Spinal Cord Compression and Bilateral Sensory Neural Hearing Loss: An Unusual Manifestation of Neurocysticercosis

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Neurocysticercosis is the most common parasitic infestation involving the central nervous system in tropical countries. Common presentations are seizure, meningitis and increased intracranial pressure. The authors report a case of a 52-year-old woman with racemose neurocysticercosis in the subarachnoid space at the cistern of the brain through the lumbar cistern. She presented with progressive paraparesis due to spinal cord compression and finally had progressive bilateral sensori-neural hearing loss. MRI brain and the whole spinal cord revealed numerous rim-enhancing cystic lesions at the basal cistern, prepontine cistern, bilateral cerebellopontine angle, internal acoustic canals, intramedullary lesion at the 5th cervical spinal level, lumbar cistern lesions and secondary syringomyelia at the thoracic spinal cord. The histopathologic examination confirmed cysticercosis. After treatment by albendazole and surgical removal, she still developed recurrent spinal compression at a higher level and obstructive hydrocephalus. Finally, she died from status epilepticus and septic shock.

Keywords : *Neurocysticercosis, Racemose, Spinal cord compression, Sensori-neural hearing loss, Cerebellopontine angle*

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Neurocysticercosis, an infestation caused by the encysted larva of the tapeworm *Taenia Solium*, is now recognized as the most frequent parasitic disease of the central nervous system (CNS) and constitutes a major public health problem for most developing countries[1-6]. In the CNS, five patterns have been described; parenchymatous, subarachnoid, intraventricular, disseminated and spinal form[7]. Brain parenchyma is generally considered as the most common site of involvement, and certain reports claim that the involvement of subarachnoid space is more common [6,8-10]. Seizures are the most common clinical presentation, but other clinical problems occur, depending upon the localization and viability of the parasite[6,11]. There were some reports of atypical presentations of neurocysticercosis such as dorsal midbrain syndrome, sensor-neural hearing loss, cerebral hemorrhage, painful cervical radiculopathy, spinal cord syndrome, dystonia and nominal aphasia masquerading as transient ischemic attact[12] However, no previous literature has reported a patient with neurocysticercosis presenting with spinal cord compression and bilateral sensorineural hearing loss in the same patient.

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Case Report

A 52-year-old woman was refered from Pitsanulok Hospital on February 16th 2002. One year before admission, she presented with numbness of both feet which slowly progressive to paraparesis within several months. Spinal cord compression was diagnosed, laminectomy and spinal decompression was done by an orthopedist. Operative findings revealed numerous cystic lesions. Histopathological findings were racemose cysticercosis. No medication was

prescribed. Her weakness improved after the operation and rehabilitation. Two months later, she developed recurrent weakness in both legs and numbness up to the nipple level. She also had urinary retention. A second laminectomy was done without improvement. The finding of the second laminectomy was numerous cystic lesions in the subarachnoid space. Approximately three months later, she developed bilateral hearing loss (left side more than right side), diffuse headache, nausea and vomiting. Physical examination revealed cachexia, generalized muscle atrophy especially the intrinsic muscle of the hands and feet. Cranial nerve examination found bilateral sensory-neural hearing loss, that was confirmed by audiogram. Motor power was graded as IV/V at the upper extremities and 0/V at lower extremities. Deep tendon reflexes were grade 4+ in the upper extremities and grade 0 in the lower extremities. Plantar reflex was no response bilaterally. There was sensory loss from the toes to the fourth thoracic level

Investigations: CBC showed; hematocrit 37 %, WBC 6,800 /mm^3 (neutrophile 68%, lymphcyte 21%, monocyte 9%, eosinophile 2%) and platelet 183,000/ mm^3. MRI brain and whole spine with gadolinium enhancement were performed. There were numerous rim-enhancing cystic lesions in the subarachnoid spaces either at the prepontine cistern, basal cistern, cerebellopontine (C-P) angle (Fig. 1,2.) and along the cervical spinal canal through the lumbar cistern, that caused secondary syringomyelia (Fig. 3-5). The C-P angle lesions demonstrated involvement of the internal acoustic canal (left side more than right side) which could explain bilateral sensory-neural hearing loss (Fig. 1,2). Gross and histologic findings from a previous hospital were multiple cystic lesions with some scolex that confirmed cysticercosis.

She received albendazole 400 mg/day (albendazole 100 mg/tab, 2 tablets after meals twice a day) for 7 days and dexamethasone 4 mg intravenous every 6 hours for a long course of albendazole treatment and after treatment for 4 days to decrease debris tissue reaction. After that she did not respond to any medical therapy and then she was operated by neurosurgery (at Siriraj Hospital). The indications of surgery were intractable to medical treatment and progressive headache and hearing loss. There were four major operations, the first procedure was a suboccipital craniotomy and removed racemose cysticercosis. Second, third and fourth procedures were laminectomies in order to the cervical spine, thoracic spine and lumbosacral spine regions. 0ne month later, her headache and weakness had improved. Finally, she had recurrent cysticercosis at the fourth to seventh cervical-spine level and C-P angle that caused obstructive hydrocephalus. She died due to refractory status epilepticus, hospital acquired pneumonia and septic shock.

Discussion and Literature Reviews

Neurocysticercosis is one of the most common parasite infestations of the CNS in the world[1,2,11]

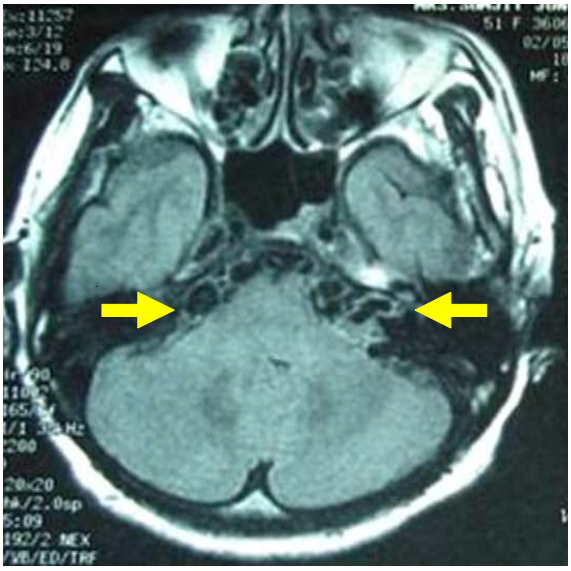

Fig. 1 Axial MRI brain T 1 weighted : multiple cystic lesions at prepontine cistern and bilateral cerebellopontine angle (arrows)

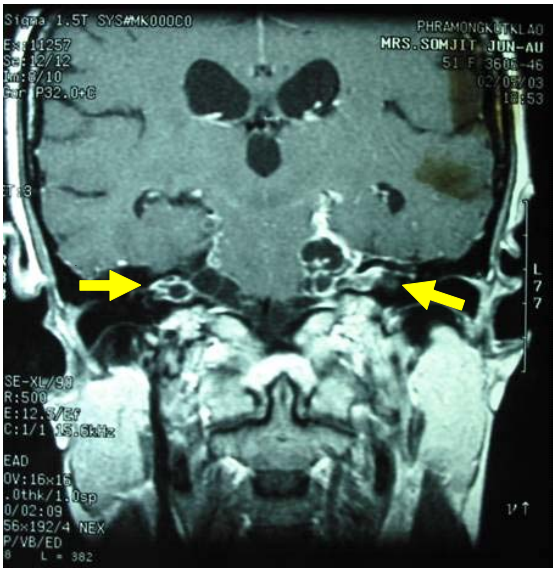

Fig. 2 Coronal MRI brain T 1 weighted with gadolinium: multiple rim-enhancing cystic lesions at subarachnoid space (arrows)

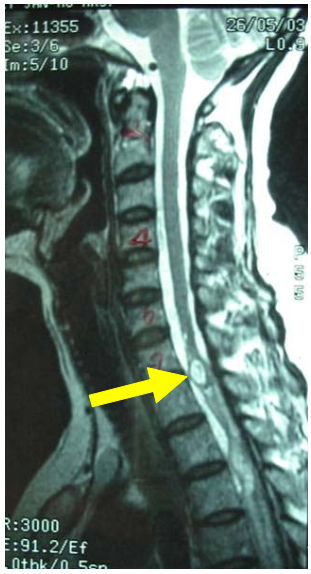

Fig. 3 Saggital MRI C-spine T 2 weighted: an intramedullary cystic lesion at spinal cord level C6 (arrow)

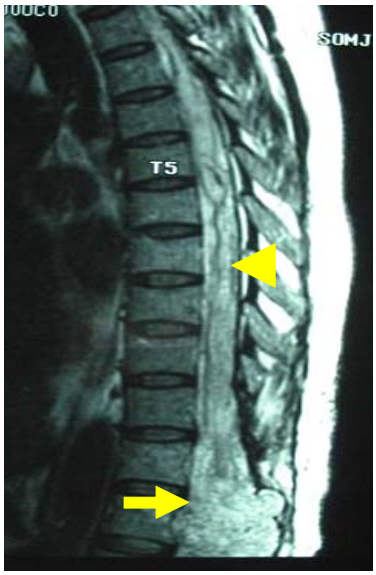

Fig. 4 Saggital MRI T-spine T 2 weighted: numerous of cystic lesions at subarachnoid space (arrow) with secondary syringomyelia (arrow head)

Pathologic classification of neurocysticercosis[11] is arachnoiditis (48.2 %), hydrocephalus secondary to meningeal inflammation (25%), parenchymal cyst (13.2%), brain infarction due to vasculitis (2.3%), intraventricular cyst (0.7%), spinal cyst (0.7%), parenchymal calcification (57%) and hydrocephalus secondary to meningeal fibrosis (3.8%).

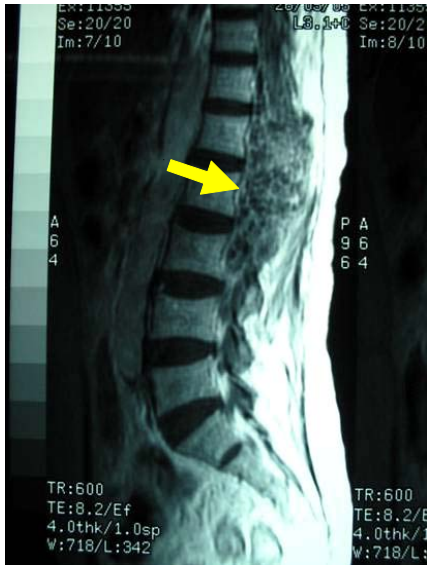

Fig. 5 Saggital MRI LS-spineT 1 weighted with Gadolinium: numerous of cystic lesions in lumbar cistern with compressed multiple nerve root (arrow)

This case presented with spinal cord compression and bilateral sensori-neural hearing loss. In a recent study, some patients were reported to have a spinal form of cysticercosis alone, which must be differentiated from an intramedullary cord tumor[13,14]. A clinical presentation of neurocysticercosis involving the spinal cord is shown in Table 1[13-24]. Most patients had an intramedullary cord lesion, whereas one case was an extradural lesion and another case was an intradural-extramedullary cord lesion that was similar to the patients. Some patients were reported with cranial neuropathy due to extra-axial compression from racemose neurocysticercosis at C-P angle alone[25,26]. Only three case reports in the literature demonstrated the racemose form of neurocysticercosis at the C-P angle, and one case presented with trigeminal neuralgia[27,28]. Other differential diagnoses of C-P angle cyst are epidermoid cyst, cystic schwanoma, cystic meningioma, lymphoma, sarcoidosis, arachnoid cyst, vascular anomaly, epidural cyst, dermoid cyst and lipoma[14,26,29]. Despite many reports of patients with spinal cysticercosis[13,14,16-17] and C-P angle racemose cysticercosis[25,26,30,31] there has been no report of these two conditions in the same patient. Presentations of spinal compression with bilateral sensori-neural hearing loss can be found in neuro-fibromatosis type 2 (bilateral acoustic neuroma and spinal cord tumor)[32]. After literature review, the authors could find no previous report, so to our knowledge this is the first case of these presentations.

Table 1. Clinical characteristics of neurocysticercosis case reports presented with spinal cord involvement or CP angle lesions

No. of case	Age (yr)	Sex	Symptom	Involve	Location of lesion	Treatment	Outcome	Ref
1	NA	NA	NA	Spinal cord	Intramedullary	NA	NA	12
1	36	male	NA	Spinal cord	Intramedullary at T- spine	Surgery, Medication	Improve	15
1	20	male	NA	Spinal cord	Intramedullary at T-spine	Surgery, Medication	Improve	
6	NA	NA	Paraplegia quadriplegia	Spinal cord	Intramedullary	Surgery & medication	NA	16
1	5	female	Back pain	Spinal cord	Intramedullary at T-spine	Surgery	NA	17
1	28	male	Back pain	Spinal cord	Intramedullary	Surgery	Recovery	18
1	55	male	Spinal cord syndrome	Spinal cord	Intramedullary	Surgery	Not improve	
1	50	male	Spinal cord syndrome	Spinal cord	Intramedullary	Surgery	Improve	
1	NA	NA	NA	Spinal cord	Intradural extramedullary Cervical subarachnoid space Basal cistern Cisterna magna	NA	NA	19
1	NA	male	Quadriparesis	Spinal cord	Extradural lesion at C4,C5 pedicle & lamina	Surgery	Improve	21
2	NA	NA	NA	Spinal cord	Intramedullary	NA	NA	22
8	NA	NA	NA	Spinal cord	Intramedullary T-spine(6 cases) C-spine(2 cases)	Surgery	Improve 7 cases	23
1	NA	NA	Paraplegia	Spinal cord	Intramedullary C-spine	Only medication (albendazole & dexamethazole)	recovery	24
1	NA	NA	NA	CP angle	Left	NA	NA	26
1	NA	NA	NA	CP angle		Albendazole	Improve	27
1	52	female	Contralateral trigeminal neuralgia	CP angle	Left	Surgery remove via left suboccipital craniotomy	Improve	28

Note: NA = not available, CP angle = cerebellopontine angle, Ref. = reference, No. = number, T = thoracic, C = cervical

Medical treatment of racemose cysticercosis has remained controversial because of no true randomized controlled trial. However, many studies showed some benefit of antiparasitic drugs both praziquantel and albendazole[6,7,11,13,33]. But surgical intervention either a shunting procedure or surgical removal and decompression gave lots of benefit in this form[11,13]. The presented case was treated with both medical and surgical methods. Finally, her weakness of both upper extremities and headache improved, but recurrent cysticercosis at the C-P angle and C-spine occurred and caused complications and death.

Conclusion

Neurocysticercosis is the most common parasitic involvement of the CNS especially in tropical countries. The authors reported a rare case of bilateral sensori-neural hearing loss due to C-P angle lesions, spinal cord compression with root involvement and secondary syringomyelia. The diagnosis was confirmed by histology. The presented case is the first case report of these presentations. After treatment by albendazole and surgical cysticercal removal, she still had recurrent spinal cysticercosis and died because of complications.

Reference

1. Nieto D. Historical noted on cysticercosis. In Flisses A, Williams K, Laclette JP, Larralde C, Ridaura C, Betran F, eds: Cysticercosis: present state of knowledge and perspective. New York: Academic Press, 1984: 1-7.
2. Delbrutto OH, Sotelo J. Neurocysticercosis: an update. Rev Infect Dis 1988; 10: 1075-87.

3. Richards F, Schantz P. Laboratory diagnosis of cysticercosis. Clin Lab MED 1991; 11: 1011-28.
4. Bia FJ, Barry M. Parasite infection of the central nervous system. Neural Clin 1986; 4: 171-206.
5. Shandera WX, White AC Jr, Chen JC, Diaz P, Armstrong R. Neurocysticercosis in Houston, Texas: a report of 112 cases. Medicine (Baltimore) 1994; 73: 37-52.
6. Garcia HH, Gonzalez AE, Gilman RH. Diagnosis, treatment and control of Taenia solium cysticercosis. Current opinion in infectious disease 2003; 16: 411-9.
7. Proano JV, Madrazo I, Avelon F, Lopez-Felix B, Diaz G, Grijalva I.Medical treatment for neurocysticercosis characterized by giant subarachnoid cysts. N Engl J Med 2001; 345: 879-85.
8. Del Brutto OH, Rajshekhas V, White Jr, et al. Proposed diagnostic criteria for neurocysticercosis. Neurology 2001; 57: 177-83.
9. McGreevy PB,Nelson GS. Larval cestode infections. In: Strickland GT, ed. Hunter's tropical medicine.7th ed. Philadelphia: WB Saunders, 1991: 843-59.
10. Earnest MP,Reller LB,Filley CM, Grek AJ. Neurocysticercosis in the United states; 35 cases and a review. Rev Infect Disease 1987; 9: 961-79.
11. Barry M,KaldjianLC. Neurocysticercosis. Seminars in Neurology 1993; 13(2): 131-45.
12. Sawhney IM, Singh G, Lekhra OP, Mathuriya SN, Parihar PS, Prabhakar S. Uncommon presentations of neurocysticercosis. J Neurol Sci 1998; 154(1): 94-100.
13. White Jr AC, Riley T. Management of neurocysticercosis. CNS drugs 2003; 17(8): 577-91.
14. Tsementzis SA. Intracranial tumor and spinal disorder. Differential diagnosis in neurology and neurosurgery. Thieme Stuttgart. New York, 2000: 111-293.
15. Muzumdar D, Nadkarni T, Desai K, Dindorkar K, Goel A. Thoracic intramedullary cysticercosis-two case reports. Neurol Med Chir 2002; 42(12): 575-9.
16. Parmar H, Shah J, Patwardhan V, et al. MR imaging in intramedullary cysticercosis. Neuroradiology 2001; 43(11): 968.
17. Homans J, Khoo L, Chen T, Commins DL, Ahmed J, Kovacs A. Spinal intramedullary cysticercosis in a five-year-old child: case report and review of the literature. Pediatr Infect Dis J 2001; 20(9): 904-8.
18. Mathuriya SN, Khosla VK, Vasishta RK, Tewari MK, Pathak AA, Prabhakar SS. Intramedullary cysticercosis: MRI diagnosis. Neurol India 2001; 49(1): 71-4.
19. Ciftci E, Diaz-Marchan PJ,Hayman LA. Intradural-extramedullary spinal cysticercosis: MR imaging findings. Comput Med Imaging Graph 1999; 23(3): 161-4.
20. Amaral L, Maschietto M, Maschietto R, et al. Unusually manifestations of neurocysticercosis in MR imaging. Arq Neuropsiquiatr 2003; 61: 533-41.
21. Mohanty A, Das S, Kolluri VR, Das BS. Spinal extradural cysticercosis: a case report. Spinal Cord 1998; 36(4): 285-7.
22. Garg RK, Nag D. Intramedullary spinal cysticercosis: response to albendazole: case reports and review of literature. Spinal cord 1998; 36(1): 67-70.
23. Mohanty A, Venkatrama SK, Das S, Das BS, Rao BR, Vasudev MK. Spinal intramedullary cysticercosis. Neurosurgery 1997; 40(1): 82-7.
24. Corral I, Quereda C, Moreno A, et al. Intramedullary cysticercosis cured with drug treatment. A case report. Spine 1996; 21(19): 2284-7.
25. Aquiar PH, Miura FK, Napoli PR, et al. Unusual cause for bilateral trigeminal neuralgia: Unilateral racemose cysticercosis of cerebellopontine angle. Case report. Arq neuropsi quitar 2000; 58(4): 1138-1141.
26. Singh S, Gibikote SV, Bannur U, Chacko G, Korah IP, Rajshekhar V. Cysticercosis of the cerebellopontine angle cistern mimicking epidermoid inclusion cyst. Acta Neurol Scand 1999; 99(4): 260-3.
27. Del Brutto OH. Albendazole therapy for subarachnoid cysticerci: clinical and neuroimaging analysis of 17 patients. J Neurol Neurosurg Psychiatry 1997; 62(6): 659-61.
28. Revuelta R, Juambelz P, Balderrama J, Teixeira F. Contralateral trigeminal neuralgia: a new clinical manifestation of neurocysticercosis: case report. Neurosurgery 1995; 37(1): 138-9.
29. Flaherty AW. Neurocutaneous syndrome. In: the Massachusetts general hospital handbook of neurology. Lippincortt Williams & Wilkins, a wolters Kluwer company 2000: 116.
30. Del Brutto OH. Cysticercosis of the cerebellopontine angle: treatment with single-dose praziquantel. Rev Neurol 2000; 31(9): 835-7.
31. Celis MA, Mourier KL, Polivka M, et al. Cisternal cisticercosis of the cerebellopontine angle. A surgically-treated case and review of the literature. Neurochirurgie 1992; 38(2): 108-12.
32. Miller VS, Steve Roach E. Neurocutaneous syndrome. In Bradley WG, ed. Neurology in clinical practice. 3th ed. Butterworth-Heinemann, 2000: 1665-1700.
33. Garg RK.Medical management of neurocysticercosis. Neural India 2001; 49: 329-37.

ภาวะไขสันหลังถูกกดทับร่วมกับภาวะการสูญเสียการได้ยินซึ่งเกิดจากประสาทหูทั้งสองข้าง: อาการที่พบไม่บ่อยในโรคพยาธิตืดหมู

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โรคถุงน้ำจากพยาธิตืดหมูเป็นโรคที่เกิดจากพยาธิตืดหมูที่มีปัญหาต่อระบบประสาทส่วนกลางที่พบบ่อยที่สุดโดยเฉพาะในประเทศเขตร้อน อาการของโรคนี้ที่พบได้บ่อยคือ ชัก อาการเนื่องจากเยื่อหุ้มสมองอักเสบและการเพิ่มความดันในกะโหลกศีรษะ คณะผู้ประพันธ์เสนอรายงานผู้ป่วยหนึ่งราย เพศหญิง อายุ 52 ปี ป่วยเป็นโรคถุงน้ำจากพยาธิตืดหมู ที่มีรอยโรคถุงน้ำคล้ายพวงองุ่น ที่ช่องน้ำไขสันหลังทั้งรอบฐานสมองจนถึงบริเวณต่ำกว่าไขสันหลัง ผู้ป่วยพบแพทย์ด้วยอาการขาสองข้างอ่อนแรงมากขึ้นเรื่อยๆเนื่องจากการกดทับที่บริเวณไขสันหลัง ร่วมกับการสูญเสียการได้ยินซึ่งเกิดจากประสาทหูทั้งสองข้าง เอ็กซเรย์แม่เหล็กไฟฟ้าของสมอง และไขสันหลังพบรอยโรคลักษณะเป็นถุงน้ำจำนวนมากที่บริเวณช่องรอบฐานสมอง รอบก้านสมอง รอยต่อของก้านสมองกับสมองน้อย รูด้านในของเส้นประสาทหู ถุงน้ำภายในไขสันหลังระดับกระดูกคอชั้นที่ 5 และยังเกิดการขยายตัวของหลอดน้ำกลางไขสันหลัง จากการตรวจทางพยาธิวิทยาเข้าได้กับโรคถุงน้ำจากพยาธิตืดหมู แม้ได้รักษาโดยการใช้ยาฆ่าพยาธิร่วมกับการผ่าตัดยังคงเกิดภาวะกดไขสันหลังซ้ำอีกที่ระดับสูงขึ้นร่วมกับการอุดตันภายในโพรงสมอง ผู้ป่วยเสียชีวิตในที่สุดจากภาวะแทรกซ้อนคือภาวะชักต่อเนื่องและภาวะความดันต่ำเนื่องจากการติดเชื้อในกระแสเลือด