalence of infection in immunosuppressed patients; it remains a major cause of human immunodeficiency virus (HIV)-related mortality worldwide. Is an opportunistic infection that causes more than 100,000 HIV-related deaths each year.^[2,5,6] Even though the infection is usually HIV-related, most of the non-HIV-related cases include patients under immunosuppressive treatments or with organic

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failure syndromes, transplants, innate immunological problems, common variable immunodeficiency syndrome, and hematological disorders.^[11]

Neurological compromise caused by this microorganism mainly debuts as a meningeal syndrome characterized by headache, altered mental status, and other signs and symptoms include lethargy along with fever, stiff neck (both associated with an aggressive inflammatory response), nausea, and vomiting. Some patients who are HIV positive may have minimal or nonspecific symptoms at presentation. The duration of symptoms from onset to a presentation is usually 1 to 2 weeks in HIV cases and 6 to 12 weeks in non-HIV cases.^[12]

Cryptococcal meningitis with spinal involvement has been reported in literature, neuropathological assessments have found Cryptococci in spinal roots and meninges, with perineuritic adhesions^[11] probably explaining compromise lower cranial nerves and even spinal nerve roots. Here, we a 39-years-old male who debuts with hydrocephalus and subsequent anterolateral spinal cord syndrome with bilateral accessory nerve palsy.

CASE PRESENTATION

A 39-year-old male, inhabitant of Mexico northeastern endemic area for *Cryptococcus* with a surgical history of

hydrocephalus treated with ventriculoperitoneal shut 1 year before, he presented 3 months of progressive weakness in the four extremities evolving to be disabling with bilateral accessory nerve palsy and loss of sensation below his neck.

On physical examination, he had increased deep tendon reflexes and 2/5 muscle strength in his bilateral upper and lower extremities with a sensory level of C3, intact proprioception and vibration, with absent pain and temperature sensation.

The magnetic resonance imaging of the brain and craniocervical junction with and without contrast showed diffuse leptomeningeal thickening both supra and infratentorial and over the spinal canal up to C5, with great enhancement to contrast administration, with involvement in ambiens, interpeduncular, prepontine, sylvian, premedullary, and Magna cisterns. Cystic formation is also shown in the cranio-cervical union causing compression of the medullary bulb [Figure 1].

The patient was seronegative for HIV.

Surgery

The patient underwent a medial suboccipital craniectomy with resection of the posterior arch of c1, intradurally found meningeal layer of the dura with multiple adhesions to the

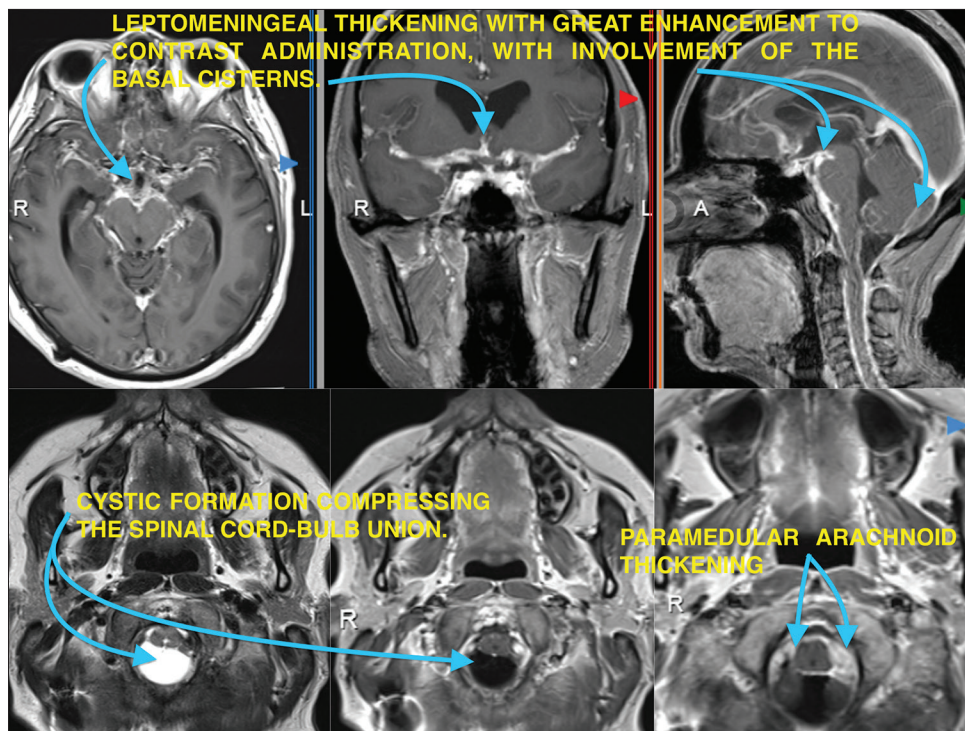


Figure 1: Preoperative MR showing diffuse leptomeningeal thickening both supra and infratentorial and over the spinal canal up to C5, with great enhancement to contrast administration, with involvement of the basal cisterns (ambiens, interpeduncular, prepontine, sylvian, premedullary, and Magna).

thickened arachnoid with multiple nodules on its surface with poor vascularization, opening of the arachnoid with partial resection on the cistern magna and on the bulbomedullary cistern with sampling for definitive pathological study, a sample of the pia mater was taken, which was thickened with the same characteristics of the arachnoid. Pathological cerebrospinal fluid (CSF) found in an arachnoid cyst formed by multiple adhesions, cerebellar tonsils were released with opening of the medullary velum until the IV ventricle was identified, choroid plexuses of calcified Lushka foramens with whitish granulations inside. Cerebellar hemisphere, biventral lobe as well as superior and inferior semilunar lobes with pial narrowing maintaining arterial integrity. Firm adhesions are observed on emergence of both accessory nerves as well as bilateral IX, X, XI, XII, and PICA [Figure 2]. The histopathology revealed numerous spherical organisms that have a thick clear capsule and are surrounded by histiocytes forming a granuloma, multinucleated giant cells with spherical structures that have a clear mucoide capsule surrounding a central pale nucleus compatible with *Cryptococcus* [Figure 3]. Postoperatively, the patient's prior neurological deficits resolved.

DISCUSSION

Cryptococcosis is an infectious disease that frequently affects the lungs and central nervous system. It is caused by

Cryptococcus neoformans and *Cryptococcus Gattii*, the first is associated with immunocompromised patients due to organ transplantation, especially in patients with advanced HIV disease and low CD4 cell count, and the second is more commonly associated with immunocompetent patients.^[2,5,7,9,11] This fungal disease occurs in 10–20% patients without detectable immunocompromising factors^[1,10] and has a mortality of 35–50% due to delay in diagnosis and treatment.^[8]

In the central nervous system, it causes encephalitis and meningitis, affecting different levels of the central nervous system, spinal arachnoiditis occurs more sporadically.^[2,3,7,8] The manifestations are established slowly and insidiously, the most common are headaches, lethargy, memory loss, meningeal irritation, fever, nausea, and vomiting.^[2,5,7,9] Our patient presented particular manifestations with long-standing significant motor and sensory involvement in all four extremities, in addition to bilateral paresis of the muscles innervated by the accessory nerve, clinical manifestations that have not been described yet.

The clinical and radiological conditions of this patient led us to consider arachnoid cysts and epidermoid cysts, including tuberculosis arachnoiditis. Offer early treatment avoids possible complications such as the development of cerebral infarcts and intracranial hypertension, which

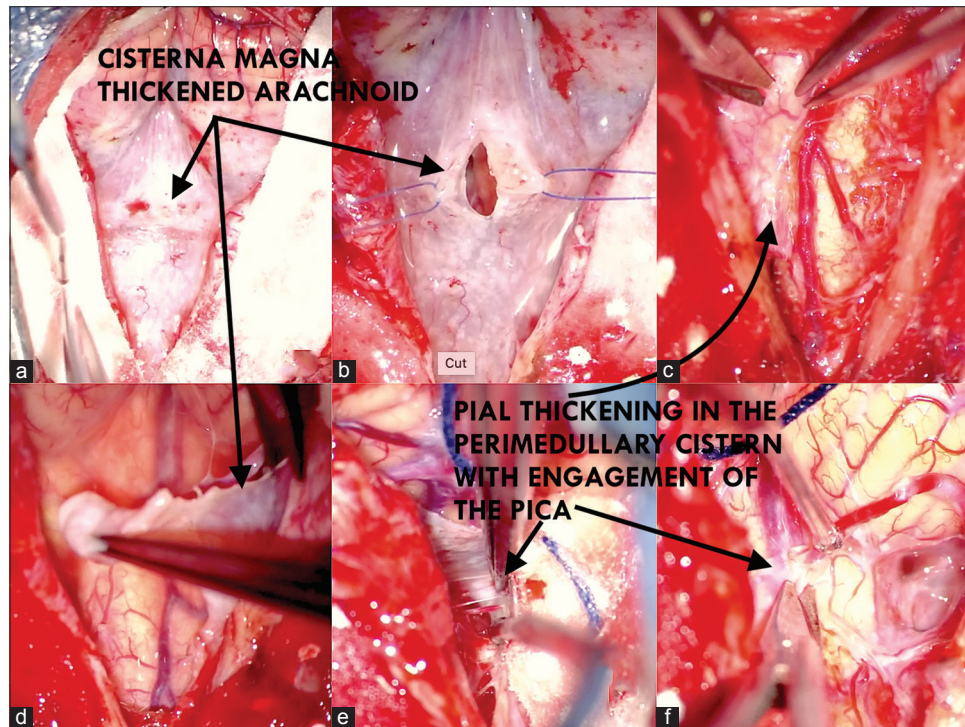


Figure 2: Intraoperative view, multiple adhesions to the thickened arachnoid with multiple nodules on its surface (a and b), the pia mater was thickened with the same characteristics of the arachnoid (c and d). Cerebellar tonsils were released with opening of the medullary velum until the IV ventricle was identified, Firm adhesions are observed on emergence of both accessory nerves as well as bilateral IX, X, XI, XII, and PICA (e and f).

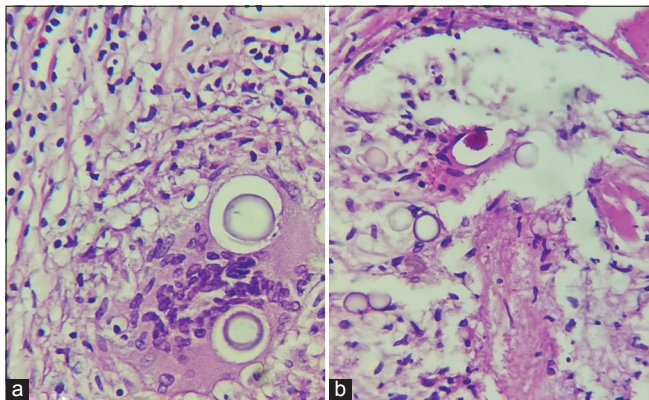


Figure 3: A multinucleated giant cell with two spherical structures that have a clear mucoide capsule (halos) surrounding a central pale nucleus. H&E $\times 40$ (a). Numerous spherical organisms that have a thick clear capsule and are surrounded by histiocytes forming a granuloma. H&E ($\times 40$) (b).

usually require CSF shunt systems in about 50% of cases.^[2-4,7,9]

First-line of treatment is medical with antifungal drugs, with in consensus with international guidelines, amphotericin followed by treatment with azoles for 12 months or more, presents a partial or total recovery of lost functions.^[1,2,7,9,10] In our case, in addition to the previously mentioned treatment, surgery was needed to solve the compression produced by meningeal thickening and cystic formations at the bulbomedullary level, causing significant deterioration of both the ascending and descending pathways, as well as the cranial nerves. The diagnosis was definitively established by histopathological study.^[2,7,9]

CONCLUSION

It is an infrequently suspected pathology in immunocompetent patients, usually requiring only antifungal treatment with adjustment of immunosuppressive or antiretroviral management. In special situations like our case as presenting with lower cranial nerve and spinal involvement, surgical treatment is a priority for the resolution of the pathology and improves disabling neurological deficit. The participation of the infectologist is essential in the establishment and monitoring of antifungal treatment.

DeARATION of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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