Case Report

Cervical Intramedullary Neurenteric Cyst in an Elderly Patient

Ho Jae Kim, M.D., Kyung Soo Min, M.D.,
Young Gyu Kim, M.D., Dong Ho Kim, M.D.

Department of Neurosurgery, College of Medicine, Chungbuk National University, Cheongju, Korea

Intraspinal neurenteric cysts are rare congenital lesions that may occur either alone or in the context of a complex malformative disorder including vertebral, visceral and cutaneous abnormalities. Most of these lesions have an intradural extramedullary location. Rarely, intramedullary neurenteric cysts not associated with other malformations have been reported. A 72-year-old woman presented with a 5-year-history of progressive paraparesis(I/II), urinary retention. A magnetic resonance(MR) image revealed a C6-C7 intramedullary cystic lesion which was located ventrally to the spinal cord and no other spinal abnormalities. The cystic lesion were subtotally removed through posterior approach. On histopathological examination, fibrous connective tissue surrounded by cuboidal cells which contained mucin vacuoles were consistent with neurenteric cyst. Postoperative MR image showed the decompression of the spinal cord. Postoperatively, neurological deficits were improved. We report a case of neurenteric cyst occured in highly old age and unusual location.

KEY WORDS : Neurenteric cyst · Intramedullary.

Introduction

Intraspinal neurenteric cyst appear to be a very rare congenital malformation, which was originated in adhesion and communication between neuroectoderm and endoderm at the 3rd weeks of gestational period. Neurenteric cysts can be an isolated lesion or complex malformations associated with abdominal, thoracic, vertebral and cutaneous lesions in about 50% of cases[4]. Intradural extramedullary locaion in cervicothoracic junction is general, and intramedullary neurenteric cysts have been reported in fewer than 5%[1,5,9,17,18]. Most of neurenteric cysts are observed in childhood and young adulthood. After subtotally resected, the surgical prognosis appear usually favorable[1,8,9]. The patient presented at an oldest age in pertinent literatures, the lesion was located at an unusual site along spinal axis.

Case Report

Presentation

A 72-year-old woman presented with a 5 year history of low back pain, progressive numbness. One month before admission she had ascending paraparesis, urinary retention. She was unable to walk at 1 week before admission. Her medical history was otherwise normal.

• Received : April 29, 2004 • Accepted : May 28, 2004
• Address for reprints : Young Gyu Kim, M.D., Department of Neurosurgery, College of Medicine, Chungbuk National University, 62 Gaesin-dong, Cheong-ju 361-711, Korea
Tel : 043) 269-6087, Fax : 043) 273-1614
E-mail : ygk@chungbuk.ac.kr

Examination

On admission, vital sign was normal and there were no cutaneous stigmata on her back. On neurological examin-ation, motor strength was grade 4/5 throughout the both upper extremity, 1/5 throughout the right lower extremity and 2/5 throughout the left lower extremity. Superficial and deep sensation were decreased below the C7 dermatome. Deep tendon reflexes of the lower extremities were increased slightly. Babinsky's sign were strong positive bilaterally. An X-ray study of the spine did not reveal abnormal finding of bony structure. A magnetic resonance(MR) image revealed a C6-7 intramedullary lobulating cystic lesion slight hyperintense to cerebrospinal fluid, hypointense to spinal cord on spin echo T1-weighted sequences and isointense to cerebrospinal fluid on T2-weighted sequences(Fig. 1). The spinal cord was displaced dorsolaterally and was flattened by the ventrally located cystic mass. The lesion was not enhanced on postcontrast T1-weighted sequences. The vertebra and soft tissue were normal. Laboratory findings of blood and urine were within normal limit.

Operation

Cervical 6-7 level laminectomy was performed. The paras-

Table 1. Reported cases of intramedullary neurenteric cysts in literatures*

Author, Year	Age(yrs)/ Sex	Other abnormality	Symptom	Location	Cyst contents	Outcome
Knight et al,1954	1/NR	Spina bifida	Anxiety, rigid neck	C7–T2	Milky fluid	Improved
Lanmaid et al,1963	11/M	Scoliosis	Sensory disturbance	NA	NR	Improved
	5/M	Diastematomyelia	NA	NA	NR	Improved
Rewcastle & Fancoeur,1964	34/F	Normal	Paraparesis	T7–L2	NR	Worse paresis
Silvernail & Brown,1972	12/M	Multiple hemivertebra	Tetraparesis	T2	Blackish fluid	Improved
Kwok & Jeffreys,1982	54/M	Hemi, Fused vertebra	NA	T2–5	NR	Improved
Fortuna et al, 1983	47/F	Normal	Lt.LEparesis	T12–L1	NR	Improved
Takemi et al, 1984	4/F	NR	Nape pain Paraparesis	C3–4	NR	Improved
Mizuno et al, 1988	6/M	Normal	UE paresis	C2–5	Clear jelly	Improved
	14/M	Normal	Neck pain	C2–5	CSF–like	Improved
Rao et al,1996	27/F	Normal	Neck pain	C3–T1	NR	Improved
Rivierez et al, 1997	46/M	Normal	LBP LE paresis	L1–2	Clear colorless	Worse paresis
Lippman et al, 2000	68/F	Normal	LBP Paraparesis	T10–11	Milky fluid	Worse paresis
Shinghal et al, 2001	67/M	Normal	Paraparesis UR	T7	Milky fluid	Improved
Rhee et al, 2001	16/M	Normal	Axillary pain,LEparesis	T3–6	Milky mucinous	Improved Recur
	17/M	Normal	UE tingling sense	T3–6	Milky	Improved
Agrawal et al, 2002	3mon/M	Normal	Paraplegia	C3	NA	NA
Kumar et al, 2002	3cases below18yrs	NA	NA	NA	NA	NA
Paolini et al, 2003	28/F	Sacral schisis	LEnumbness	T8–9	Milky	Improved
Present case, 2004	72/F	Normal	Paraparesis UR	C6–7	Clear jelly–like	Improved

* : Abbreviations : M=male; F=female; LBP=low back pain; LE=lower extremity; mon=month; Lt=left; NR=not reported; NA=not available; UE=upper extremity; UR=urinary retention; yrs=years

pinal soft tissue, spinous processes, laminae, dura were all normal. On opening the dura, partially transparent pia matter and thinned cord in the dorsal midline were seen. Standard posterior median myelotomy exposed a thin walled translucent membrane.

Incision of membrane released clear jellylike contents. After evacuating of cystic contents, the anterior pia and dura through transparent ventral wall of cyst was seen and membrane of cyst was partially excised(Fig. 2).

Histopathological examination

Light microscopy revealed that the folded cyst wall was lined by simple, partially pseudostratified cuboidal and columnar epithelial cells. These cells abutted on loose fibrous tissue and contained mucin vacuoles which showed a strong positive reaction for mucic-armine and periodic acid-Schiff stains(Fig. 3).

Postoperative course

The postoperative MR image revealed that cystic lesion was

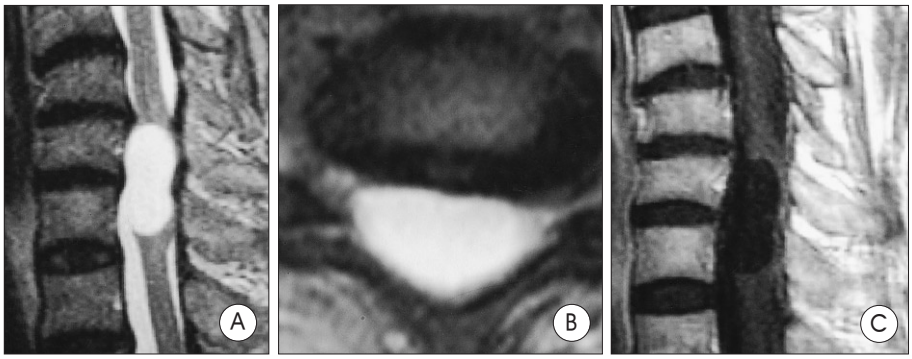

Fig. 1. Preoperative spin echo sagittal(A), axial(B) T2 weighted magnetic resonance(MR) image, and postcontrast sagittal(C) T1 weighted magnetic resonace image showing C6−C7 intramedullary cystic lesion which locates ventrally to the spinal cord and compresses the spinal cord. There is no enhancing part of cystic lesion. Bony, soft tissue abnormalities are not seen.

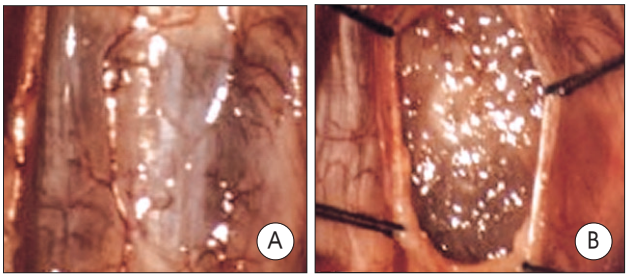

Fig. 2. Intraoperative photograph. Buldged dura is opened then bluish cyst wall appears under transparent pia matter(A). After posterior midline myelotomy, jelly−like contents are seen(B).

absent and dorsally located cord was expanded(Fig. 4). Postoperatively, the neurological deficits showed steady improvement. One month after the operation, the patient was again able to walk unaided.

Discussion

Intraspinal neurenteric cysts are congenital lesion derived from endodermal remnants during period of notochordal formation at the 3rd week of fetal development[2,4,15]. Bentley and Smith[4] explained embryological aspects as split notochord theory. Rhaney and Barclay[15] reviewed three cases of enterogenous cysts and introduced causal explanation as intercalation of endoderm between the notochord and the neuroectoderm. In about 50% of cases, neurenteric cysts may be combined with vertebral abnor-malities such as cleft vertebra, hemivertebra, spina bif-ida, absent or fused vertebra, or diastemat-omyelia, Klippel-Feil anomaly and with anomalies which may be fatal[15,19]. Their occurences have been reported by other authors but are rare malformations comprising 0.7 to 1.3% of all spinal cord tumors and 16% of cysts[9,13]. These cysts occur in men 1.5 to 3 times as often as in women and usually present in the patients second and third decades of life[1,12]. Clinical presentation more than 50 years of age are very rare[1,9,18]. Authors was unable to find highly old age patients(the 8th decade of life) like presenting case in pertinent literatures.

At their most severe lesions, they are associated with other major congenital malformations that may be incompatible with life[2,4]. Patients who present in later childhood or young adulthood are usually found to have isolated cysts of spine with or without anterior or posterior dysraphic abnormalities[9,16]. The clinical features are in no way different from those of other space-occupying lesions of the spinal canal, the specific symptom depending soley on the level affected[1]. The clinical courses may progress rapidly or intermittently be progressive[3,17]. In some patients, symptomatic presentations is characterized episodically by exacerbation and remission[3,6,10,17], mimicking an inflammatory disease like multiple sclerosis. These clinical patterns have been explained as periodic rupture of the cyst contents or changes in the rate of production, reabsorption by the cyst walls, and hemorrhagic event within cys-ts[12,17]. An event of trauma may in-itiate or exacerbate symptoms in some patients[6,8,16]. In our prese-nting case, paraparesis have sho-wn patterns that waxed and waned for 5 years. There were neither histories of trauma or hemo-rrhagic findings in operation. Causal mechanisms of

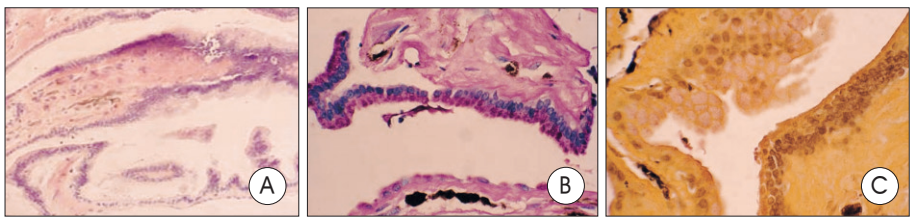

Fig. 3. Photomicrograph of the intramedullary neurenteric cyst of the spinal cord. The cyst is lined by non−ciliated cuboidal and columnar epithelial cells. The cells contain apical mucin vaculoes which show a strong positive reaction for periodic acid−Schiff(PAS), mucicarmine stains. The wall of the cyst is composed by loose fibrous tissue.(A : H&E,×100 ; B : PAS×400 ; C : mucicarmine stain ; original magnification ×400).

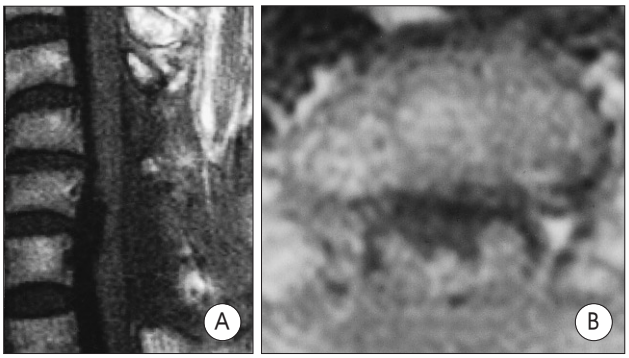

Fig. 4. Postoperative spin echo sagittal(A), and axial(B) T1 weighted magnetic resonance image showing disappeared cystic lesion and dorsolateral expansion of the spinal cord. Adjacent cerebrospinal fluid space widening are only seen.

delayed symptomatic progression in old ages such as our case are vague.

In cases more than 50%, these lesions was located in cervical region[9,20]. Greater than 90% of these cysts have been reported to be located in the intradural extramedullary compartment of the spinal canal[1]. Intramedullary location of these cysts are rare(Table 1)[1,9,14,16,17]. Moreover, intramedullary neurenteric cysts not associated with other congenital abnormalities are very rare[13,16]. In domestic literatures, 1 case(16 years old male) who had intramedullary neurenteric cyst not associated with other congenital abnormalities was reported by Rhee et al.[16].

Myelography with postmyelographic CT, and MRI are available techniques at present for visualizing intraspinal neurenteric cysts and their coexisting abnormalities. The MR pattern is more indicative of the cystic nature of the lesion than was myelography or postmyelographic CT[3,10], increase sensitivity, and provides information as to best surgical approach[10]. MR image in our cases and literatures have shown neurenteric cysts to be isointense to hyperintense relative to cerebrospinal fluid and hyperintense to spinal cord on T2-weighted sequences. On T1-weighted sequences, most are isointense or mildly hyperintense to cerebrospinal fluid and hypointense relative to spinal cord. Intensity pattern of cystic lesion on MR image may vary with the nature(particularly protein contents) of the intra-cavitary fluid[3,7,17]. Cystic morphology of the lesion, the kind of signal in the two sequences, and the absence of mural nodule, lack of contrast enhancement usually differentiate these lesion from the more common spinal cord tumors[14,17].

Many authors have reported favorable results after complete or partial resections of cyst wall via posterior approach, although cysts were located ventrally to the spinal cord[1,6,8,9,12,18]. Extramedullary neurenteric cysts are a clear plane of dissection between lesion and the neural tissue. In such cases, complete resection can be achieved. Because intramedullary lesion such as presenting case may have no clear plane of cleavage between the spinal cord and the cyst wall, complete resection of cyst wall may be complicated. In these cases, although subtotal resection and wide fenestration can retain favorable results[9,14,17,18], a few authors have reported recurrence after simple aspiration or subtotal resection[11,16]. Complete resection is recommended, if possible, but overly aggressive cyst removal that may be risky should be avoided[9]. Authors' case is favorable during one year period after subtotal resection and wide fenestration. Several authors have reported on the anterior approach for cervical neurenteric cysts because it provides good visualization and safe removal although anterior approach is technically difficult and complicated[3,14,19]. In these appro-aches, relative locations between cyst and cord, retraction of cord should be considered. Ultrasound, spinal cord monitoring, endoscope can be helpful intraoperatively in some operative conditions[9,12].

Conclusion

Intramedullary neurenteric cysts not associated with other congenital abnormalities are very rare, moreover in old age. A 72 year old female presented with paraplegia was admitted in our institute. A MR image revealed low cervical intramedullary cystic lesion causing cord compression. After C6-7 laminectomy, subtotal resection of cyst was performed and on histopathologic examination the cyst was consistent with neurenteric cyst. The patient was able to walk unaided. In view of their rarity, peculiarity in terms of presenting age and location, authors report this case with the review of literatures.

• Acknowledgement

This study was financially supported by a 2004 clinical research grants from Chungbuk National University Hospital, Korea.

References

1. Agnoli AL, Laun A, Schonmayr R : Enterogenous intraspinal cysts. **J Neurosurg 61** : 834-840, 1984
2. Anderson FM : Occult spinal dysraphism. **J Pediatr 73** : 163-177, 1968
3. Bae KJ, Kim IM : Anterior surgical approach in recurrent cervical neurenteric cyst. **J Korean Neurosurg Soc 29** : 1258-1261, 2000
4. Bentley JF, Smith JR : Developmental posterior enteric remnants and spinal malformations : The split notochord syndrome. **Achi Dis Child 35** : 76-86, 1960
5. Brooks BS, Duvall ER, Gammal EI, Garcia JH, Gupta KL, Kapila A : Neuroimaging features of neurenteric cysts ; analysis of nine cases and review of the literature. **Am J Neuroradiol 14** : 735-746, 1993

6. D'Almeida AC, Stewart DH : Neurenteric cyst. **Neurosurgery 8** : 596-599, 1981
7. Geremia GK, Russel EJ, Clasen RA : MR imaging characteristics of a neurenteric cyst. **Am J Nueroradiol 9** : 978-980, 1988
8. Jae CB, Kim TS, Park JT, Lim YJ, Leem W, Kim GK : Intradural extramedullary enterogenous cyst in cervical spine. **J Korean Neurosurg Soc 27** : 1741-1745, 1998
9. Lippman CR, Arginteanu M, Purohit D, Naidichi TP, Camins MB : Intramedullary neurenteric cysts of the spine. **J Neurosurg 94** : 305-309, 2001
10. Miyagi K, Mukawa J, Mekaru S, Ishikawa Y, Kinjo T, Nakasone S : Enterogenous cyst in the cervical spinal canal. **J Neurosurg 68** : 292-296, 1988
11. Mizuno J, Fiandaca MS, Nishio S, O'Brien MS : Recurrent intramedullary enterogenous cyst of the cervical spinal cord. **Childs Nerv Syst 4** : 47-49, 1988
12. Osenbach RK, Godersky JC, Traynelis VC, Schelper RD : Intradural extramedullary cyst of the spinal canal : clinical presentation, radiographic diagnosis, and surgical management. **Neurosurgery 30** : 35-42, 1992
13. Palma L, Di Lorezo N : Spinal endodermal cyst without associated vertebral or other congenital abnormalities. Report of four cases and review of the literature. **Acta Neurochir 33** : 283-300, 1976
14. Paolini S, Ciappetta P, Domenicucci M, Guiducci A : Intramedullary neurenteric cyst with a false mural nodule. **Neurosurgery 52** : 243-246, 2003
15. Rhaney K, Barclay GPT : Enterogenous cysts and congenital diverticula of the alimentary canal with abnormalities of the vertebral column and spinal cord. **J Pathol Bact 77** : 457-471, 1959
16. Rhee JJ, Ra YS, Khang SK, Roh SW, Rhim SC : Recurrent intramedullary neurenteric cyst of the spine. **J Korean Neurosurg Soc 30** : 1422-1426, 2001
17. Rivierez M, Buisson G, Kujas M, Ridarch A, Mignon E, Jouannelle A, et al : Intramedullary neurenteric cyst without any associated malformation. **Acta Neurochir 139** : 887-890, 1997
18. Singhal BS, Parekh HN, Ursekar M, Deopujari CE, Manghani DK : Intramedullary neurenteric cyst in midthoracic spine in an adult. **Neurol India 49** : 302-304, 2001
19. Takase T, Ishigawa M, Nishi S, Aoki T, Wada E, Owaki H, et al : A recurrent intradural cervical neurenteric cyst operated on using an anterior approach. **Surg Neurol 59** : 34-39, 2003
20. Wilkins RH, Odom GL : Spinal intradural cyst in Vinken PJ, Bruyn GW(eds); **Handbook of clinical neurology**. Amsterdam, North-Holland, 1976, vol20, pp55-102