NEUROCYSTICERCOSIS IN ASPLENIC PATIENT, CASE REPORT

NEUROCISTICERCOSE EM PACIENTE ASPLÊNICO, RELATO DO CASO

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ABSTRACT

Neurocysticercosis is an important cause of neurologic and psychiatric disorders; it is a frequent etiology for acquired epilepsy worldwide. The parasitic infection of *Taenia solium* (including larval dissemination to the nervous system) can be avoided by effective means of prevention. Nonetheless, this disease remains endemic in many regions of the world.

To demonstrate the importance of prophylaxis this paper reports the case of a patient without spleen, who was treated for neurocysticercosis manifested by epilepsy. In twenty years of follow up, the patient did not experience a repeat occurrence of neurocysticercosis, despite of immunological impairment (absence of spleen) and environmental exposure (living in an endemic area).

Prevention was guided by a regular use of anthelmintic (Albendazole) and health education.

Key words: Neurocysticercosis, Splenectomy, Spleen, Epilepsy

RESUMO

Neurocisticercose é uma importante causa de doenças neurológicas e psiquiátricas, é uma frequente etiologia de epilepsia adquirida, no mundo. A infecção parasitária da *Taenia solium* (incluindo a disseminação das larvas para o sistema nervoso) pode ser evitada por meios eficazes de prevenção; no entanto, esta enfermidade ainda é endêmica em muitas regiões do mundo.

Para demonstrar a importância da profilaxia relata-se o caso de um paciente sem baço, o qual foi tratado para a neurocisticercose manifestada por epilepsia. Em vinte anos de seguimento, o paciente não repetiu a ocorrência de neurocisticercose, apesar de dano imunológico (ausência de baço) e exposição ambiental (habitação em área endêmica).

A prevenção foi guiada pelo uso regular de anti-helmíntico (Albendazole) e medidas educativas em saúde.

Palavras-chaves: Neurocisticercose, Esplenectomia, Baço, Epilepsia

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REVIEW

Neurocysticercosis is considered the leading cause of preventable neurological diseases in the world. Endemic areas include South and Central America, Asia, Sub-Saharan Africa, and India. It is also emerging in non-endemic areas due to migration and travelers, particularly in European countries, the United States of America, and Canada.

In Brazil, epidemiological studies consider the prevalence of the disease as underestimated, reporting incidence of 1.5% in autopsies and 3% in clinical trials.[1]

Case reports of neurocysticercosis in the international literature are more common than in Brazil. However, Brazilian publications add both quality and rarity to the body of literature. For example:

a) Rare presentations: Bruns syndrome, i.e., episodic hydrocephalus with cyst movement in brain ventricles,[2] frontotemporal dementia with mutism,[3] spastic paraplegia,[4] seizures and psychiatric manifestations of temporal lobe,[5] trigeminal neuralgia,[6] Lennox-Gastaut syndrome,[7] and presented with giant multiple cysts.[8] [9]

b) Atypical locations of cysts: brainstem (occurring in 8% of cases),[10] intramedullary,[11] (occurring in 1.2 to 5.8% of cases).[12]

Although appropriate prophylaxis for neurocysticercosis exists, the disease remains as a common cause of epilepsy and is also associated with a variety of sensory and motor deficits. Estimated 7.6 million cases of epilepsy are associated with neurocysticercosis, considering the world population.[13]

In studies of prophylaxis the use of 400 mg Albendazole for three consecutive days, 2 times a year, has been demonstrated to produce significant reduction of taeniasis (and other helminthiasis) when administered to participants who are over six years old, who are not pregnant or breastfeeding, and who do not suffer from acute diseases.[14]

Taenia solium eggs can be transmitted between humans. Such interpersonal transmission can lead to neurocysticercosis.

Animals, particularly pigs, are intermediate hosts, because they harbor the parasite in its larval forms. Humans are the definitive hosts, because they house the *Taenia solium* in its adult form. The adult worm is able to disseminate thousands of fertilized eggs daily.

The eggs can be ingested in water or on contaminated food. The eggs hatch in the human intestine after exposure to bile and pancreatic juice. The larvae (*metacestodae*) become mature in the digestive tract, causing taeniasis. The larvae can also spread throughout the bloodstream, causing cysticercosis in various organs such as eyes, brain, muscles, subcutaneous tissue, etc. Another route of contamination is by ingestion of undercooked pork containing the living larvae.[15]

Contamination can be prevented by thorough cooking of meat, sanitation of fresh foods (such as fruits, vegetables…), washing of hands, as well as by animal's health inspection.

The immune system, in order to combat the *metacestodae*, recruits the participation of several cells (eosinophils, lymphocytes, monocytes, plasma cells, macrophages...) and chemical mediators (lymphokines, interferons, tumor necrosis factor...).[16]

In the brain, the formation of granulomas around the cysts illustrates the importance of these cellular mechanisms in combating neurocysticercosis. Granulomas are formed by macrophages and their derived cells (giant cells, epithelioid). Other defensive mechanisms include the perivascular mononuclear infiltration, the fibroblast multiplication, and the microglia proliferation. The phagocytic capacity of macrophages and microglial cells highlights the participation of reticuloendothelial system (mononuclear phagocyte system) in the defense against neurocysticercosis. [17]

The spleen is the largest of the lymph organs and part of the reticuloendothelial system. Therefore, it forms an integral part in the prevention of neurocysticercosis infections.

Spleen is more efficient than liver in the removal of non-opsonized bacteria and encapsulated organisms. Encapsulated pathogens such as *Streptococcus pneumoniae*, *Neisseria meningitidis*, or *Haemophilus influenzae* type B can cause severe infections in post-splenectomy patients. The spleen not only purifies blood from fungi, viruses, and foreign bodies but also participates in hematopoiesis and immunoglobulin synthesis.[18] [19]

The literature proposes that the absence of spleen weakens the mononuclear phagocyte system. This evidence suggests that asplenia may hinder successful prevention of long-term neurocysticercosis recurrence in spenectomized patients and inhabitants of endemic areas.

OBJECTIVE

The objective of this paper is to report both the follow-up of a patient with neurocysticercosis and asplenia

as well as to describe the prophylaxis against helminthiasis used in this case.

METHODOLOGY

In this case report, we present the evolution of neurocysticercosis in a splenectomized patient, observing the patient twice a year for over 20 years.

CASE REPORT

The patient was referred to neurology department of Our Lady of Conception Hospital in Porto Alegre, Brazil, to investigate epilepsy. The patient was 24 years old, male, with professional activities in pig farming, from a rural area, where several cases of neurocysticercosis have occurred.

The history of the patient revealed seizures beginning at 21 years of age. The crises were characterized as tonic-clonic and occurred on a monthly basis; phenobarbital could not control the seizures. Episodes of holocranial headache emerged at the same time as the seizures.

The patient reported splenectomy surgery at 11 years of age, due to a physical trauma caused by a fall from height. Other conditions such as drug addiction, AIDS, or head trauma were excluded. General clinical and laboratory examinations revealed no abnormalities. Neurological examination found no motor or sensory impairments. Electroencephalography showed bursts of sharp and polyspike waves generalized with asymmetric amplitude.

Computed tomography (CT) of the head documented multiple cystic lesions forming a "Swiss cheese pattern", which refers to the significant amount of disseminated cysts in the brain parenchyma. The multiple lesions had ovoid shapes with dimensions of approximately 5-10 mm, filled with liquid. Within a few cysts, visible scolices were present with a characteristic "comma" shape and altered intensity compared to the vesicular fluid. The cysts were in various stages of evolution, predominantly in stage I (minimal tissue reactions or absent) and II (with perilesional edema). CT findings characterized the neurocysticercosis lesions.

Based on the revised diagnostic criteria,[20] the case met: a) an *absolute* diagnostic criterion; b) a *major* criterion; c) as well as *minor* and *epidemiological* criteria (Table 1). This set established definitive diagnosis.

Table 1. Diagnostic criteria

Criteria	Presentation in the patient
Absolute	**Visualization of the scolex by CT**
Major	**Resolution of the cystic lesions after therapy with Albendazole**
Minor	**Clinic manifestation of neurocysticercosis (seizures and headache)**
Epidemiologic	**Origin from endemic area**

We treated parenchymatous neurocysticercosis with: a) use of antiparasitic drugs, b) management of seizures, and c) management of cerebral edema. The patient was hospitalized at the hospital of origin; we prescribed Albendazole 5 mg/kg/ day for 4 weeks; and Phenobarbital 100 mg /day continuous, and Dexamethasone 12 mg/day for 1 week; in consensus with the treatment recommendations for neurocysticercosis.[21]

The treatment maintained the initial Phenobarbital monotherapy because the cause of the uncontrolled seizures lied in the brain edema controlled by corticosteroid.

After treatment, seizures had a complete remission. The follow-up CT, after one year of treatment, showed regression of lesions to calcification, reduction in the diameter of the cysts from 5 to 1 mm on average, perilesional resolution of edema, absence of ventricular dilatation, or subsequent gliosis.

DISCUSSION

Patient's origin from an endemic area and the professional activity in swine farming were significant in the case reported; however, neurocysticercoses are known to occur also in in non-endemic areas among migrants and travelers.

The patient's prior splenectomy has led to prophy-

lactic care that included the use of 400 mg Albendazole every 6 months. Also, other means of prophylaxis were included such as thorough cooking of pork, evaluating the origin of fruits and vegetables, and proper hygiene and diseases prevention practices.[22]

In the following 20 years, the patient did not develop subsequent neurocysticercosis.

An interesting finding was the occurrence of another parasitic disease, the ganglionic form of toxoplasmosis, twenty years after neurocysticercosis hospitalization.

The literature documents in mice experiments that the *Taenia* species inhibits certain defense mechanisms of spleen cells. Infection by *Taenia taeniaeformis* decreases the ability of spleen cells to respond to the mitotic factor concavaline A.[23] Similar effect occurs in toxoplasmosis. *Toxoplasma gondii* infections in mice inhibit mitotic effect of concavaline A on T cells in the spleen;[24] toxoplasma decreases also the production of interferon gamma and other cytokines in spleen cells.[25] Similarities between these two parasitic infections highlight the importance of macrophagic defence mechanisms and need for prophylaxis and adequate follow-up in asplenic individuals.

CONCLUSIONS

Treatment and prophylaxis of neurocysticercosis were effective, despite of the splenectomy factor.

This case report of neurocysticercosis is associated with asplenia, which brings attention to the role of mononuclear phagocyte system in the defenses against neurocysticercosis and toxoplasmosis.

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REFERENCES

1. Agapejev S. Clinical and epidemiological aspects of neurocysticercosis in Brazil: a critical approach. *Arq. neuropsiquiatr* 2003; *61*(3B), 822-828.
2. Rodriquez, RD, Crestani DNDS, Soares JOD, Franceshini PR, Alves RP, Zimerman, R, Ferreira N, Barea LM. Bruns’ syndrome and racemose neurocysticercosis: a case report. *Revista da Sociedade Brasileira de Medicina Tropical* 2012; *45*(2), 269-271.
3. Satler C, Maestro ES, Tomaz, C. Frontotemporal dementia and neurocysticercosis: a case report. *Dement. neuropsychol* 2012; *6*(1).
4. Holanda LFD, Pereira BJA, Holanda RR, Silva Filho APD, Segundo L, da Costa J, Oliveira P, Holanda, GN. Paraplegia espástica causada por cisticercose intramedular: apresentação de caso e breve revisão bibliográfica. *Arq. bras. neurocir* 2010; *29*(4).
5. Costa FADO, Fabião OM, Schmidt FDO, Fontes AT. Neurocysticercosis of the left temporal lobe with epileptic and psychiatric manifestations: case report. *Journal of Epilepsy and Clinical Neurophysiology* 2007; *13*(4), 183-185.
6. Aguiar PH Miura FK, Napoli PR. Sendenski M, Rotta JM, Cescato VA, Teixeira MJ, Marino Junior R. Neuralgia do trigêmeo bilateral por cisticerco racemoso unilateral no ângulo-ponto cerebelar: relato de caso. *Arq. neuropsiquiatr* 2000: *58*(4), 1138-41.
7. Agapejev S, Padula NAR, Morales NMO, Lima MMF. Neurocysticercosis and Lennox-Gastaut syndrome: case report. *Arq. neuropsiquiatr* 2000: *58*(2B), 538-547.
8. Mattana PR, Michelon AT, Bender A, Dillenbur, GA, Michielin JH, Michalski L, Ortigara M, Mantovani, R. Neurosisticercose múltipla gigante: apresentação de caso. *Rev. cient. AMECS* 1997: *6*(1), 96-101.
9. Zerati E, Góes MJ, Nascimbeni, MA. Cisto gigante neurocisticercótico: relato de caso. *Arq. bras. neurocir* 1994: *13*(1), 46-8.
10. Wanderley FLER, Lelis LC, Sander JC, Barbosa E, Wilson FJ, and Bastos CAG. Neurocisticercose em tronco cerebral: relato de caso e revisão da literatura. *Arq. bras. neurocir* 2013; *32*(4).
11. Araujo A B S, Cambraia M B R, Azevedo R, Filho M M, Rezende G L, & Vanderlei A S. Spinal intramedullary cysticercosis: a case report and literature review. Arq. Bras. Neurocir 2013; v. 32, n. 4.
12. Yamashita S, Mesquita M, Machado G, Miranda AH, Morceli, J. Cisticercose intramedular: relato de caso e revisão de literatura. *Radiol. Bras* 2003; *36*, 255-257.
13. Berkowitz AL, Raibagkar P, Pritt BS, Mateen FJ. Neurologic manifestations of the neglected tropical diseases. *Journal of the neurological sciences* 2015; *349*(1), 20-32.
14. Ash A, Okello A, Khamlome B, Inthavong P, Allen J, Thompson R A. Controlling Taenia solium and soil transmitted helminthes in a northern Lao PDR village: Impact of a triple dose albendazole regime. *Acta tropica* 2015.
15. Del Brutto OH, García HH. Taenia solium Cysticercosis—The lessons of history. *Journal of the neurological sciences* 2015; *359*(1), 392-395.
16. Sciutto E, Cárdenas G, Adalid-Peralta L, Fragoso G, Larralde C, Fleury A. Human neurocysticercosis: immunological features involved in the host’s susceptibility to become infected and to develop disease. *Microbes and Infection* 2013; *15*(6), 524-530.
17. Dametto E. Histopathology of the Human Brain in Neurocysticercosis. J Mol Histol Med Physiol 2016; 1: 106.
18. Browning MG, Bullen N, Nokes T, Tucke, K, Coleman, M. The evolving indications for splenectomy. *British Journal of Haematology* 2016; 1-3.
19. De Matos Filho AS, Petroianu A. Spleen function after preservation in a physiological solution. *Journal of Surgical Research* 2015; *199*(2), 586-591.
20. Del Brutto OH, Rajshekhar V, White Jr AC, Tsang VCW, Nash TE, Takayanagui OM, Schantz PM, Evans CAW, Flisser A, Correa D, Botero D, Allan JC, Sarti E, Gonzalez AE, Gilman RH, García HH. Proposed diagnostic criteria for neurocysticercosis. *Neurology* 2001; *57*(2): 177-183.
21. García HH, Evans CA, Nash TE, Takayanagui OM, White AC, Botero D, Flisser A. Current consensus guidelines for treatment of neurocysticercosis. *Clin Microbiol Rev* 2002; *15*(4): 747-756.
22. Mascarini-Serra L. Prevention of soil-transmitted helminth infection. *J Glob Infect Dis* 2011; *3*(2): 175.
23. Letonja T, Hammerberg C, Schurig G. Evaluation of spleen lymphocyte responsiveness to a T-cell mitogen during early infection with larval Taenia taeniaeformis. *Parasitol Res* 1987; *73*(3): 265-270.
24. Neyer LE, Kang H, Remington JS, Suzuki Y. Mesenteric lymph node T cells but not splenic T cells maintain their proliferative response to Concanavalin - A following peroral infection with Toxoplasma gondii. *Parasite Immunol* 1998; *20*(12): 573-581.
25. Candolfi E, Hunter CA, Remington JS. Roles of gamma interferon and other cytokines in suppression of the spleen cell proliferative response to concanavalin A and toxoplasma antigen during acute toxoplasmosis. *Infect Immun* 1995; *63*(3): 751-756.