Intramedullary spinal cysticercosis

Case report and review of the literature

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Cysticercosis is the most common parasitic infection of the central nervous system. It infrequently affects the spine, but when it does, it can present with symptoms similar to other more common spinal diseases. The authors present a case of isolated intramedullary cysticercosis of the cervical spine and review the literature.

KEY WORDS • cervical spine • cysticercosis • intramedullary lesion

Cysticercosis is the most common parasitic disease to affect the human central nervous system.^{1,7} Neurocysticercosis occurs through one of the following mechanisms: ingestion of water or food contaminated by the ova of the cestode *Taenia solium*; inadvertent autoinfection with adult *T. solium* worms via an anus-to-finger-to-mouth route; or regurgitation of ova from the lower to upper small intestine and stomach where the shells of ova are digested and embryos or oncospheres are liberated.^{7,8} The embryos burrow into the gastric or intestinal wall and reach the circulation. These embryos may be deposited almost anywhere in the human body. In approximately 2 months' time, the embryos form the encysted larval stage termed *Cysticercus cellulosae*.

Most often the brain is affected and is involved in 60 to 92% of all patients with cysticercosis.^{7,13} The cysticerci tend to accumulate at the base of the brain in multicystic or grapelike structures in a form called *C. racemose*.^{5,10} In only rare cases is the spinal cord involved.^{2,7,12,14} Other common locations for cysticercosis include subcutaneous tissue (17.8%), skeletal or heart muscle (8.5%), and eye (5.1%).^{7,14} More rare sites include liver, lung, kidney, pancreas, intestine, diaphragm, spleen, peritoneum, parotid gland, mouth, and peripheral nervous system.^{1,7,8} Presenting symptomatology depends on the number of parasites, their location(s), duration of infection, and the host's immune response.

We describe a 16-year-old right-handed young woman who presented to the University of Virginia Health Sciences Center with complaints of progressive bilateral hand numbness following a skiing accident approximately 4 months previously. She was found to harbor an isolated cervical C1–2 intramedullary spinal cysticercosis. The presentation, diagnosis, and treatment are detailed, and we provide a review of the literature regarding intramedullary spinal cysticercosis.

CASE REPORT

Presentation and Examination. This 16-year-old young woman suffered injuries in a skiing accident. Over the following 4 months, she experienced progressive bilateral hand paresthesias. Thereafter, the patient experienced periods characterized by a decreased respiratory rate. At the time of her presentation, her neurological status was significant only for slightly decreased sensation to light touch in her hands bilaterally (worse on the left), but these deficits did not conform to particular dermatomal distributions. Other sensory modalities (pinprick, temperature, and proprioception) remained intact.

Upon arrival, the patient underwent emergency head and cervical spine CT scanning performed with and without contrast medium. Multiple contiguous axial CT scans were obtained from C2–3 to the vertex at 5-mm intervals with and without intravenous contrast. The brain parenchyma, ventricles, sulci, and cisterns were shown to be free of lesions. A low density lesion, however, was noted

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computerized tomography; MR = magnetic resonance.

within the spinal cord at C1–2; this lesion demonstrated minimal mass effect and mild contrast enhancement (Fig. 1 *upper left*).

To visualize the cervical cord better, MR imaging was performed. We observed an intraparenchymal lesion at C1–2. The cord was focally enlarged at this level, and signs of edematous change were demonstrated. The lesion appeared cystic in nature, measured 1.5 cm in widest diameter, and had a thin rim of enhancement (Fig. 1 *upper right* and *lower*). The content of the lesion appeared isointense with CSF on T₂-weighted MR images.

Operation. Surgery was believed to be warranted because of the patient's progressive neurological symptoms and the worrisome location of the lesion (that is, at C1–2). In the operating room, somatosensory evoked potential monitoring was conducted; no paralytic agents were given so as to allow the patient to breathe spontaneously throughout the procedure. A standard C-1 laminectomy was performed, and the underlying dura mater was

Fig. 1. Contrast-enhanced neuroimaging studies. *Upper Left:* Axial CT scan revealing a low density lesion at C1–2 with minimal mass effect and mild contrast enhancement. *Upper Right:* Axial MR image revealing a cystic lesion at C1–2 with irregular contrast enhancement of the lesion's periphery. *Lower:* Sagittal MR image demonstrating the cysticercosis cyst at C1–2 with irregular contrast enhancement at the periphery (*arrow*).

opened. An expansile nodule over the posterior surface coincided with a cystic structure on intraoperative ultrasonography. A myelotomy was made, and the lesion was immediately identified. A cleavage plane between the spinal cord and lesion was defined. A large mucoid lobule was extirpated, and circumferentially the cyst wall was then removed. These specimens were sent for neuropathological analysis. The dura was closed primarily. The patient maintained regular spontaneous ventilation throughout the operation; somatosensory evoked potential monitoring demonstrated no changes.

Neuropathological Examination. Analysis showed cyst wall remnants from intramedullary cysticercosis (Fig. 2 *upper*). The wall consisted of a cuticular layer with brush-like border and an inner reticular layer demonstrating degenerative changes (Fig. 2 *lower*). The *T. solium* scolex could not be identified. The remaining specimens showed dense fibrotic capsules with accompanying lymphocytes and macrophages. Examination of spinal cord samples demonstrated reactive gliosis.

Postoperative Course. Postoperatively, the patient experienced mild but improved hand numbness bilaterally. Steroid therapy was slowly tapered, and the patient received a 2-week course of praziquantel. Postoperative anticysticercal treatment is generally recommended because cysticercosis is considered to be a generalized disease with focal symptoms.¹¹ Mohanty, et al.,¹¹ treated 50% of their patients with a course of anticysticercal therapy even after the cyst was surgically removed. Praziquantel or albendazole are the most commonly used pharmacological agents to treat neurocysticercosis.^{1,9,11}

The patient underwent full craniospinal axis MR imaging evaluation, which demonstrated no evidence of other cysticercosis lesions. She was discharged to home on postoperative Day 2.

DISCUSSION

Despite being the most common parasitic entity affecting the central nervous system, neurocysticercosis is a rare clinical entity in the United States. The condition is more endemic to Brazil, Peru, Mexico, Korea, and India.^{9,15} Our patient lived just outside of Washington, D.C. She adhered to a Kosher diet and denied consuming pork. Ten years previously, she had briefly traveled to Mexico.

Neurocysticercosis-related mortality rates range from 6 to 50%.^{1,13,17} Spinal involvement occurs in only 0.7 to 5.85% of patients afflicted with the disease, and the low incidence of spinal involvement is believed to be related to the fact that the blood flow to the brain is approximately 100-fold greater than to the spine.^{2,11,13} In fact, the locations of spinal lesions appear to be proportional to regional spinal cord blood flow. As such, De Souza Queiroz, et al.,² estimated that spinal distribution of cysticerci occurs as follows: 34% in the cervical; 44.5% in the thoracic; 15.5% in the lumbar; and 6% in the sacral region. In an older study² patients afflicted with spinal cysticercosis, the authors reported that 30% also had intracerebral involvement and 25% had intramuscular involvement, whereas in more a more recent study⁶ the authors noted 100% concurrent intracerebral involvement.

The incubation period for cysticercosis is usually less

Fig. 2. Photomicrographs. *Upper:* Section of remnants of a cysticercosis cyst showing degenerative changes. The scolex could not be found in the specimen. The cyst is composed of a thick integument and an outer surface covered with microvilli. *Lower:* Tegumental cells displaying degenerative changes in the inner layer of the cyst wall. H & E, original magnification $\times 40$ (*upper*) and $\times 200$ (*lower*).

than 10 years, and most larvae survive for only 5 years before dying.^{8,16} In cases of spinal cysticercosis, most patients exhibit symptoms lasting between 7 days and 7 years before the disease is diagnosed; there is a report of one patient who experienced symptoms for more than 10 years before appropriate care was rendered.¹⁵ Common symptoms included spastic paraplegia, sphincter disturbances, and pain.^{2,15} In the two most recent series, all patients with spinal intramedullary cysticercosis had definite sensory levels and experienced significant motor symptoms including paraparesis, quadriparesis, or paraplegia.^{3,11} Subacute symptoms are likely a result of mass effect and inflammation whereas acute worsening may result from meningitis or vascular insufficiency. These neurological symptoms could just as easily be manifestations from a spinal cord tumor. In the literature, most patients were male and between age 20 and 40 years at presentation.¹⁵ Our case is atypical in that the patient was a young female who exhibited no motor symptoms, pain, or sphincter dysfunction and in whom no distinct sensory

level was evident. Additionally, she had not been in an endemic area for more than 10 years, a period that is thought to be the extent of the incubation for cysticercosis.

Laboratory and neuroimaging evaluation can be helpful in establishing the diagnosis.¹ Magnetic resonance imaging of intact spinal intramedullary lesions typically demonstrates cystic areas within the parenchyma and cyst fluid similar to that of CSF on both T₁- and T₂-weighted images.⁶ A subtle hypointensity may appear at the rim of the cyst on T₂-weighted sequences. Infrequently, the scolex can be visualized on T₁-weighted images as a mural nodule isointense to cord parenchyma and is located in the cyst itself.¹² Irregular areas of peripheral enhancement after intravenously administered gadolinium have also been observed. With CT myelography, partial to complete myelographic blockage and focal cord widening are common over one to two vertebral segments; additionally soft-tissue calcification may be observed.^{11,15} Spinal intramedullary cysts usually range in size from 0.5 to 1 cm in diameter.¹¹ The imaging findings for cystic intramedullary disease must include in the differential diagnosis other parasitic diseases, neoplasia with primary or secondary cysts, and posttraumatic spinal cord changes.^{6,8} Once a diagnosis of cysticercosis had been made, the entire neuraxis should be evaluated and serial images obtained to assess treatment effectiveness. In the present case, an isolated intramedullary cystic lesion was demonstrated at C1–2. Magnetic resonance imaging revealed no evidence of cranial cysticercosis. The absence of cranial cysticercosis is in contrast to findings in the only series in which MR imaging was used to perform complete evaluation of 16 patients; in this series the authors found concomitant intracranial cysticercal lesions in all 16 patients.⁶ Serial craniospinal MR imaging is planned for our patient over the next 12 months.

When a patient does not come from an endemic area and spinal cord symptoms are the first manifestations of the disease, the diagnosis may only become apparent at the time of surgery. It is of greater importance to determine a tentative diagnosis without or prior to surgery because of the high surgery-related morbidity (85%), associated mortality rate of the disease, the potential earlier detection with neuroimaging evaluation, and the increasing effectiveness of medical management.^{4,9,14,17} The role of medical treatment is increasing in the management of neurocysticercosis. Albendazole (15 mg/kg/day) and praziquantel (50 mg/kg/day) regimens have been used for both 8- and 30-day periods.^{1,4,9,10}

Medical management is often an adjunct to surgery. The effectiveness of cysticidal therapy does not obviate the need for resection of symptomatic intraparenchymal lesions. Current therapy for leptomeningeal spinal cysticercosis is laminectomy and resection followed by medical management. In cases of the intramedullary form of spinal cysticercosis, resection is usually required to obtain a definitive diagnosis as well as to ameliorate some neurological deficits related to mass effect of the cyst.^{4,8} In fact, with most cases of intramedullary spinal cysticercosis, histopathological evaluation was the method used to achieve a correct diagnosis of the cysticercal nature of the lesion.^{2,4,6,11,15} Intramedullary spinal cysticercosis cysts tend to be located superficially within the parenchyma (that is, < 3 mm deep), and this was in fact the case in our

patient.¹¹ After performing a superficial myelotomy, a cleavage plane can usually be developed with ease, thereby separating the cyst from the adjacent, albeit reactive, cord parenchyma.

Surgery-related results of surgery for intramedullary spinal cysticercosis are mixed. Mohanty, et al.,¹¹ have reported that seven of eight patients improved neurologically following surgery, and one patient, who had presented with paraplegia, was even able to perform her activities of daily living on follow-up examination.¹¹ Outcomes reported in other series have not been as favorable. For instance, of the 20 patients treated surgically by Sharma, et al.,¹⁵ 12 improved neurologically to some degree, five did not improve, and three died; a variety of motor, sensory, and urological deficits were demonstrated in all surviving patients. In a similar fashion, De Souza Queiroz, et al.,² noted that of 11 surgically treated patients, complete or partial recovery was observed in eight, no change was observed in one, and two died. Sharma, et al.,¹⁵ hypothesized that sensory and urological improvements likely were a result of decreased mass effect following decompressive surgery, but the refractoriness of motor deficits might have been a result of parenchymal gliosis from toxic parasitic metabolites. Nevertheless, the rate of neurological improvement resulting from decompressive surgery alone in those afflicted with intramedullary spinal cysticercosis was 60 to 75% in these three series.^{2,11,15} Our patient was no exception: she experienced no further apneic spells and suffered only mild residual sensory deficits. Furthermore, she has been able to return to school and continues to participate in sports.

CONCLUSIONS

Intramedullary spinal cysticercosis is a rare clinical entity and is frequently associated with intracranial involvement. Patients tend to present with motor, sensory, and urological deficits not unlike those observed in patients with other parasitic, neoplastic, and posttraumatic origins. Preoperative evaluation can include hematological and CSF testing as well as neuroimaging evaluation, preferably MR imaging. Suspicion should also be heightened in patients who have traveled to endemic areas in the preceding 10 years. Definitive diagnosis and amelioration of neurological sequelae can generally be achieved by resecting the intramedullary spinal cysts. Medical treatment and serial craniospinal neuroimaging should be undertaken after the diagnosis has been made.

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