Isolated Primary Intradural Extramedullary Spinal Neurocysticercosis: A Case Report and Review of Literature

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Abstract-

Background: In spite of being the most common parasitic infestation of central nervous system (CNS), spinal cysticercosis remains a rare entity.

Case Report: We report an unusual case of a 45-year-old-male with primary isolated localization of spinal intradural extramedullary cysticercosis at thoracic 3/4 level. The lesion was surgically addressed to decompress the cord in combination with administration of oral albendazole. The weakness improved after treatment but the pain and numbness persisted. The available treatment options, diagnostic strategies and the pathophysiology of this rare condition are discussed here with a brief review of literature.

Conclusions: Clinicians should be aware of the diagnostic possibility of such a rare pathology. Neurosurgeons may face surgical challenges due to dense arachnoiditis associated with the degenerating lesion which may also account for the incomplete resolution of the symptoms even after treatment.

Key Words: Neurocysticercosis, Spine, Primary, Isolated, Intradural, Extramedullary

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INTRODUCTION

Human cysticercosis is a systemic infestation caused by Cysticercus cellulosae, the larval form of Taenia solium. Pigs are the intermediate while humans are definite (or occasionally accidental intermediate) host[(1)]. It is the most common parasitic infection affecting the CNS[(1)]. Neurocysticercosis (NCC) typically involves the brain parenchyma, intracranial subarachnoid space or ventricular system and is often self-limiting[(1)]. Spinal NCC, even in endemic regions, is rare with a reported incidence of around 1-3%[(2,3)]. Most of the cases described in world literature have a concomitant cranial involvement, suggested as a rationale for entire neuraxis evaluation[(2,4)]. The current case presents a unique example of isolated primary cysticercosis of spine, without an evidence of cranial involvement.

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CASE REPORT

History and examination

A 45-year-old man otherwise fit and well, presented with mid-back pain of three months duration, radiating along a right fifth intercostal space, exacerbated by coughing. He subsequently developed progressive difficulty while walking with no evidence of bladder and bowel dysfunction. Physical examination revealed muscle power of MRC grade 4/5 in right lower limb, thoracic-7 sensory level and lower extremity hyperreflexia with a spastic gait. Other sensations including joint position and vibration were intact. The Babinski sign was present on the right side with a normal down going big toe on the contralateral side. Magnetic resonance imaging (MRI) study revealed multiple intradural extramedullary cystic lesions displacing the spinal cord anteriorly and to left at thoracic 3/4 level (Fig. 1). The lesions were well-circumscribed and located in spinal subarachnoid space with signal intensity similar to cerebrospinal fluid (CSF), low in T1 weighted (Fig. 2) while high in T2 weighted sequence, measuring approximately 2.5 to 3.0 cm in diameter (Fig. 3). The cysts showed mild enhancement on gadolinium administration (Fig. 3). MRI of the brain was normal. A complete blood count revealed the presence of eosinophilia showing >8% eosinophils in peripheral blood with an absolute count of >1300 mm^3. Other blood counts were within normal limits. The results of CSF analysis, obtained by a lumber puncture, showed an increased eosinophil count (>30 cells) and high proteins (>1.8 g/l). A serum ELISA test was positive for the presence of anti-Taenia solium antibodies of IgG and IgM type.

Operation

The patient subsequently underwent a thoracic 3/4 laminectomy and resection of the mass lesion. Dura was opened in midline under operating microscope and dense arachnoid scarring was seen around the lesion. Majority of the lesion was cystic with a thin and friable cyst wall.

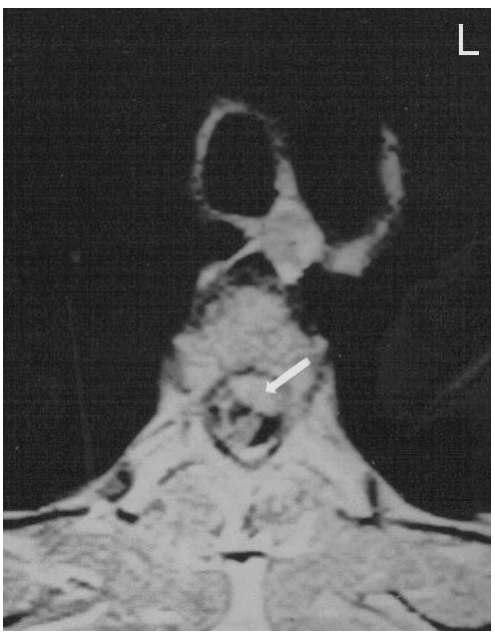

Figure 1. T1-weighted image showing transverse section at the level of thoracic 3/4. A multiloculated cystic lesion of variable intensity measuring approximately 2.5 to 3.0 cm in diameter can be seen in the posterolateral location pushing the cord to left and anteriorly (arrow).

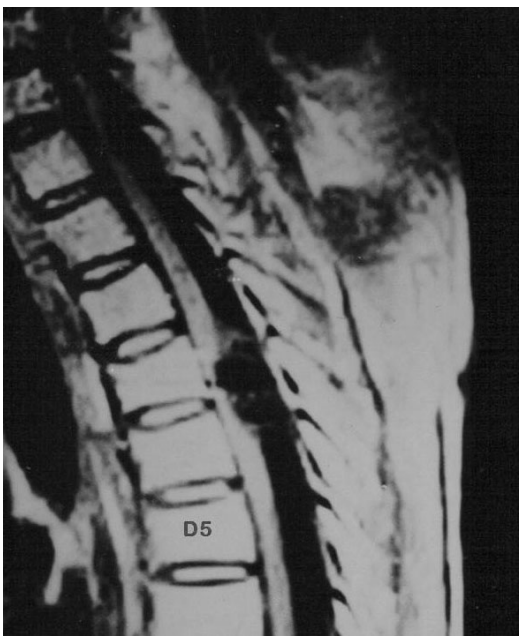

Figure 2. T1-weighted sagittal image showing the cyst compressing the cord at thoracic 3/4 level.

No scolics were identified within the cyst. Solid nodular part lying more rostrally was found densely adherent to the cord tissue. Electrophysiological monitoring was done throughout the procedure and used as a guide to decide the extent of excision. A fully integrated electrophysiological monitor (EPM064™, E-Trolz's® Inc., North Andover, MA, USA) was used for the purpose. An electrical stimulus in the form of square waves was used at a pulse of 200 cycles per sec with an intensity of 10-15 mAmp. The stimuli were delivered at a rate of 4.7 per sec via 3 channels. One set of electrode was placed over the scalp along with two sets of electrodes on the spinal cord and one on each side of operating area. The posterior tibial nerve was stimulated and the responses were recorded at scalp. An initial somatosensory evoked potential (SSEP) of 30-40 msec was recorded from the scalp electrodes, to be used as reference value. Any changes in the SSEP during surgery were carefully monitored. An attenuation in SSEP up to 40-50% of baseline was considered acceptable. The attenuation was usually encountered during cord manipulation, excessive retraction and excision or extensive dissection of cyst wall off the spinal cord. Any of these procedures were abandoned immediately as soon as a 50% or more fall in SSEP was noted. The surgery was withheld until the SSEP returned back to baseline, which usually took 10-15 min. Most of the lesion was removed with the help of Cavitron® ultrasonic surgical aspirator (CUSA® Excel™, Integra Radionics, Burlington, MA, USA) but the part of solid portion, and very small part of cyst wall adherent to the cord tissue left behind.

Postoperative course

Histological evaluation showed translucent cyst wall with an eosinophilic lining, clear fluid and chronic inflammatory cells, consistent with cysticercosis (Fig. 4). Albendazole 300 mg three times daily was continued for a period of four weeks. The steroid cover was given in the form of prednisolone 40 mg per day for first week, tapered over the next week. Postoperatively, the power improved in the lower limbs, but pain persisted and was still present at a one year follow-up. The pain was controlled with the help of routine pain killers like ibuprofen and paracetamol.

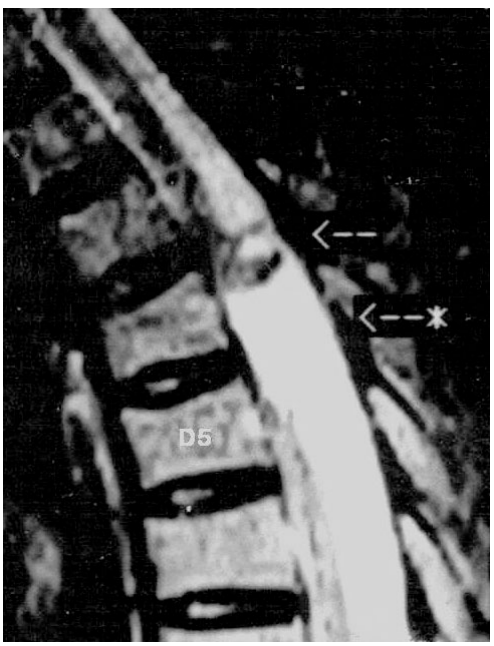

Figure 3. T2-weighted sagittal sequence. The lesion is present posterior to cord pushing it anteriorly, enhancing on contrast administration. The oedema can be seen surrounding the lesion.

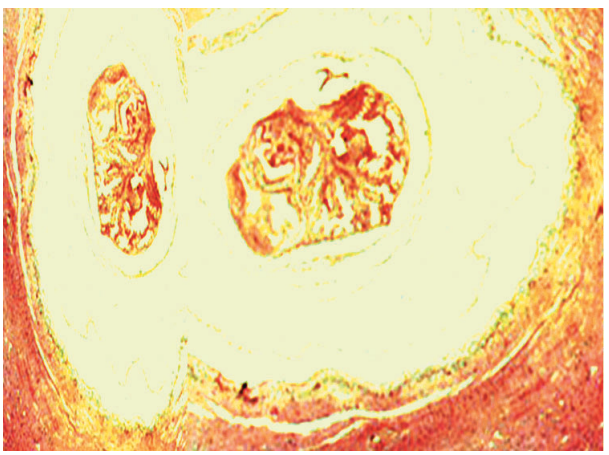

Figure 4. Photomicrograph showing two dead translucent cysts with eosinophilic lining surrounded by clear fluid and chronic inflammatory cells viz. neutrophils and eosinophils. The meningeal thickening can be seen clearly, secondary to inflammatory response (hematoxylin and eosin bleached stain, x600)

DISCUSSION

Human NCC was first described by Paranoli Rumi in 1550[1,2] whereas the first reference of intraspinal cysticercosis is attributed to Rockitansky in 1856[5]. Contrary to the cranial variety, incidence of spinal NCC remains very low with less than 200 cases being reported[1]. Rositti et al.[6] found an incidence of 1.4% in 205 NCC patients while Cenelas et al.[7] reported the incidence of 2.7% among 296 patients of NCC. Despite of some variations, an incidence of 1.5-3.0% is often agreed by most authors[1,3,7].

The basis of rarity for spinal cysticercosis as compared to cranial NCC remains a matter of debate. Approximately two thirds cases of spinal NCC occur in presence of coexisting cranial NCC with primary isolated spinal involvement constituting a minority[1]. Quieroz et al.[8] proposed that the CSF reflux at craniovertebral junction can prevent the spinal dissemination by propelling the floating cysts back to intracranial space. Another hypothesis given for Cysticercus larvae descending into intradural extramedullary space is retrograde flow of these larvae through valveless epidural venous plexus, which may conduct blood in any direction under the influence of intra-abdominal and intrathoracic pressure variations[9]. This explains the occurrence of spinal NCC even without the involvement of brain, as in our case. Whereas on the one hand the larval migration is prevented by CSF reflux, on the other hand this portal remains the most important mode of entry for Cysticercus larvae to the spinal territory[7,8]. Consequently the most common location of NCC in spine is subarachnoid space accounting for the 80% all cases. Intramedullary lesions, which are thought to be secondary to haematogenous spread[8,10], constitute the remaining 20% of cases. The extradural occurrence of NCC in spine is exceedingly rare[8].

The sign and symptoms produced by spinal NCC largely remain a function of size and location of the lesion as well as the products released by the cyst degeneration. The important underlying pathophysiological mechanisms are mass effect, inflammatory reaction causing arachnoiditis and meningitis or obstruction of subarachnoid pathways[1]. Myelopathy caused by the cord compression commonly leads to progressive weakness[7,11,12]. Extramedullary NCC of lumbar region tends to give rise relatively slow and insidious onset of symptoms whereas an intramedullary lesion in cervical canal produces fast and early deterioration[1,12]. The inflammatory reaction evoked by cyst degeneration when a parasite dies may induce severe symptoms[1]. Live cysts cause less inflammation and therefore are easy to excise surgically[13-15].

MRI is the diagnostic modality of choice for evaluating the spinal NCC. Ratnalkar et al.[16] described the MRI findings according to the different stages of disease. The initial stage or vescicular stage, where the parasite is live, is characterised by cystic lesions isointense to CSF appearing hypointense in T1 while hyperintense in T2 weighted images, without any surrounding oedema. In second stage (colloidal vesicular stage), an immune response is generated due to dying parasite resulting in peri-lesional oedema. Due to breach in blood-CNS-barrier the cyst appears as ring enhancing lesion in contrast enhanced CT scan. The previously hypointense cyst in T1 weighted images, now appears mild hyperintense in T1 while hyperintense in T2. The perilesional oedema begins to appear now and is seen as hypointense in T1 while hyperintense in T2 weighted sequences. During the third stage, known as granulonodular stage, the capsule thickens and calcification begins. Finally, in the fourth stage, when larva is dead (calcified nodular stage) the densely calcified scolics and cysts are difficult to visualise in MRI. The changes can be seen on CT scan more readily as areas of calcification. In the present case, the lesions were present at the level of thoracic 3 to 4 level. Mild enhancement was seen in T2 weighted MRI sequence after gadolinium injection, presumably secondary to dead cysts. In spite of the classical description of different stages of NCC, it is important to remember that all the stages of Cysticercus larva can be present simultaneously.

Rosas et al.[17] demonstrated that ELISA of CSF is helpful in confirming the diagnosis of NCC carrying a high sensitivity of 87% with a specificity of 97% as opposed to sensitivity of 50% and specificity of 70% for

serum serological studies. However, excision and histopathological examination remains the only definitive method of confirming the diagnosis[18,19]. The typical histopathological findings of NCC, as seen in our case, are presence of dead or active translucent cysts with eosinophilic lining. The cysts are usually surrounded by clear fluid and chronic inflammatory cells including neutrophils, eosinophils and giant cells. Calcified cysts can be seen in late and inactive stages. The meningeal thickening and signs of arachnoiditis are also common, as seen in the case presented here.

Other common conditions such as simple or complex arachnoid cyst, hydatid cysts, tuberculosis, sarcoidosis or subarachnoid metastatic neoplasm should be considered in the differential diagnosis[4].

The parenchymal variety is considered to be most responsive to pharmacological therapy[20]. The classical anticysticercal drugs, albendazole and praziquental can be tried as a first line of treatment[19,21,22]. Steroid cover and strict neurological monitoring is necessary during the medical management to avoid the acute neurological deterioration from the inflammatory response as the parasites die[21]. Due to natural confines of the spinal canal, a low threshold should be kept in performing a surgical decompression if a clinical deterioration is observed during the pharmacotherapy[1]. Due to higher CSF penetration[23] and increased serum levels by concomitant administration of steroids[19], albendazole is claimed to be superior to praziquental. Mohanty et al.[24,25] believe that the spinal NCC represents focal manifestation of a systemic disease and recommend medical therapy in all patients with spinal NCC.

Surgery has got a definitive place in the management of spinal NCC mainly due to two reasons: first; the efficacy of medical treatment in subarachnoidal, cisternal and extradural NCC remains unclear[14,15], secondly; the acute exacerbations of neurological deficits in the course of disease need prompt surgical attention[1]. The indications for a surgical intervention are presence of severe and progressive symptoms and failure of medical management or acute neurological deterioration during pharmacotherapy[1]. The excision of the extramedullary lesion is often difficult due to preformed dense adhesions from previous arachnoiditis[1,12,26]. The aim should be maximum possible excision without compromising the neurological status. Operating microscope and CUSA can be useful adjunct to the surgery to aid the dissection and excision. We recommend continuous intraoperative physiological monitoring which should be used as a guide to decide the extent of excision. Due to severe inflammatory process intraoperative ultrasonography may help in localization of the lesion[12,26] and it may be safer to leave behind a small part of capsule densely adherent to the cord tissue. A number of measures have been suggested which may assist in cyst extirpation including sharp dissection, gentle irrigation and Valsalva manoeuvres[1,12,26]. Duroplasty may be required to re-establish the CSF flow in cases where arachnoiditis induced CSF blockade is suspected[1]. Cyst migration is a well documented phenomenon[27] and if the interval between the surgery and imaging is long, surgeon should consider repeating the scan in order to make sure that the targeted lesion lies within the planned surgical field.

The final outcome of this potentially curable condition is reported to be unsatisfactory[28]. Patients with chronic arachnoidal scarring or spinal cord inflammation may suffer suboptimal outcomes despite surgical interventions, as in our patient. Other factors responsible for poor outcomes are parenchymal gliosis, pachyleptomeningitis causing cord degeneration, and vascular compromise[8,29].

In conclusion, spinal NCC represents a rare manifestation of a common parasitic infestation of CNS. It should be considered in differential diagnosis of spinal space occupying lesion in endemic areas. Medical management can be tried with a low threshold to operate should any clinical deterioration is observed. The possibility of cyst migration should be kept in mind while planning the surgery. Due to dense arachnoiditis, despite the best surgical and medical measures, the possibility of suboptimal outcome should be discussed with patient.

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