

INTRAMEDULLARY SPINAL CYSTICERCOSIS SIMULATING A CONUS MEDULLARIS TUMOR

Case report

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ABSTRACT - Cysticercosis is an endemic condition in many developing countries. Although it is the most common parasitic disease of the central nervous system, cysticercal involvement of the spinal cord is rare. It may occur as intradural extramedullary, intramedullary, intramedullary associated with intradural-extramedullary or as the vertebral presentation. We report the case of a 53-year-old woman who presented with low back pain of acute onset and no other symptoms. Magnetic resonance imaging (MRI) showed an intramedullary cyst of the conus medullaris region which, at pathological examination, was diagnosed as a cysticercal cyst. She refused anticysticercal agents and steroids postoperatively. After an eight-year follow-up, the patient performs the activities of her daily living with no difficulties, and annual spinal MRIs show no residual signs of the disease. Clinical, pathophysiological, diagnostic and therapeutic aspects of spinal cord intramedullary cysticercosis are discussed.

KEY WORDS: conus medullaris, cysticercosis, intramedullary cysticercosis, spinal cord.

Cisticercose intramedular simulando tumor do cone medular: relato de caso

RESUMO - Cisticercose é uma doença endêmica em vários países em desenvolvimento. Embora seja a doença parasitária mais freqüente do sistema nervoso central, o acometimento medular por cisticercos é raro. Pode ocorrer nas formas intradural extramedular, intramedular isolada, intramedular em associação com intradural extramedular, além da forma vertebral. Relatamos o caso de mulher de 53 anos de idade que se apresentou com dor lombar de início agudo, sem outros sintomas. Ressonância magnética (RM) identificou imagem cística na região do cone medular que, no estudo histopatológico, foi diagnosticada como cisticercose. A paciente recusou tratamento pós-operatório com anti-helmínticos e corticosteróides. Após oito anos de seguimento, a paciente exerce suas atividades cotidianas sem dificuldades, e estudos de RM anuais não mostram sinais de doença residual. Aspectos clínicos, fisiopatológicos, diagnósticos e terapêuticos da cisticercose intramedular são discutidos.

PALAVRAS-CHAVE: cone medular, cisticercose, cisticercose intramedular, medula espinhal.

Cysticercosis is the infection caused by *Cysticercus cellulosae*, the larvae of the tapeworm *Taenia solium*, which affects humans mainly by accidental ingestion of eggs containing infective oncospheres¹⁻⁴. It is the most common parasitic disease of the central nervous system^{1-3,5-13}, and its related mortality rates range from 6 to 50%¹². It is an endemic condition to Brazil^{8,12}, Peru¹⁴, Mexico^{1,12}, Korea, India^{11,12} and other South American, Tropical African, and Southeast Asian countries^{1,6}. Its frequency in developed countries

is increasing, as migration rates increase from endemic areas^{3,6,12,15}. Cysticercal involvement of the spinal cord is rare^{1,4-8,11,15-21}, even in endemic areas¹, and accounts for 0.7 to 5.85% of all cases^{2,8,10-13,15,19,21-23}. Its prevalence may be underestimated, since brain cysticercosis, which is a more common condition, frequently occurs concomitantly^{5,8,15,18,21,24}, spinal canal is not routinely examined in necropsies^{1,8,20,21,24} and small asymptomatic cysts may be easily overlooked¹. Intradural extramedullary (subarachnoid) cysticercosis

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sis is more prevalent than intramedullary cysticercosis (IC)^{5,6,11,20,21,23} and than the association of intramedullary and intradural-extramedullary presentations (54% vs. 17% vs. 17%, respectively)²⁵. Extradural location is even more rare^{1,5,6,8,15,16,20,21,25}. The vertebral type has also been reported^{2,22}.

We report the case of a 53-year-old woman who presented with low back pain of acute onset and no other symptoms. Magnetic resonance imaging (MRI) revealed an intraparenchymal cyst of the conus medullaris region which, at pathological examination, was diagnosed as a cysticercal cyst. Based on a brief review of the literature, clinical, pathophysiological, diagnostic and therapeutic aspects of IC are discussed.

CASE

An otherwise healthy 53-year-old woman wakes up in the early morning with intense incapacitating low back pain with lower limbs irradiation. No predisposing condition for cysticercal infection was present, and she is resident in a non-endemic region of Rio de Janeiro, at the Sou-

theast of Brazil. At the Emergency Department she presented afebrile with stable vital signs and normal mental status. General, ophthalmologic and neurologic examinations, routine blood, urine analysis, plain x-ray films of her lumbosacral spinal cord and chest were normal. There were no signs of subcutaneous nodules, nor meningeal irritation. As pain relieved after non-steroidal anti-inflammatory drugs administration, she was discharged for ambulatory investigation and follow-up. Twelve hours after hospital discharge, she returned with recurrence of the symptoms. Again, no abnormality was noted on physical and laboratory examinations.

The patient underwent lumbosacral spine MRI, which showed a cystic lesion in the conus medullaris. On sagittal T1-weighted image (Fig 1), cyst fluid was isointense to cerebrospinal fluid (CSF) and there was a hyper- to isointense nodule attached to the interior of the cyst wall. Further images were not available for publication. As definitive diagnosis was missing, pain rapidly worsened and did not resolve with progressive more potent analgesic agents, an ordinary T11 to T12 laminectomy was performed 36 hours after the first episode. Dural sac was tough and tense (Fig 2A). After longitudinal dural opening at posterior midline, an enlarged



Fig 1. Sagittal T1-weighted MRI of the spine showing an intramedullary well delimiting cyst at the conus medullaris region (arrow). Cyst fluid is isointense to CSF and the scolex is visualized as a hyper- to isointense nodule attached to the interior of the cyst wall.

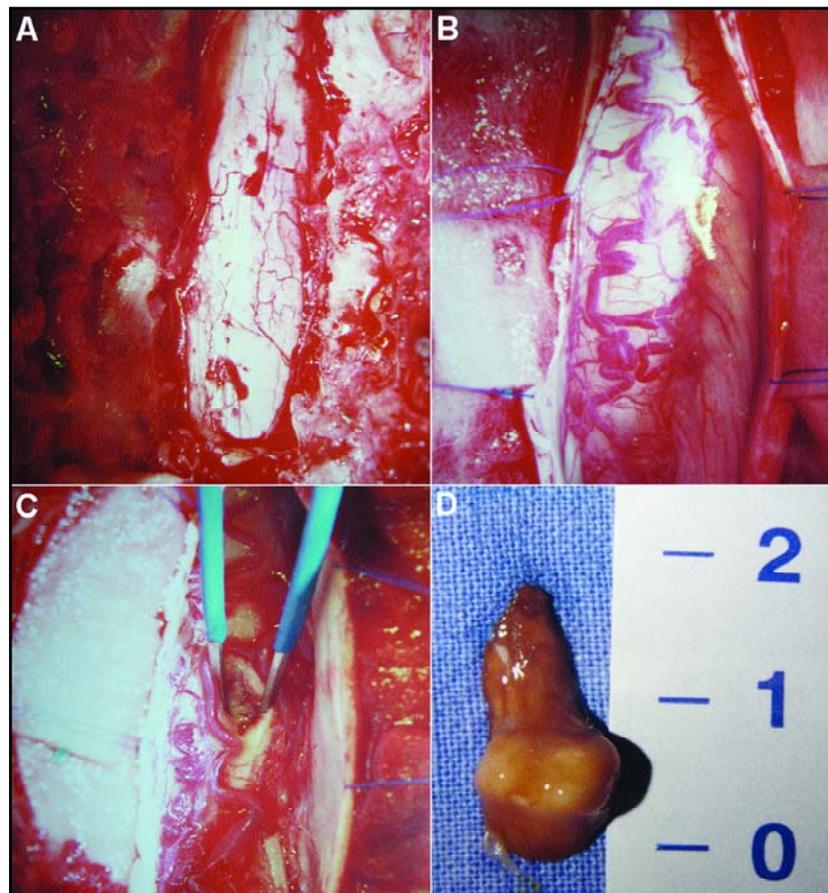


Fig 2. Surgical excision of the intramedullary cyst. A) The tough and tense dural sac. B) The enlarged spinal cord after longitudinal dural opening at posterior midline. C) Mielotomy at the posterior median sulcus showing the intramedullary parasite. D) Photograph of the parasite at the operating room.

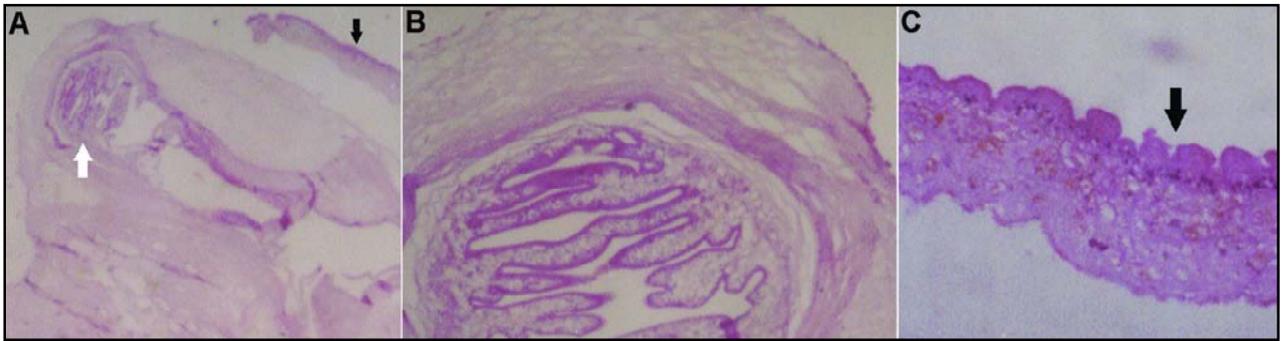


Fig 3. Photomicrographs of the histopathological specimen. A) The scolex (white arrow) and a portion of the cyst wall (black arrow). H&E, original magnification X 40. B) A close view of the scolex. H&E, original magnification X 100. C) Membrane of the cyst showing the outer layer covered with microvilli (arrow). H&E, original magnification X 400.

spinal cord was visualized (Fig 2B). Five milliliters of a xanthochromic fluid were then aspirated with puncture at spinal posterior midline, leaving some fluid inside the cyst. Fluid examination did not reveal malignant cells. Under microscope vision and with microsurgical techniques, myelotomy was made at the posterior median sulcus (Fig 2C), followed by extirpation of the cyst (Fig 2D). Histological findings at the operating room showed a non-tumoral, inflammatory process. Postoperative histological diagnosis of cysticercosis was made by means of hematoxylin and eosin-stained samples of the surgical specimen (Fig 3).

There was no evidence of cysticercal infection elsewhere. The patient, a mathematics teacher married to a doctor and mother of two other ones, refused postoperative treatment with anticysticercal agents and steroids. After an eight-year follow-up, she presents with hypesthesia over the S1 dermatome and absent Achilles tendon reflex bilaterally, preservation of anal sphincter and bladder function, no motor deficit and no difficulty on deambulation, without compromise of the activities of her daily living. Annual spinal MRIs show no residual signs of the disease.

DISCUSSION

Intramedullary cysticercal involvement, usually solitary^{2,23}, most probably occurs through arterial blood circulation^{2,5,8,9,11,12,15,16,18,20,21,24,25}. The site of infection could be proportional to regional blood flow^{2,8,10-12,15,16,20-22}, and this would help explain why the most common region of IC is thoracic, followed by cervical, lumbar and sacral regions^{2,8,12,15,20,22,23,25}. The cysticercus can cause direct mass effect, induce regional or distant inflammatory reaction, and medullar degeneration due to meningitis or vascular compression and insufficiency^{5,8,10,12,14,16,26}. Inflammatory reaction against the dead parasite is associated with perilesional edema, which can damage medullar parenchyma^{15,18} and, therefore, worsen symptoms and predict a poorer outcome¹. We believe that mass effect, inflammation and, mainly, ischemia of the sacral re-

gion, an area of low blood flow, would explain the onset of symptoms in the present case.

Most reported cases of IC ranged between 20-45 years of age, and symptoms duration varied from one week to 10 years^{10,12}. Common symptoms include pain, para- or quadriparesis, spasticity, bowel and bladder incontinence⁸ and sexual dysfunction. They can be accompanied by a variety of sensory deficits^{15,23}. These symptoms may be secondary to mass effect lesions, and their occurrence, as happened in the present case, should not be primarily attributed to IC. This can be the cause of delayed diagnosis, mainly in non-endemic areas, unless there is evidence of concomitant cysticercal infection elsewhere. Our patient presented with pain of acute onset and progressive worsening, without other signs and symptoms of neurologic disease. As she is resident in a non-endemic area of the country, has good educational and economical status and no evident risk factors for cysticercosis, the presumptive diagnosis was an intramedullary tumor of the conus.

Differential diagnosis of an intramedullary cystic lesion in a patient presenting with back pain is extensive and includes neoplastic^{2,12,15,18,19}, infectious (e.g. abscess)²⁷, inflammatory (e.g. multiple sclerosis), post-traumatic spinal cord changes (e.g. syringomyelic cavitation)^{12,15} and parasitic infestations (e.g. IC and hydatidic cysts)¹⁵. MRI can identify characteristic features of IC⁹, and this would be of great diagnostic importance. It is the preferred image method for intramedullary lesions visualization¹ and correlates with cysticercal pathological stages (vesicular, colloidal vesicular, granular nodular and calcific nodule)¹⁰, but visualization of the scolex is sometimes not possible. Although a nodule was present inside the cyst, clinical and epidemiological history did not lead us to IC as the primary diagnostic hypothesis in the present case.

The optimal treatment for IC remains controversial²³. As surgical excision can give definitive diagnosis and alleviate compressive symptoms^{5,6,12}, it is recommended by the majority of authors as the treatment of choice^{5,6,10,12,13,18,20}. As cysts usually have a superficial location (3 mm deep or less)¹², adhere weakly to medullar parenchyma even in the degenerative stage⁵ and their walls are made of a dense layer of fibrous tissue^{4,10,15}, total resection is possible in most cases^{6,17}. In our case, aspiration of part of the cyst fluid was necessary in order to reduce its tension, preventing the medullary cavity from closing above a completely evacuated cyst. Then, microsurgical techniques and gentle manipulation made total resection possible.

According to the American Society for Microbiology Current Consensus Guidelines for Treatment of Neurocysticercosis³, the treatment of spinal cysticercosis, intra- or extramedullary, is primarily surgical (evidence III: opinions of respected authorities, based on clinical experience; descriptive studies and case reports; or reports of expert committees). Some authors state that postoperative treatment with anticyclicercal agents would be warranted, since cysticercosis is a generalized disease with focal symptoms^{1,11,12,22}. Our patient was operated on in an emergency basis, 36 hours after the onset of the symptoms and, despite postoperative anticyclicercal and steroids refusal, her postoperative deficits were non-significant.

Total resection, nevertheless, is not always feasible. Adhesions to nervous tissue and vessels can make intramedullary degenerating cysts resection technically difficult in some cases^{1,5,6,10}. Besides, while surgery has high related mortality and morbidity rates (15% and 85%, respectively)^{2,12,18,28}, good results after 8- and 30-day regimens of albendazole (15 mg/kg/day) and praziquantel (50 mg/kg/day) have been reported^{9,18,23,27}. It seems that albendazole is more effective for IC than praziquantel, as it is for cerebral cysticercosis¹⁸. Dexamethasone is also used because it increases albendazole blood levels and attenuates treatment-associated inflammatory reactions²³.

IC is a rare condition, mainly at the sacral region, and the difficulties in making a definitive diagnosis are greater in non-endemic areas. Once spread to medullar parenchyma, most probably by arterial embolization, cysticercus may cause mass effect, inflammatory reaction and medullar degeneration. The majority of symptoms are non-specific, and MRI is the preferred diagnostic method. The optimal treatment for IC (surgical exeresis versus anticyclicercal drugs) remains controversial. High surgery-related

mortality and morbidity have been reported, while medical regimens have achieved good results in recently reported cases. Clinical studies on this issue are still lacking.

REFERENCES

- Alsina GA, Johnson JP, McBride DQ, Rhoten PRL, Mehringer CM, Stokes JK. Spinal neurocysticercosis. *Neurosurg Focus* 2002;12:1-7.
- Dantas FLR, Fagundes-Pereyra WJ, Souza CT, Veja MG, Souza AA. Intramedullary cysticercosis: case report. *Arq Neuropsiquiatr* 1999;57:301-305.
- García HH, Evans CAW, Nash TE, et al. Current consensus guidelines for treatment of neurocysticercosis. *Clin Microbiol Rev* 2002;15:747-756.
- Hesket KT. Cysticercosis of the dorsal cord. *J Neurol Neurosurg Psychiatry* 1965;28:445-448.
- Colli BO, Assirati JA Jr, Machado HR, Santos F, Takayanagui OM. Cysticercosis of the central nervous system: II. Spinal cysticercosis. *Arq Neuropsiquiatr* 1994;52:187-199.
- Colli BO, Valença MM, Carlotti Jr CG, Machado HR, Assirati JA Jr. Spinal cord cysticercosis: neurosurgical aspects. *Neurosurg Focus* 2002;12:1-7.
- Egberts JH, van der Horst C, Bannowsky A, Junemann KP, Braun PM. Micturition dysfunction triggered by spinal intramedullary neurocysticercosis. *Aktuelle Urol* 2004;35:58-61.
- Gallani NR, Zambelli HJL, Roth-Vargas AA, Limoli C Jr. Cysticercosis of the spinal cord: report of two cases, literature review and comments on pathogenesis. *Arq Neuropsiquiatr* 1992;50:343-350.
- Gaur V, Gupta RK, Dev R, Kathuria MK, Husain M. MR imaging of intramedullary spinal cysticercosis: a report of two cases. *Clin Radiol* 2000;89:311-314.
- Mathuriya SN, Khosla VK, Vasishtha RK, Tewari MK, Pathak A, Prabhakar S. Intramedullary cysticercosis: MRI diagnosis. *Neurol India* 2001;49:71-74.
- Mohanty A, Venkatrama SK, Das S, Das BS, Rao BR, Vasudev MK. Spinal intramedullary cysticercosis. *Neurosurgery* 1997;40:82-87.
- Sheehan JP, Sheehan J, Lopes MB, Jane JA. Intramedullary spinal cysticercosis: case report and review of the literature. *Neurosurg Focus* 2002;12:1-4.
- Singh NN, Verma R, Pankaj BK, Misra S. Cauda-conus syndrome resulting from neurocysticercosis. *Neurol India* 2003;51:118-120.
- Trelles JO, Caceres A, Palomino L. La cysticercose médullaire. *Rev Neurol (Paris)* 1970;123:187-202.
- Leite CC, Jinkins JR, Escobar BE, et al. MR imaging of intramedullary and intradural-extramedullary spinal cysticercosis. *Am J Roentgenol* 1997;169:1713-1717.
- Akiguchi I, Fujiwara T, Matsuyama H, Muranaka H, Kameyama M. Intramedullary spinal cysticercosis. *Neurology* 1979;29:1531-1534.
- Barini O. Cisticercos macrocístico intramedular: extração cirúrgica. *Arq Neuropsiquiatr* 1954;12:264-266.
- Corral I, Quereda C, Moreno A, et al. Intramedullary cysticercosis cured with drug treatment: a case report. *Spine* 1996;21:2284-2287.
- Vázquez MAS, Carachure IJ, Maltos URC, Herrera FS. Cisticercosis intramedular. *Arch Neuroci* 2001;6:36-38.
- Siqueira MC, Koury LS, Boer CAA, Rezende-Filho CP. Cisticercos solitário intramedular: relato de caso e revisão da literatura. *Arq Bras Neurocir* 1987;38:131-139.
- Canelas HM, Ricciardi-Cruz O, Escalante OAD. Cysticercosis of the nervous system: less frequent clinical forms; III - Spinal cord forms. *Arq Neuropsiquiatr* 1963;21(2):77-86.
- Singh P, Sahai K. Intramedullary cysticercosis. *Neurol India* 2004; 52:264-265.
- Torabi AM, Quiceno M, Mendelsohn DB, Powell CM. Multilevel intramedullary spinal neurocysticercosis with eosinophilic meningitis. *Arch Neurol* 2004;61:770-772.
- Sperlescu A, Balbo RJ, Rossitti SL. Brief comments on the pathogenesis of spinal cysticercosis. *Arq Neuropsiquiatr* 1989;47:105-109.
- Amaral L, Maschietto M, Maschietto R, et al. Unusual manifestations of neurocysticercosis in MR imaging: analysis of 172 cases. *Arq Neuropsiquiatr* 2003;61:533-541.
- Yamashita S, Mesquita MVG, Machado JCM, Miranda AH, Morceli J. Intramedullary spinal cysticercosis: a case report and review of the literature. *Radiol Bras* 2003;36:255-257.
- Robertson HJ, Watson J. Case 4: Neurocysticercosis with cervical meningeal involvement. *Am J Roentgenol* 1998;171:879-880.
- Holtzman RNM, Hughes JEO, Sachdev RK, Jarenwattananon A. Intramedullary cysticercosis. *Surg Neurol* 1986;26:187-191.