Case Report/Relato de Caso

Bruns' syndrome and racemose neurocysticercosis: a case report

Síndrome de Bruns e neurocisticercose racemosa: relato de caso

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ABSTRACT

Cysticercosis is an infection caused by the larval stage of the tapeworm *Taenia solium*. The parasite may infect the central nervous system, causing neurocysticercosis (NCC). The clinical manifestations depend on load, type, size, location, stage of development of the cysticerci, and the host's immune response against the parasite. The racemose variety occurs in the ventricles or basal cisterns and is a malignant form. Mobile ventricular mass can produce episodic hydrocephalus on changing head posture with attacks of headache, vomiting, and vertigo, triggered by abrupt movement of the head, a phenomenon called Bruns' syndrome (BS). We report a patient with racemose NCC and BS.

Keywords: Racemose neurocysticercosis. Cysticercotic meningitis. Bruns' syndrome.

RESUMO

A infecção por cisticercose é causada pelo estágio larval da *Taenia solium*. O parasita pode infectar o sistema nervoso central, causando neurocisticercose (NCC). As manifestações clínicas dependem da quantidade, tipo, tamanho, local, estágio de desenvolvimento do cisticerco e resposta imune do hospedeiro contra o parasita. A variedade racemosa ocorre nas cisternas ventriculares ou basais e é considerada uma forma maligna. O cisticerco móvel no ventrículo pode produzir hidrocefalia episódica com ataques de cefaléia, vômitos e vertigem, provocados pelo movimento abrupto da cabeça, fenômeno chamado de síndrome de Bruns (SB). Relataremos o caso de uma paciente com NCC racemosa com SB.

Palavras-chaves: Neurocisticercose racemosa. Meningite cisticercótica. Síndrome de Bruns.

INTRODUCTION

Cysticercosis is an infection caused by the larval stage of the tapeworm *Taenia solium*. The parasite may infect the central nervous system, causing neurocysticercosis (NCC). NCC is the single most common cause of acquired epileptic seizures in the developing world[1]. Despite the advances in diagnosis and therapy, NCC remains endemic in most low-income countries[2]. It is estimated that 50 million people in the world today are infected by the taeniasis-cysticercosis complex and 50,000 die every year[3].

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The clinical manifestations of NCC largely depend on load, type, size, location, and the stage of development of the cysticerci, as well as on the host's immune response against the parasite[4]. There is neither a pathognomonic feature nor a typical NCC syndrome.

The racemose variety occurs in the ventricles or basal cisterns and is characterized by abnormal growth of cystic membranes followed by degeneration of the scolex (parasite´s head). As ventricular and basal cisternal locations are considered malignant forms of NCC, when hydrocephalus secondary to cysticercotic meningitis is present, mortality rate is high (50%), and most patients die within 2 years of cerebrospinal fluid (CSF) shunting[5]. Attacks of severe headache, vomiting, and vertigo triggered by abrupt movement of the head due to a mobile ventricular mass producing episodic hydrocephalus on changing head posture happen. This unusual life-threatening phenomenon is called Bruns' syndrome (BS)[6-8].

In this paper, we report a patient with racemose NCC who presented symptoms compatible with Bruns' syndrome and an atypical cyst location, whose diagnosis was only possible through magnetic resonance imaging (MRI).

CASE REPORT

A 43-year-old female maid from the urban region of the south of Brazil was admitted to the Santa Casa emergency department in April 2008 presenting tonic-clonic seizure. She had no prior history of seizures. Over the previous 60 days, she had been presenting severe holocranial headaches with a tightening quality and progressive bilateral visual and hearing loss.

She denied any other signs or symptoms and reported no other possible trigger factor for a seizure than type II diabetes mellitus. The patient had a two-year history of direct contact with pulmonary tuberculosis at home.

When examined in the emergency room, she was disoriented with complete visual loss on the left eye and significant loss of visual accuracy on the right eye, as well as signs of meningeal irritation and bilateral papilledema. A brain computed tomography scan (CT) revealed hydrocephalus hypertensive tetraventricular. A lumbar puncture was performed with opening pressure of 750mmHg. The analysis of the CSF revealed a glucose concentration of 4mg/dl, a protein concentration of 72.8mg/dl, and 25 cells with 40% polymorphonuclear leukocytes and 40% lymphocytes. Results of the direct analysis and cultures for fungi and bacteria were negative. After 4 days, the patient was put on a combined treatment of rifampicin, isoniazid, and pyrazinamide (RHZ) plus dexamethasone. On the 14th day of hospitalization, a brain MRI was performed and

pointed to inflammatory granulomatosis and aracnoiditis and cystic lesions beside the pontine cistern, the interpeduncular cistern, and the bulbo-cerebellar cistern with hydrocephalus (**Figure 1A**).

Her symptoms did not improve throughout the RHZ treatment, and on the 21st day of hospitalization, a ventriculoperitoneal shunt (VPS) was performed. There was clinical improvement after the procedure. However, there were no changes in the CSF parameters.

Further investigation into the possible diagnosis of NCC was then initiated due to the findings on the MRI. A Weinberg reaction was, therefore, performed on the CSF, with **low reactors** result (1:2). The RHZ treatment was stopped and replaced by one with albendazole. The patient received albendazole for 15 days simultaneously with dexamethasone for 30 days.

Approximately 2 weeks after being discharged, the patient returned to the hospital with symptoms suggestive of VPS infection. Antibiotic treatment was prescribed along with the removal of the VPS, followed by the endoscopic third ventriculostomy. Twenty four hours after the procedure, the patient presented with intracranial hypertension. Subsequently, an external ventricular drainage was performed. A new VPS was placed later, and since then, the patient had been asymptomatic for 14 months.

In January 2010, the patient was admitted because she had complained of sudden vertigo and vomiting, associated with head rotation and daily headaches in the past 30 days prior to her admission. She then underwent brain MRI that showed intra-fourth ventricular cystic lesion and cystic lesion of the pineal region (**Figure 1B**). The patient underwent surgery for removal of the cysticercus (**Figures 1C** and **1D**). Anatomopathological examination confirmed the diagnosis of cysticercosis.

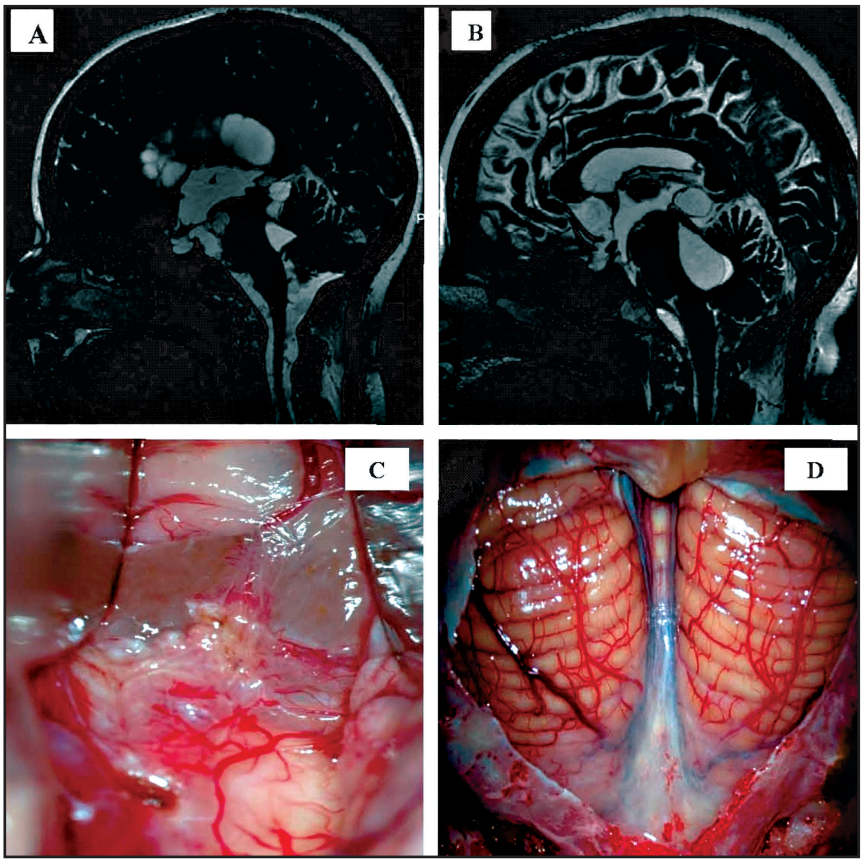

FIGURE 1: A - Sagital magnetic resonance Imaging (MRI) with a fast imaging employing steady state acquisition (FIESTA) acquisition demonstrating inflammatory granulomatosis, aracnoiditis and cystic lesions inside the lateral ventricles, third ventricle, pineal region and basal cisterns. B - T2 weighted sagital MRI showing a large cyst inside the fourth ventricle and in the pineal region and cysts in the basal cisterns. C, D - Surgical approach. In D the cerebellum with cisterna magna intact demonstrating aracnoiditis in this region. In C after opening of the cistern magna and spatulation of the cerebellum hemispheres in the middle line also showing aracnoiditis. In C the surgeon is approaching the forth ventricle.

DISCUSSION

Extraparenchmal disease varies in its symptoms and prognosis according to the location of the parasites. The prognosis for intraventricular neurocysticercosis is worse than that for the intraparenchymal forms of the disease[5]. Intracranial hypertension is a common manifestation and may be the result of a mass effect, distortion of the normal anatomy of CSF pathways, direct obstruction of the ventricular system by a cyst, or an inflammatory reaction in the meninges leading to arachnoiditis. Sometimes, like our patient, an intermittent or positional CSF obstruction with increasing intracranial pressure produces relapsing/remitting symptoms. The pathomechanism of the BS is not clear. Bruns believed the symptoms resulted from a change in the position of the cyst in the fourth ventricle with periodic blocking of the ventricular system on change of head posture and elevation of intracranial pressure due to a ball-valve mechanism[6,8]. The diagnosis of NCC is difficult because clinical manifestations are nonspecific. Most neuroimaging findings are non-pathognomonic, while some serologic tests have low specificity and sensitivity. A set of diagnostic criteria was proposed in 1996 and recently revisited to help with the diagnosis of NCC. Proper interpretation of these criteria permits two degrees of diagnostic certainty: definitive or probable[9]. Magnetic resonance imaging (MRI) is more accurate than CT for the diagnosis of most cases of NCC.

Therapeutic measures include antiparasitic drugs, surgery, and symptomatic medications. As inflammation is the conspicuous accompaniment in most forms of NCC, corticosteroids represent the primary form of therapy for meningitis, cysticercal encephalitis, and angiitis. Albendazole is considered the antiparasitic drug of choice for NCC because it has better penetration into the CSF and because its concentration is not affected when administered along with steroids[5,10].

Extra-parenchymal cysticercosis is associated with poor prognosis and requires a more aggressive approach. When feasible, complete surgical excision of lesions remains the definitive therapy[5-6]. In patients with hypertension, the priority is to manage the hypertension problem before considering any other form of therapy. In our case, this procedure was not an option due to the location of the lesions found in the first hospitalization but was the proposed treatment in the second hospitalization, as an abrupt permanent obstruction of the CSF flow can occur in patients with BS, which causes hydrocephalus leading to stupor, coma, and death due to brain herniation. The most common surgical indication for NCC is

ventricular shunting to resolve the hydrocephalus. Hydrocephalus secondary to NCC is associated with high rates of shunt dysfunction.

Neurocysticercosis is a longstanding disease that remains endemic in most low-income countries and has increasingly affected high-income countries due to increased migration, tourism, and travel to endemic areas. We report this case because NCC is a highly prevalent infectious disease, but is potentially eradicable, and is a public health issue that entails many social and economic consequences. Control programs are urgently needed to reduce the burden of this disease.

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