

Article

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Abstract: *Introduction:* Neurocysticercosis (NCC) is a common parasitic infection of the central nervous system caused by the larvae of the *Taenia solium*. Spinal cord involvement is very uncommon. *Clinical case:* A female patient with a history of NCC presented with chronic and recurrent headache associated with motor and sensory deficit, which develops tonic-clonic convulsion, with spatial disorientation. She also had intracranial hypertension syndrome, meningitis syndrome, and pyramidal signs suggestive of spinal NCC. *Conclusions:* Neurocysticercosis usually occurs in developing countries and should be considered as a differential diagnosis of neurological diseases. Early diagnosis and treatment are mandatory, as well as education to the community to primary prevention.

Key words: Neurocysticercosis, Spine, *Taenia solium*

Introduction

Cysticercosis is the most common parasitic disease affecting the Central Nervous System in under developing countries, caused by the larvae of the *Taenia solium*. Neurocysticercosis - NCC is unusual form of cysticercosis (1–6), which can be categorized according to its main

CNS involvement as: parenchymal, leptomeningal, intraventricular or spinal, according to the location of the lesions. Spinal NCC accounts for about 0.7 to 0.85% of all cases, and most of these are accompanied by concomitant intracranial involvement (2,7–9); it also may occur with extra-spinal involvement

(vertebral) or intraspinal (intradural, extramedullary or intramedullary) (4), being the extramedullary-intradural form the most common, with an incidence of almost 80% of the spinal forms (5).

Case

A female patient with a history of NCC diagnosis characterized by intense oppressive headache and periods of amnesic, which had received a peritoneal ventricle bypass system – SDVP - because of presenting obstructive hydrocephalus secondary to NCC. Prior to the shunt surgery, she developed an uncontrolled convulsive syndrome for the last 2 months.

One month later, she presented with a moderate to severe pulsatile headache, irradiated in the frontal region of the evening predominance, triggered by stress factors, which improved with the intake of NSAIDs. A week later, severe intensity headache (10/10 on the analogue pain scale), which was irradiated to the frontal region of the evening predominance, reappeared, accompanied by nausea, subjective vertigo, and an approximate weight loss of 5 kg. The patient presented progressive weakness in the left lower limb, which then became bilateral. It also refers to frequent falls with inversion of the right foot to ambulation. Following this, it presents paresthesias (numbness and cramps) with increased difficulty in walking and sensory deficit. At the initial neurological examination are found the mental functions preserved, the cranial nerves without alterations, strength, tone, trophism and normal sensitivity, neurophatic gait, muscular stretching reflexes +++ are symmetrical and generalized, without data of meningeal irritation. Cerebellar examination is normal. There are no

pathological or ataxic reflexes. After admission, she performs tonic-clonic convulsive crisis with disorientation in time and place, encountering endocranial hypertension syndrome, meningeal irritation syndrome and pyramidal syndrome. In addition, are the reflexes of Hoffman, Tromner and palmomenta.

In the initial CT scan, it was observed enlargement of the ventricular system and a mild right frontal cortical hypodensity, suggestive of NCC, An MRI of the lumbar and thoracic spine was performed, as well as CSF sample for cytochemical analysis. The glucose was 37 mg/dL, with an increase of proteins levels 49 mg/dL and increase of the number of cells at the expense of lymphocytes, with an ELISA test for diagnosis cysticercosis positive. Spinal myelography was performed, showing altered contrast suggestive of spinal cysticercosis (Figure 1). It was decided to start treatment with albendazole. Patient is managed on an outpatient basis by family decision.



Figure 1 - Spinal myelography showing contrast alteration suggestive of spinal cysticercosis, with many levels of stenosis due to the cysts

NCC is a common parasitic infection of the central nervous system - CNS, being the main cause of acquired epilepsy and other neurological morbidities in many areas of the world, especially in under developed countries (1,10). The peak of incidence is between the 3rd and 4th decade of life (11). This infection is common in Central and South America, most of Southeast Asia, part of China, many non-Muslim regional of Africa (10). The NCC was first described by Paranoli in 1550, and its causative agent, the *Taenia solium*, was recognized by Leuckart y Kuchenmeister in the 19th century (6,12,13). Spinal involvement is rare, and it has been subdivided in extradural, intradural-extramedullary and intramedullary, being the most rare form of presentation, accounting for <20% of all cases (1,4,7,14–16). Spinal NCC occurs frequently with a diagnosis of intracranial NCC, these cases are approximately 75%, however, isolated cases of spinal NCC are uncommon (6,11,16).

NCC transmission form is fecal-oral. The parasite is the cestode, *Taenia solium*, pigs are the intermediate host and humans are the definitive host (6,9). Life cycle starts in pigs, the larvae is encapsulated in muscles, subcutaneous tissues, liver, lungs, and brain, and then in humans starts with the ingestion of cysticercal eggs in food contaminated (6,14,17). In the stomach thanks to gastric acid the larvae is release from the eggs, and then it penetrate the intestinal mucosa and it goes to bloodstream, where is able to migrate to brain, muscle, and the eyes. Through small capillaries into the parenchyma or through the choroid plexus into the ventricles, the larvae is

disseminated to the subarachnoid space (6,14). Inside the human the larvae produce proglottids that contain many eggs, which are expulse in feces and may be ingested by intermediate hosts. Also humans can be infected by themselves (17).

NCC patients generally experience prolonged periods without symptoms and may present neurological manifestations such as local neurological deficit and seizures (17). In addition, they may present various symptoms such as generalized pain, radiculopathies, paresthesia, numbness, quadriparesis, frank equine tail syndrome, anatomical dysfunction of the intestine and bladder; radicular pain and progressive weakness of the extremities; the latter thow being the most common manifestations (4,14,18,19). In contrast, parenchymal NCC is characterized by epileptic convulsions, subarachnoid NCC manifestations with headache and intraventricular NCC sings as an acute or intermittent syndrome of intracranial hypertension (11).

These manifestations are produced by the cyst size, location and degeneration products (14,16). In addition, it triggers pathophysiological mechanisms such as mass effect, which causes compression of the spinal cord and produces myelopathy that causes progressive weakness in patients (20–22); And inflammatory reaction that may lead to the appearance of arachnoiditis, meningitis and obstruction of the subarachnoid pathways (4,14). The intact cyst causes less inflammatory reaction and is easy to surgical excision; on the other hand, degeneration of the cyst when parasite death occurs can cause

severe symptoms (4,16,23,24). The localization of these is more common at the thoracic level, but can occur in any region of the spine (4).

The extradural NCC presented in the lumbar region causes relatively slow symptoms, compared to the presence of an intramedullary NCC in the cervical region that causes a fast and early deterioration (16).

Three clinical stages of NCC have been defined: active, inactive, and transitory; and 4 pathological stages: vesicular, colloid vesicular, granular nodular and calcified nodular. The vesicular stage is the active form; The nodular and colloidal vesicular granules represent the transitory stage; And the calcified nodular pathological stage is an inactive stage of neurocysticercosis (11,25).

Differential diagnoses for spinal cystic lesions are spinal tumors, epidermoid tumor, echinococcosis, arachnoid / colloid cyst, hydatid cysts, tuberculosis, sarcoidosis, and meningocele. The diagnosis of NCC is made from neuroimaging, analysis of CSF and the detection of antibodies in serum (7,15,16). Several criteria have been proposed to facilitate its diagnosis; the major criteria include neuroimaging findings suggestive of NCC (cystic lesions or large calcifications), detection of anticystic antibodies by immunoblotting and resolution of cystic lesions after treatment with albendazole or praziquantel. Minor criteria include clinical manifestations suggestive of NCC, positive CSF ELISA for detection of anticystic antibodies or cysticercan antigens and presence of cysticercosis outside the CNS. Epidemiological criteria include residence or

travel to an endemic area or exposure to a *T. solium* carrier. The diagnosis is made with the presence of 2 major criteria plus 1 minor and 1 epidemiological (11,15,16).

The imaging study of choice for the detection of spinal lesions is MRI, however, it is difficult to detect small calcifications (5,7,15). MRI findings vary depending on the stages of the disease. The initial or vesicular stage is characterized by CSF isointense lesions that appear hypointense in T1 and hyperintense in T2, without surrounding edema. The colloidal vesicular stage is characterized by the presence of hypointense perilesional edema in T1 and hyperintense in T2, the cyst now appears as a ring, slightly hyperintense in T1 and hyperintense in T2. In the nodular granule stage, the capsule thickens and calcifications begin. In the calcified nodular stage, densely calcified cysts are difficult to visualize in MRI, unlike CAT scan where they are easily seen (4,14,16).

The CSF ELISA test is useful to confirm the diagnosis of NCC in patients with neurological signs, but in whom neuroimaging findings are inconclusive. The typical histopathological findings of NCC are the presence of dead or active translucent cysts with eosinophilic lining, usually surrounded by clear fluid and chronic inflammatory cells including neutrophils, eosinophils, and giant cells. The presence of calcified cysts can be observed in late and inactive stages (7,15,16). CSF examination may show low or normal levels of glucose, increased protein, lymphocytic pleocytosis, and eosinophilia (4).

For spinal NCC treatment there are decisive factors as activity of the disease and

location of parasites (11,15). A therapy with praziquantel and albendazole (at dose of 15mg/kg/day for 4 to 6 weeks) are efficient in neurologically stable patients (4,5,11,15–17). Antiparasitic treatment, due to the death of parasites, may lead to exacerbation of neurological symptoms (17,26). Surgical excision is the treatment of choice in patients with symptoms due to mass compression in the spinal cord, large cystic lesion or acute neurological deterioration during pharmacotherapy, in order to avoid irreversible neurological damage (4,9,11,15–17). Preoperative adjuvant treatment with albendazole allows to improve dissection planes during surgery (4,15). Steroid administration as well as extensive irrigation of the spinal canal helps to minimize the post-surgical inflammatory process (9,15,17,26). During the medical management a strict neurological monitoring is vital to avoid the acute neurological deterioration secondary to inflammatory response. To make sure the targeted lesion within the planned surgical field, the surgeon should consider recent scan (16).

To reduce NCC transmission the best way is by community education on endemic areas and those who wish to travel to areas where NCC prevails (26). Outcomes in patients with spinal NCC depend on cyst location, severity of inflammation, chronicity of symptoms and time of treatment (6,16). Intramedullary spinal NCC has a poorer outcome than patients with NCC-induced extrinsic spinal cord and cauda equine compression in the absence of severe arachnoidal scarring. A complete improvement can be achieved, however in

some cases the symptoms may be continuous or recurrent due to of arachnoid inflammation (6). Spinal intradural NCC is a rare variant of cysticercosis, which usually occurs in developing countries with poor practices in food preparation. The early detection of manifestations suggestive of spinal NCC allows the clinician an early diagnosis for a specialized management in the search to prevent permanent neurological complications. Disease promotion and prevention practices become an important tool for reducing the incidence of NCC, especially in endemic areas.

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