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.

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**Cervical Intramedullary Neurenteric Cyst in an Elderly Patient**

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**Introduction**

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**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.

**Examination**

On admission, vital sign was normal and there were no cutaneous stigmata on her back. On neurological examination, 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-C7 intramedullary lobulating cystic lesion slight hypointense 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-
Neurenteric Cyst

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

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Age(yrs)/ Sex</th>
<th>Other abnormality</th>
<th>Symptom</th>
<th>Location</th>
<th>Cyst contents</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knight et al, 1954</td>
<td>1/NR</td>
<td>Spina bifida</td>
<td>Anxiety, rigid neck</td>
<td>C7–T2</td>
<td>Milky fluid</td>
<td>Improved</td>
</tr>
<tr>
<td>Lalmaid et al, 1963</td>
<td>11/M</td>
<td>Scoliosis</td>
<td>Sensory disturbance</td>
<td>NA</td>
<td>NR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>5/M</td>
<td>Diastematomyelia</td>
<td>NA</td>
<td>NA</td>
<td>NR</td>
<td>Improved</td>
</tr>
<tr>
<td>Rewcastle &amp; Hancock, 1964</td>
<td>34/F</td>
<td>Normal</td>
<td>Paraparesis</td>
<td>T7–L2</td>
<td>Blackish fluid</td>
<td>Improved</td>
</tr>
<tr>
<td>Silvemai &amp; Brown, 1972</td>
<td>12/M</td>
<td>Multiple hemivertebra</td>
<td>Tetraparesis</td>
<td>T2</td>
<td>Clear jelly</td>
<td>Improved</td>
</tr>
<tr>
<td>Kov &amp; Jeffrey, 1982</td>
<td>54/M</td>
<td>Hemi, Fusion vertebra</td>
<td>NA</td>
<td>T2–5</td>
<td>NR</td>
<td>Improved</td>
</tr>
<tr>
<td>Fortuna et al, 1983</td>
<td>47/F</td>
<td>Normal</td>
<td>L1-L2 paresis</td>
<td>T12–L1</td>
<td>NR</td>
<td>Improved</td>
</tr>
<tr>
<td>Takemi et al, 1984</td>
<td>4/F</td>
<td>Normal</td>
<td>Nape pain</td>
<td>C3–4</td>
<td>NR</td>
<td>Improved</td>
</tr>
<tr>
<td>Mizuno et al, 1988</td>
<td>6/M</td>
<td>Normal</td>
<td>UE paresis</td>
<td>C2–5</td>
<td>Clear jelly</td>
<td>Improved</td>
</tr>
<tr>
<td>Rao et al, 1996</td>
<td>14/M</td>
<td>Normal</td>
<td>Neck pain</td>
<td>C2–5</td>
<td>CSF-like</td>
<td>Improved</td>
</tr>
<tr>
<td>Rivierez et al, 1997</td>
<td>27/F</td>
<td>Normal</td>
<td>Neck pain</td>
<td>C3–T1</td>
<td>NR</td>
<td>Improved</td>
</tr>
<tr>
<td>Lippman et al, 2000</td>
<td>46/M</td>
<td>Normal</td>
<td>LBP</td>
<td>L1–2</td>
<td>Clear colorless</td>
<td>Improved</td>
</tr>
<tr>
<td>Shinghal et al, 2001</td>
<td>67/M</td>
<td>Normal</td>
<td>Paraparesis</td>
<td>T7</td>
<td>Milky fluid</td>
<td>Improved</td>
</tr>
<tr>
<td>Rhee et al, 2001</td>
<td>16/M</td>
<td>Normal</td>
<td>Axillary pain, L1 paresis</td>
<td>T3–6</td>
<td>Milky mucinous</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>17/M</td>
<td>Normal</td>
<td>LBP</td>
<td>T3–6</td>
<td>Milky</td>
<td>Improved</td>
</tr>
<tr>
<td>Agrawal et al, 2002</td>
<td>3mon/M</td>
<td>Normal</td>
<td>Paraplegia</td>
<td>C3</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Kumar et al, 2002</td>
<td>3cases below 18yrs</td>
<td>Normal</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Paolini et al, 2003</td>
<td>28/F</td>
<td>Sacral schisis</td>
<td>L1 numbness</td>
<td>T8–9</td>
<td>Milky</td>
<td>Improved</td>
</tr>
<tr>
<td>Present case, 2004</td>
<td>72/F</td>
<td>Normal</td>
<td>Paraparesis</td>
<td>C6–7</td>
<td>Clear jelly-like</td>
<td>Improved</td>
</tr>
</tbody>
</table>

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

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

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).
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. Bentley and Smith explained embryological aspects as split notochord theory. Rhaney and Barclay 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 abnormalities such as cleft vertebra, hemivertebra, spina bifida, absent or fused vertebra, or diastematomyelia, Klippel-Feil anomaly and with anomalies which may be fatal. Their occurrences 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. Clinical presentation more than 50 years of age are very rare. 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. 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. The clinical features are in no way different from those of other space-occupying lesions of the spinal canal, the specific symptom depending solely on the level affected. The clinical courses may progress rapidly or intermittently be progressive. In some patients, symptomatic presentations is characterized episodically by exacerbation and remission, 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 cysts. An event of trauma may in-itiate or exacerbate symptoms in some patients. In our prese-nting case, paraparesis have shown patterns that waxed and waned for 5 years. There were neither histories of trauma or hemo-rhagic findings in operation. Causal mechanisms of
delayed symptomatic progression in old ages such as our case are vague.

In cases more than 50%, these lesions were 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 is rare\(^{1,9,14,16,17}\). Moreover, intramedullary neurenteric cysts not associated with other congenital abnormalities are very rare\(^{13,16}\). In domestic literatures, 1 case\((16 \text{ years old male})\) 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\(^3,10\), increases 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\(\text{(particularly protein contents)}\) of the intra-cavitary fluid\(^{3,17}\). 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\(^{1,9,14,17}\), 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\(^{1,14,19}\). In these approaches, 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.

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**References**