www.jkns.or.kr http://dx.doi.org/10.3340/jkns.2012.52.1.67 Print ISSN 2005-3711 On-line ISSN 1598-7876
J Korean Neurosurg Soc 52 : 67-70, 2012

Case Report

Spinal Intramedullary Ependymal Cysts : A Case Report and Review of the Literature

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We report a rare case of a spinal intramedullary ependymal cyst in a 46-year-old female and review the 17 pathologically proven cases in the literature. The patient presented with a two-week history of gradually increasing tingling in her left posterior thigh and calf. A preoperative magnetic resonance image revealed a well-defined intramedullary cystic lesion on the ventral side of the spinal cord at the T11 to T12 levels. The lesion was hyper intense in T2-weighted images and hypointense in T1-weighted. The patient underwent a right-side hemilaminectomy at the T11 to T12 levels and fenestration of the cyst wall. After having the cyst wall partially removed and communication established between the cyst and the subarachnoid space, the patient improved neurologically. A histological study of the surgical specimens revealed that the cyst wall consisted of glial cells lined by a simple cuboidal to columnar epithelium. An immunohistochemical examination of the cells lining the cyst wall was positive for S-100 protein, glial fibrillary acidic protein, epithelial membrane antigen, and cytokeratin. We suggest that the optimal treatment of intramedullary ependymal cysts creates adequate communication between the cyst and the subarachnoid space.

Key Words : Ependymal cyst · Intramedullary · Spinal cord · Immunochemistry.

INTRODUCTION

The different varieties of developmental intradural spinal cord cysts include enterogenous cysts, teratomatous cysts, arachnoid cysts, neurenteric cysts, foregut cysts, bronchogenic cysts, epithelial cysts, colloid cysts, and ependymal cysts[4,5,10,11]. All of these cysts are infrequent, but spinal intramedullary ependymal cysts are particularly rare. Only 17 pathologically proven cases have been reported since the first was published in 1938[1,5,6,10,14,16]. Here, we report an additional case : a 46-year-old female with an intramedullary ependymal cyst in the conus medullaris. Additionally, we discuss the diagnosis, operative management, and pathological features and review the literature.

CASE REPORT

A 46-year-old female presented with a 2-week history of a gradually increasing tingling sensation in her left posterior thigh and calf. A neurological examination showed grade IV/V motor strength for left hip flexion and knee extension. The femoral nerve stretch test was positive for the left leg. A magnetic resonance image (MRI) revealed a well-defined cystic lesion on the ventral side of the spinal cord at the T11 and T12 levels. The lesion was oval in shape and 36×15 mm in size. The spinal cord was compressed and displaced posteriorly and leftward. The lesion was hyperintense in T2-weighted images and hypointense in T1-weighted images (Fig. 1).

The patient underwent a T11-T12 right hemilaminectomy and fenestration of the cyst wall. A midline myelotomy revealed a glistening white cyst without hemorrhage (Fig. 2) on the central part of the conus medullaris. A 23-gauge needle was inserted into the lesion, and cerebral spinal fluid (CSF)-like clear fluid was aspirated, resulting in the immediate collapse of the cyst. The cyst wall and compressed cord tissue was removed to achieve adequate communication between the cyst and the subarachnoid space. On the day after the surgery, the patient's sensory symptoms disappeared, and her motor weakness recovered to normal muscle strength. On the 2nd postoperative day,

• Received : April 9, 2012 • Revised : May 23, 2012 • Accepted : June 18, 2012
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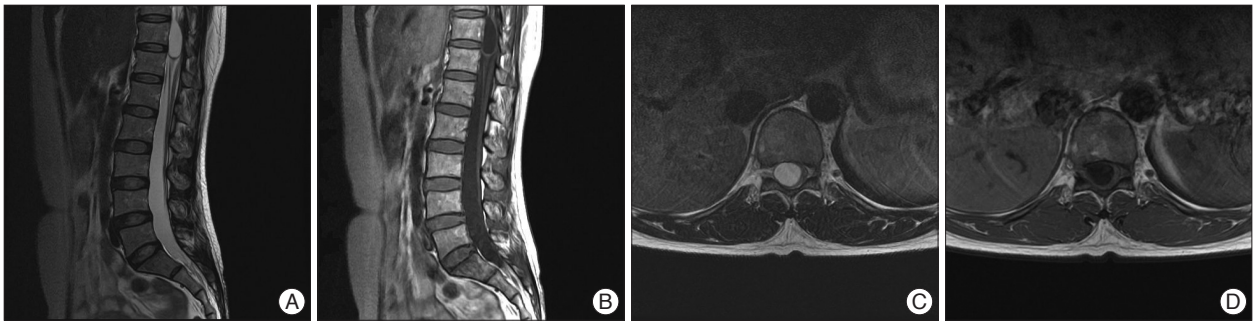

Fig. 1. The magnetic resonance image revealing a well-defined cystic lesion on the ventral side of the spinal cord at the T11 to T12 levels. The lesion is a 36×15 mm oval intramedullary cystic mass. The lesion is hyperintense in T2-weighted images (A and C) and hypointense in T1-weighted images (B and D). In the axial imaging, the cyst is observed to compress the spinal cord on the left dorsal side (C and D).

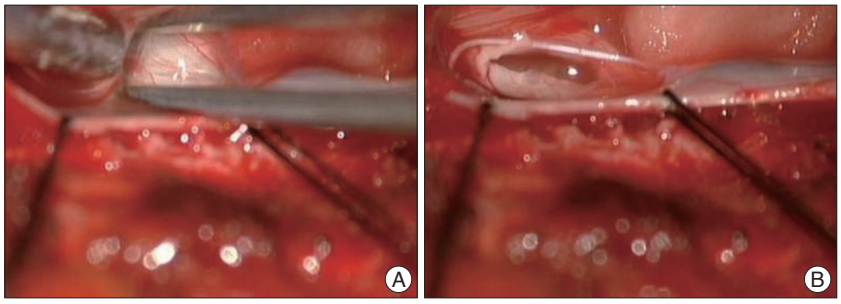

Fig. 2. A glistening white cyst without hemorrhage or parenchyma (A). The cyst wall is fenestrated to obtain adequate communication between the cyst and the subarachnoid space (B).

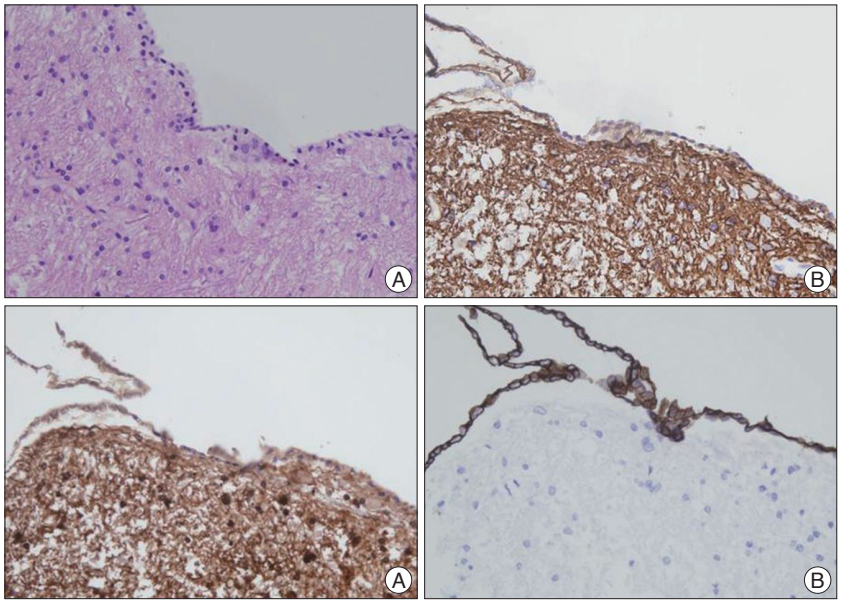

Fig. 3. In the H&E staining (A), the cyst wall consists of glial cells lined by a simple cuboidal to columnar epithelium. An immunohistochemical examination of the cells lining the cyst wall is positive for glial fibrillary acidic protein (B), S-100 protein (C), and cytokeratin (D). These findings are consistent with an ependymal cyst diagnosis.

a follow-up MRI revealed that the cystic lesion had diminished to 29×9.6 mm and that the cord deformity had improved. There was no new signal change in the adjacent spinal cord (Fig. 4). A histological study of the surgical specimens revealed that the cyst wall consisted of glial cells lined by a simple cuboidal to columnar epithelium. An immunohistochemical examination of the cells lining the cyst wall was positive for S-100 protein, glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), and cytokeratin. These findings were consistent with an ependymal cyst diagnosis (Fig. 3).

DISCUSSION

Ependymal cells line the ventricular spaces and the central canal. They have a simple ciliated cuboidal morphology. During spinal cord development, the walls of the recently closed neural tube consist of neuroepithelial cells. These cells extend over the entire thickness of the wall and form a thick pseudostratified epithelium. They are connected at their lumens by junctional complexes. They divide rapidly during the neural groove stage and immediately after tube closure. During this time period, these cells produce an increasing number of epithelial cells. Collectively, they constitute the neuroepithelial layer[9]. Once the neural tube closes, the neuroepithelial cells begin to give rise to another cell type that is characterized by a large round nucleus with a pale nucleoplasm and a dark-staining nucleolus. These cells are the primitive nerve cells or neuroblasts. Neuroepithelial cells following the other pathway differentiate into ependymal cells, which line the ventricular spaces and the central canal[9].

The most widely accepted hypothesis regarding the genesis of ependymal cysts holds that the floor plate of the neural tube is evaginated on the ventral side and becomes isolated to form an ependymal cyst[5,14,16]. The locations of the isolated ependymal cells determine whether the cyst presentation is intramedullary

or extramedullary[5]. Additionally, the nature of the cyst presentation may cause the cyst to be present anywhere along the cranio-spinal axis[5]. There have been frequent reports of extramedullary spinal ependymal cysts. However, intramedullary spinal ependymal cysts are rare lesions.

We have summarized the 18 pathologically proven ependymal cyst cases, including our case of a 46-year-old female, in Table 1. There is female predominance (11 females and 7 males), and a wide range of ages (from one to 71 years) is represented. The ependymal cysts are mostly located in the conus medullaris[10,14]. Nine cases concern the thoracic spine, 3 cases the thoraco-lumbar junction, 3 cases the cervical spine, 1 case the cervico-thoracic junction

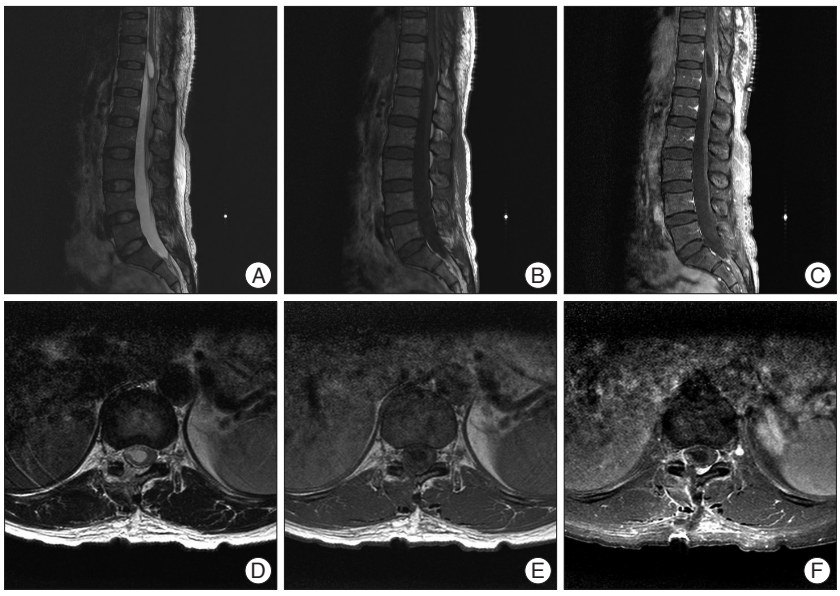

Fig. 4. A postoperative magnetic resonance image reveals that the cystic lesion's size had decreased to 29×9.6 mm and that the spinal cord had decompressed.

Table 1. A summary of the reported intramedullary ependymal cyst cases

No.	Year	Author	Age/Sex	Clinical presentation	Duration	Location	Op. type*	Result/Recurrence†
1	1979	Dharker et al.[2]	38/F	Backache, paraparesis	1 month	T6	Op	No improvement
2	1983	Rousseau et al.[13]	71/F	Radicular pain	53 years	T2	Op	Little improvement
3	1983	Fortuna and Mercuri (case 1)[4]	67/F	Intermittent paresis	5 years	T12	Op	Self-sufficient with crutches
4	1983	Fortuna and Mercuri (case 2)[4]	57/F	Medullary pain	6 years	T12	Op	Recovery
5	1985	Findler et al.[3]	6/M	Cervical pain and weakness	3 weeks	C7-T1	Op	Complete recovery
6	1987	Sharma et al.[15]	7/M	Urinary hesitancy and paraparesis	1 year	T4	R	Complete recovery
7	1991	Pagni et al.[11]	39/M	Paresthesia of the C6-C7 dermatomes	-	C5-C6	R	Self-sufficient with crutches
8	1991	Robertson et al. (case 1)[12]	48/F	Paresthesia	3 years	T12-L1	Op	Little improvement
9	1999	Iwahashi et al.[5]	1/F	Weakness	1 month	T6	Op	Complete recovery
10	2001	Kumar et al. (case 1)[7]	5/M	Excessive falls	-	T4-T6	R	Recovery
11	2001	Kumar et al. (case 2)[7]	4/M	Neck pain and weakness	5 months	C1-C5	Op	Improvement
12	2003	Chhabra et al.[1]	15/M	Quadriparesis	-	C2-C3	Op	Recovery/recurrence
13	2005	Saito et al.[14]	44/F	Weakness	1 year	T11-L1	Op+shunt	Complete recovery
14	2006	Takci et al.[16]	33/M	Back pain and LE paresthesia	2 months	L3	Op	Gradual improvement
15	2006	Lalitha et al.[8]	8/F	LE pain and gait difficulty	7days	T2-T3	R	Complete recovery
16	2008	Kato et al.[6]	31/F	LBP and numbness in the left leg	2 months	L2-L3	R	Complete recovery
17	2010	Nagano et al.[10]	64/F	Numbness and pain in both feet	5 years	T11-L1	Op	Complete recovery
18	2012	Park et al.	46/F	Left LE paresthesia	2 weeks	T11-T12	Op	Complete recovery

*Type of surgery, †Recurrence : case no. 12 was the only instance of cyst recurrence. R : resection of cyst, Op : biopsy or partial removal of the cyst wall, with opening and marsupialization of the cyst, shunt : cystosubarachnoid shunt, No : number, F : female, M : male, LE : lower extremity, LBP : low back pain, C : cervical, T : thoracic, L : lumbar

and 2 cases the lumbar spine. Based on the location and size of the lesion, we have reported a variable clinical presentation, including intermittent paresis of the lower limbs, radicular pain, paresthesia, and quadriparesis.

MRI is the best method for evaluating spinal cord cysts. The borders of the cyst appear to be smooth and well defined. Ependymal cysts are iso-intense with CSF in T1- and T2-weighted and proton density images without contrast-enhanced lining[5,10,14]. However, a differential diagnosis among the various types of intramedullary cystic lesions[4,5,10] is difficult to make based on MRI findings alone[11]. Therefore, pathological diagnosis is mandatory. A conventional histological examination is not sufficient to differentiate between ependymal and endodermal cysts because their microscopic morphologies are often similar[6]. Therefore, electron microscopy or immunohistochemical staining is necessary[6]. In an ependymal cyst, electron microscopy displays the ependymal character of the epithelium by showing intercellular junctions, membrane-bound granules in non-ciliated cells, and a lack of coating on the luminal surfaces of the cells[6]. In immunohistochemical staining, ependymal cysts are positive for GFAP and S-100[6,10]. Additionally, the cells lining the cyst wall stain positive for CAM5.2, AE1/AE3 keratin (low- and high-molecular-weight keratins), and EMA[6,10,14].

The surgical approaches to spinal intramedullary ependymal cysts include fenestration and resection of the cyst wall, placement of cyst-to-pleural space and cyst-to-subarachnoid space shunts, and biopsies of the cyst wall[1,5,6,10,11,14,16]. The aim of surgical treatment should be neural decompression and prevention of the cyst from refilling, which is best accomplished by completely resecting the cyst and closing the communication between the cyst and the subarachnoid space[6,16]. In our review of the literature, partial removal of the cyst wall or marsupialization of the cyst was performed in thirteen patients, whereas total enucleation was possible in five patients. Twelve of the 13 cases experienced gradual or complete functional improvement, and there was only one case of recurrence[1]. Because the cyst wall usually cannot be separated from the cord parenchyma[7,10], total excision of cyst can leads to neurologic deterioration. The only possible treatment may be to create communication between the cyst and the subarachnoid space by means of fenestration or marsupialization[7,10,14].

In our case, the cyst wall could be partially removed and biopsied, because there was no discernible plane between the cord parenchyma and the cyst wall. We agree that adequate decompression and communication between the cyst and subarachnoid space may be necessary and sufficient for ependymal cysts. However, re-accumulation of the cyst may be possible with fenestration or marsupialization; hence, these patients require close follow-up and imaging studies[5,7,16].

CONCLUSION

We report a rare spinal intramedullary ependymal cyst case. These cysts are benign in nature, and there is usually good functional recovery and a low recurrence rate after fenestration. Hence, we consider adequate decompression and communication between the cyst and subarachnoid space to be the optimal treatment for intramedullary ependymal cysts.

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