INTRADURAL-EXTRAMEDULLARY SPINAL CYSTICERCOSIS WITH BRAIN INVOLVEMENT: A CASE REPORT AND LITERATURE REVIEW*

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Abstract **Report on a rare case of infestation by cysticercosis involving the cerebral subarachnoid space and the epidural space in the cervical and thoracic regions of a 59-year old female patient who presented nausea, signs of cerebelar ataxia and gradual loss of the sensibility in both legs. The diagnosis was based on magnetic resonance imaging of brain and cervical-thoracic spine demonstrating the presence of cysts in the subarachnoid spaces. An enzyme-linked immunosorbent assay (ELISA) performed on cerebrospinal fluid resulted positive and demonstrated a protein level of 420 mg/dl indicating the disease activity. Parasites were surgically removed for the necessity of thoracic spinal cord decompression. Based on findings of literature review, brief comments were made on the pathogenesis of the intradural-extramedullary spinal cysticercosis cystic form, magnetic resonance imaging findings and treatment.**

Keywords: Cysticercosis; Brain; Spine; Magnetic resonance imaging.

Resumo *Cisticercose intradural-extramedular cerebral e espinhal: relato de caso e revisão da literatura.*

Relato de um caso raro de apresentação de infestação por cisticercose do espaço subaracnóide cerebral e intra-raquiano nas regiões cervical e torácica em mulher de 59 anos de idade, com náuseas, sinais de ataxia cerebelar e perda gradual da sensibilidade nas pernas. O diagnóstico foi feito por meio de imagens por ressonância magnética do cérebro e da coluna cérvico-torácica, que evidenciaram a presença de cistos nos espaços subaracnóides. O exame do líquido cefalorraquiano revelou teste imunológico ELISA positivo e elevado nível de proteína (420 mg/dl), indicativo de atividade da doença. Os parasitos foram removidos cirurgicamente pela necessidade de descompressão da medula espinhal torácica. Breve comentário sobre a patogênese da forma cística da cisticercose espinhal intradural-extramedular, aspectos das imagens de ressonância magnética e tratamento foram feitos com base nos achados de revisão da literatura.

Unitermos: Cisticercose; Cérebro; Coluna; Ressonância magnética.

INTRODUCTION

Human cysticercosis is the most common parasitic disease to affect the central nervous system, caused by the larval stage of *Taenia solium* (cysticercus), especially in countries where the sanitary infrastructure is deficient, particularly in Latin America, Africa and Asia and yet, with relative frequency, in Portugal, Spain and European Eastern countries and endemic in developed countries with high rates of immigration from endemic areas[1–3].

The adult worm develops in humans after ingestion of pork, rare, infected meat containing encysted larvae. The human cysticercosis is acquired through oral-fecal pathway after contact with member of the family or community with the adult worm, as well as by auto-infestation in patient with the intestinal adult worm. After ova ingestion, the shells of ova are digested and the larvae develops and lodge in soft-tissues, primarily in brain, spinal cord, heart/extra-ocular/skeletal muscle, eyeball, fat tissue and skin[1–3].

Cerebral and spinal-medullary intraparenchymatous cysticercosis is caused by cysts and/or granulomatous, inflammatory nodules, whereas extraparenchymatous cysticercosis like those located inside periencephalic liquoric spaces (including the subarachnoid space in the region of cortical sulci and encephalic cisterns), cerebral ventricles and intradural-extramedullary space of the vertebral column, is typically cystic.

The case report includes clinical-radiological-pathologic aspects of cerebral and spinal intradural-extramedullary cysticercosis in cystic form, with a brief literature review.

CASE REPORT

A 59-year old, white woman was admitted for evaluation, presenting nausea, cerebelar ataxia, a gradual loss of sensibility in both legs, micturition and evacuation disorders. At general physical and neurological examination decreased reflexes in lower extremities and hyperesthesia at the T6 dermatome where characterized.

Magnetic resonance imaging (MRI) has revealed cystic lesions in brain, cervical-thoracic spine subarachnoid spaces with signs of compression on the thoracic spinal cord (Figures 1 and 2).

Laboratory tests on cerebrospinal fluid collected by means of lumbar puncture have indicated a high protein level (420 mg/dl) and the research of antibodies for cysticercus, ELISA test, was positive.

Because of the cysts compressive effect on the thoracic spinal cord, surgical treatment was indicated, which has evidenced several cystic lesions in subarachnoid spaces immediately after dura mater incision, such lesion being removed (Figure 3).

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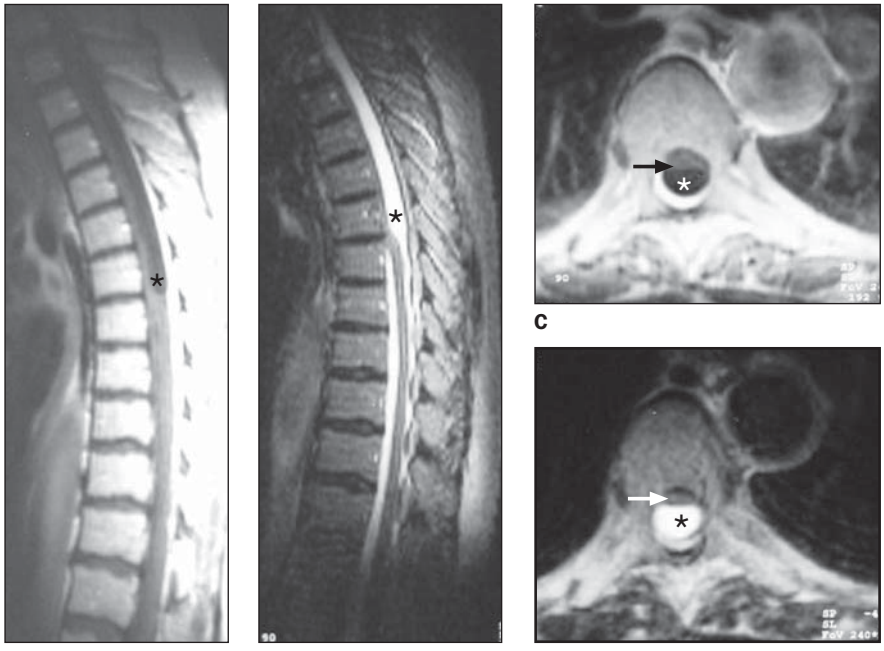


Figure 1. Dorsal column MRI. **A:** Sagittal plane, T1-weighted sequence showing clustered cysts (asterisk) corresponding to an area with hyposignal. **B:** Sagittal plane, T2-weighted sequence showing hyperintense clustered cysts (asterisk) compressing the dorsal spinal medulla. **C:** Axial plane, T1-weighted sequence showing hypointense clustered cysts (asterisk). **D:** Axial plane, T2-weighted sequence showing hyperintense clustered cysts (asterisk) compressing the dorsal spinal medulla (arrows).

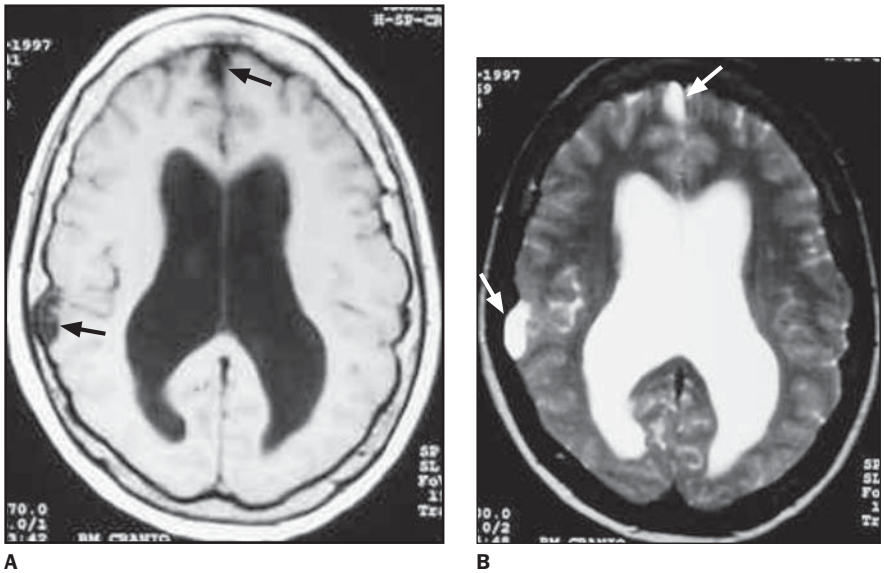


Figure 2. Axial brain MRI. **A:** T1-weighted showing hypointense cysts (arrows) in subarachnoid space in parietal and right frontal regions. **B:** T2-weighted sequence at the same level, showing hyperintense cysts (arrows).

DISCUSSION

A case of vesicles in the corpus callosum was first described by Paranoli in 1550. Paracelso correlated cerebral vesicles and convulsive attacks in 1650, while Ridi and Malpighi characterized these parasitic lesions in 1686. The first reference to intra-rachidian cysticercosis is attributed to Rokitansky in 1856[4].

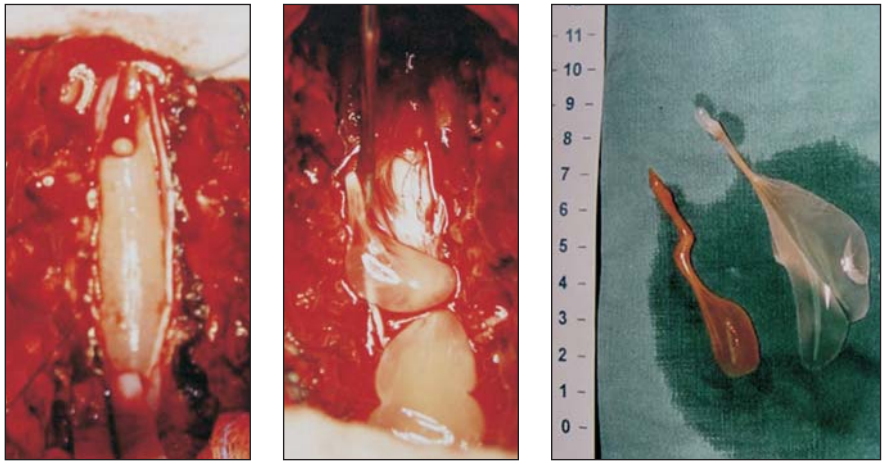
The neurocysticercosis intra-rachidian form is quite rare, the intradural-extramedullary presentation being the most frequent and the intramedullary, the less frequent. Among 205 inpatients at Hospital Vera Cruz de Campinas, SP, Neurology Service, in the period between 1961 and 1987, only 18 (9.0%) had a definite diagnosis of neurocysticercosis, and only three (1.4%) presented intradural-extramedullary spinal infestation by cysticercosis[5].

In the literature, we have found an excellent review study analyzing 95 cases of spinal cysticercosis published since 1865 (Chart 1), 61 (66.0%) of them presenting intradural-extramedullary localization, and 33 (34.0%) presenting intramedullary localization[4]. Many studies included in this review and other more recent studies[1,3,6], likewise the present case, have shown the simultaneous involvement of liquoric, intradural-extramedullary spinal spaces in different segments and cerebral spaces.

The cysticerci larvae dissemination into the intradural-extramedullary spinal space has been explained by the accumulation of a great volume of blood originating from valveless epidural venous plexus, with extremely thin-walled veins, which may conduct the blood towards any direction, under the influence of intra-abdominal and intrathoracic pressure variations. Two surgically-confirmed cases of primary cysticerci larvae dissemination into the intradural-extramedullary spinal canal without cerebral involvement corroborate the hypothesis of retrograde flow of those larvae through the veins of the above mentioned epidural venous plexus, differently from the Isamat de La Riva's postulate on the parasites descending migration from the cerebral subarachnoid space into the spinal space[7].

The term racemose cysticercosis (*cysticercus racemosus*) has been utilized by several authors and characterizes the development of large translucent vesicles, with big tails, multiloculated or branched off, absent scolex, subarachnoid space-occupying[1,3,5,6,8].

The intra-rachidian intradural-extramedullary cysticercosis may be asymptomatic or presenting few symptoms over long periods of time. Symptoms may vary, depending on the vertebral column segment (cervical, dorsal, lumbar or sacral) were cysts are lodged[1–3,6]. When the cyst is



A B C

Figure 3. A: Surgical aspect following dura mater incision in the dorsal region, exposing clustered cysticercotic cysts. **B:** Surgical sequence showing cysts individual removal. **C:** Macroscopic aspect of two removed cysts.

localized in the spine cervical-thoracic segment, the symptoms are more apparent and usually cause medullary compression syndrome, as described in this case: gradual loss of sensibility in both legs, hyperestesia at the corresponding dermatome and micturition and evacuation physiological disorders. However, in the spine lumbosacral segment, frequently the clinical picture evidences localized radiculopathy that may be confounded with discopathy[1–3,6].

Before the MRI advent, computed mielotomography, notwithstanding its invasive nature, was the imaging method employed for investigating lesions producing medullary and/or radicular compression symptoms. This procedure showed intra-rachidian, round, hypodense lesions

Chart 1 Intradural-extramedullary spinal cysticercosis: published cases.

Author	Year	Country	Age	Sex	Brain*	Diagnosis	Localization
Westphal	1865	?	?	?	?	?	Cauda equina
Richter	1891	?	?	?	+	Necropsy	C4/T10
Minor	1899	?	?	?	+	Necropsy	Thoracic
Hartman	1902	Germany	?	?	?	Lumbar puncture	?
Wollenberg	1905	?	?	?	+	Necropsy	Cervical-lumbar
		?	?	?	+	Necropsy	Thoracic-lumbar
Sterz	1910	Germany	?	?	?	Lumbar puncture	?
Vasiliu	1921	Italy	?	?	?	?	?
Redalie	1921	France	?	?	?	?	Cervical-thoracic
Castex	1926	Argentina	25-year-old	F	+	Lumbar puncture	?
Verga	1926	Italy	?	?	+	Necropsy	?
Morawieka	1927	?	?	?	+	Lumbar puncture	Cauda equina
Bertrand	1945	France	?	?	?	?	Lumbar
Loyo	1955	Mexico	?	?	?	?	Cauda equina
Pennybaker	1956	?	?	?	+	Surgery	Cervical
Fracassi	1956	Argentina	49-year-old	M	+	Lumbar puncture	?
De la Riva	1957	Spain	?	?	+	Necropsy	Lumbar
			?	?	+	Lumbar puncture	Lumbar
Rocca	1959	Peru	(15 cases)			?	Cervical (13), thoracic (1), lumbar (1)
Cabieses	1959	Peru	12-year-old	M	–	Surgery	T3
Cruz	1961	Brazil	42-year-old	M	–	Surgery	T11/L2 e L2 L3
Calzado	1960	Mexico	?	?	?	?	Thoracic
			26-year-old	M	–	Surgery	T6/T9
Canelas	1963	Brazil	32-year-old	F	+	Surgery	T11/L2
			42-year-old	M	+	Surgery	T12/L2
			27-year-old	M	+	Surgery	T5/T9
Staimle	1964	Mexico	57-year-old	M	–	Surgery	Cauda equina
Absalon	1965	Mexico	45-year-old	M	–	Surgery	T8/T9
			35-year-old	M	–	Surgery	C5/C7
			63-year-old	M	–	Surgery	C2/C4
			40-year-old	M	+	Surgery	L1/L2
Alanis	1967	Mexico	57-year-old	F	?	?	L4/L5
Trelles	1968	Peru	56-year-old	M	+	Necropsy	C4
			46--year-old	M	+	Necropsy	C4
			58-year-old	F	+	Necropsy	Cauda equina
Castano	1969	Colombia	34-year-old	F	–	Surgery	Cauda equina
Parker	1988	France	35-year-old	M	+	Surgery	L5/S2
			46-year-old	F	–	Surgery	L5/S1
Bandres	1992	USA	34-year-old	M	–	Lumbar puncture	C2/S1
Leite	1997	Brazil, USA, Colombia	(9 cases)			Lumbar puncture, surgery	Cervical-thoracic (4), cervical-thoracic-lumbar (4), cervical (2), thoracic-lumbar (1), lumbar (1)
Çiftçi	1999	USA	30-year-old	F	+	Lumbar puncture	C2/C4

* Coexistence of cysts in liquoric cerebral and spinal spaces.

marginated by intrathecally-injected iodine contrast[2,5,6].

Presently, the imaging method of choice for intradural-extramedullary cysticercosis diagnosis is the MRI, that accurately demonstrates cystic lesions in subarachnoid liquoric spaces of periencephalic or spinal regions and, principally because is a non-invasive procedure[1,3,8,9].

MRI images indicate the precise localization and contours of these cysts whose signal intensity is similar to CSF, hyposignal in T1-weighted sequences and hypersignal in T2-weighted sequences[1,3,8,9], identical to those demonstrated in this patient studies.

However, these characteristics may differ when cysts present inflammatory alterations, resulting in hyperintense images both in T1-weighted and T2-weighted sequences, as a consequence of the high protein content[3]. After paramagnetic contrast agent endovenous infusion, both the cyst and the meninges surrounding the adjacent medulla may be enhanced (arachnoiditis)[1–3,8], which has not occurred in this case report.

A differential diagnosis spectrum should be considered when cystic, intrarachidian lesions are detected in MRI images: congenital lesions (simple or complex arachnoid cysts, dermoid cysts), cysts of other parasitary etiology (hydatid), tuberculosis, sarcoidosis or subarachnoid metastatic neoplasm[1,3].

A positive ELISA serological test and high rates of protein in CSF may be of help in the diagnosis, indicate the disease activity and serve as a follow-up and control of medicamentous treatment[6]. In the present case, CSF laboratory tests reveal high protein levels (420 mg/dl) and positive for ELISA.

Usually, treatment is conservative, presenting good results with administration of cysticidal drugs like praziquantel or albendazole, the later demonstrating higher treatment efficacy[8,9]. In cases where signs of medullary and/or radicular compression are detected, surgical indication becomes necessary, likewise in the case of our patient. An interesting aspect of the decompressive surgery in this case was the photographic register of a careful and individual surgical removal of cysts, allowing the radiological-surgical-pathological correlation.

Concluding, cysticercosis is a severe global public health problem, including developed countries because of the high rates of immigration from endemic areas. The disease severity is determined when cystic cysticerci involve subarachnoid spaces, producing medullary symptoms when located in the cervico-thoracic segment and radicular symptoms, when located in the lumbosacral segment, demanding surgery due to frequent unsuccessful conservative clinical treatments. The diagnosis may be completed by a findings combination: epidemiological history, classical clinical signs and symptoms, MRI images, laboratory tests and CSF serological tests.

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