Neuroparacoccidiomicosis Simulating Posterior Fossa Tumor

ABSTRACT

Paracoccidioidomycosis (PCM) is a systemic granulomatous disease caused by a fungus known as Paracoccidioides brasiliensis. Central nervous system (CNS) is affected in 12% of the cases, but posterior fossa involvement is rarer. We report a case of a patient who presented an expansive lesion at the posterior fossa causing a right hemispheric cerebellar syndrome. After lesion excision and analysis, the diagnosis of Neuroparacoccidiomicosis was confirmed. Our case highlights this rare presentation of this pathology and its capacity to simulate neoplastic lesions.

Key-words: Paracoccidioidomycosis; Micosis; Brain tumor; Abscess; Infection

INTRODUCTION

Paracoccidioidomycosis (PCM) is a systemic granulomatous disease caused by a dimorphic fungus known as Paracoccidioides brasiliensis. This fungal infection is endemic in the Southern America and, among the countries with higher prevalence rates, Brazil is the one with the majority of reported cases (80%)².

The disease is acquired by inhalation of the conidia produced in the mycelial form of the P. brasiliensis, classically causing a benign pulmonary infection. Subsequently, it can disseminate through the hematologic and/or lymphatic route to other organs, forming secondary lesions in the mucous membranes, skin, lymph nodes, and adrenal glands³,⁴.

Central nervous system (CNS) is affected in around 12% of the cases, and in these, the supratentorial lesions are the majority¹. More specifically, cerebellar involvement is rarer and account for 29% of the neuroparacoccidioidomycosis (NPCM) – only 3% of the overall patients³.

We present a very rare case of a fungal abscess caused by P. brasiliensis with cerebellar involvement simulating a posterior fossa tumor.
CASE REPORT

A 62-year-old male presented to our clinic with a history of progressive headache, dizziness and gait instability, falling forward to the right, for about 2 months. He lived in a rural area in southern Brazil and worked as a farmer. The patient had a history of smoking and primary hypertension. At neurological examination, he presented right dysmetria, kinetic tremor and gait ataxia. No other abnormalities were detected. Routine laboratory studies were normal. The patient had a magnetic resonance imaging (MRI) of the brain that showed a cystic-solid intra-axial lesion in the right cerebellar hemisphere with perilesional enhancement in the contrasted phases and obliteration of the fourth ventricle (Fig. 1). Based on clinical history and radiological findings the hypothesis of a neoplastic lesion was considered.

The patient underwent microsurgical resection of the lesion. After the craniotomy a cerebellar abscess was found, which ruptured intraoperatively. The content of the lesion was thick and greenish. The cyst fluid was aspirated and the capsule was resected without intercurrences. The postoperative period was unremarkable and the follow-up MRI suggested complete resection of the lesion (Fig. 2). The patient was discharged after 3 days, without deficits and in use of Vancomycin, Cefepime and Metronidazole. After the histopathological analysis, the diagnosis of Paracoccidioidomycosis brasiliensis was confirmed and Amphotericin B was prescribed to the patient.

Six weeks later the patient returned to our clinic due to recurrence of symptoms. A new MRI demonstrated the recurrence of the lesion (Fig. 1C). The patient was submitted to a new surgical resection and evolves without deficits. Currently the patient is asymptomatic, in use of antifungal and in follow-up with infectologist.

DISCUSSION

It is estimated that, in endemic areas, the incidence of PCM ranges from 3 to 4 cases per 100,000 inhabitants per year. PCM can occur in individuals of any age group, from 2 years of age, but mainly between 30 and 50 years. It is more prevalent in the male gender and in the rural workers. Furthermore, there is frequent association of PCM with smoking and alcoholism.

Neurological infection is described as secondary, and the main form of dissemination is hematologic, due to the predominance of lesions in areas with high blood flow.

Clinical Presentation

The clinical forms of NPCM are meningeal, meningoencephalic and pseudotumoral and the presence of the neurological symptoms is related to the region of the brain affected. The pseudotumoral form is the most frequent, corresponding to approximately 96% of the cases, and may mimic other pathologies, such as abscesses, granulomas, nodules or intraparenchymal cysts. It preferably occurs at supratentorial location, affecting the cerebral hemispheres.

In the infratentorial cases, common symptoms include gait disturbances, dizziness, nausea, vomiting and diplopia, as well as rapid progression to intracranial hypertension syndrome due...
to ventricular system compression\(^7\).

In addition, motor and sensory deficits, paralysis of cranial nerves, seizures, disturbances of consciousness and mood, aphasia, signs of hydrocephalus may also occur depending of the location of the impairment\(^2,4\).

Our patient presented with a right cerebellar hemispheric syndrome, which may have several etiologies that cannot be rule out without proper evaluation with imaging and histological analysis.

**Evaluation**

Neuroimaging methods (CT and MRI) are considered essential in demonstrating the neurological involvement of the disease. However, it may appear as other intracranial expansive processes in imaging studies, being necessary histological analysis to confirm the diagnosis\(^4\).

MRI is considered the best non-invasive method to evaluate lesions of the meninges or parenchyma, since it is more sensitive in the identification of intraparenchymal lesions, especially infratentorial ones, including cerebellum, brainstem and spinal cord\(^7\). Therefore, it should be the gold standard for the assessment of suspected NPCM. The lesions were characterized by iso or hyposignal in T1, hyposignal in T2, with peripheral edema and ring enhancement or after contrast infusion\(^4,7\).

**Treatment**

The treatment was based on antifungal therapy; the association of sulfamethoxazole-trimethoprin was used as the first option because of the facility in their administration, the high levels reached in the CNS, and the low cost\(^1\). However, Amphotericin B is indicated for severe cases, at least in the initial phase of treatment, in order to prevent unfavorable outcomes\(^5,6,8\). Duration of treatment was based on clinical improvement and regression or stabilization of the radiologic findings\(^5,9\).

The indications for neurosurgical intervention are similar to any brain abscess case and include signs of significant mass effect, associated hydrocephalus, proximity to the ventricular system or neurological deterioration\(^1,4,10\). In addition, surgery can be performed when diagnosis is dubious or in the failure of antibiotics course\(^10\).

Even with the option of surgical treatment, drug therapy should be started early, since the manipulation of granulomas can lead to the spread of the disease\(^2,4\).

We chose treatment with amphotericin B because the lesion occupied a large area in a delicate region of the brain, which could progress to hydrocephalus and severe neurological complications. Moreover, disruption of the cyst during the surgical procedure could cause a fungal spread to other parts of the brain, which highlights the need for more effective treatment.

The indication of surgery to our patient was to obtain elucidation about the etiology of the lesion. Initially, due to the history of smoking and the age range, the main hypothesis raised was of metastatic lesion of pulmonary origin, which shares similar radiologic characteristic and site of involvement.

Finally, we believe that the early recurrence, in our case, resulted probably due to the suboptimal resection of the lesion and/or spread of the cyst fluid.

**Outcomes**

Even with aggressive surgical and drug intervention, NPCM is a high morbidity pathology, and neurological sequelae are very frequent. Mortality is high, estimated at around 53%\(^7\). These are worse prognostic factors: advanced age, comorbidities, multisystemic impairment presence of hydrocephalus\(^2\).

Even with two surgical interventions, our patient had a good progression, recovering from established neurological deficits prior the diagnosis. This can be considered an uncommon outcome considering the unfavorable results reported in the literature.

**Conclusion**

In conclusion, NPCM should be promptly considered in the differential diagnosis of brain tumors in endemic areas when a ring-enhancing mass associated with perilesional edema is observed on MRI. Due to the high mortality rates, the treatment should be introduced as early as possible.
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DISCLOSURES
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