Craniospinal Neurenteric Cysts: Various MR Imaging Features

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Purpose: Craniospinal neurenteric (NE) cysts are rare developmental non-neoplastic cysts of the central nervous system with diverse MR imaging findings. The purpose of this study was to evaluate various MR imaging findings of intracranial and intraspinal NE cysts.

Materials and Methods: We retrospectively reviewed the MR imaging findings of six NE cysts that were confirmed by pathology. We evaluated anatomic location, signal intensity, size and enhancement pattern of NE cysts.

Results: Two intracranial lesions were located extra-axially in the cerebellopontine angle and quadrigeminal cisterns. Three spinal lesions were intradural-extradural cysts, located ventral to the spinal cord, but one thoracic lesion was an intramedullary cyst. The signal intensity of the cysts was hyperintense on T1-weighted images as compared with the cerebrospinal fluid (CSF) for two intracranial lesions and one cervical lesion. In addition, all intracranial lesions showed diffusion restriction. For the remaining three spinal lesions, the signal intensity was nearly the same as the signal intensity of the CSF as seen on both T1- and T2-weighted images. On contrast-enhanced studies, two intracranial cysts showed a small nodular enhancement and one thoracic spinal lesion showed rim enhancement.

Conclusion: NE cysts have various locations, signal intensities, and possible focal nodular or rim enhancement. Therefore, NE cysts can be included in the differential diagnosis of various craniospinal cystic or tumorous cystic lesions.

Index words: Neurenteric cyst
Magnetic resonance (MR)

Introduction

A neurenteric (NE) cyst is a rare benign endodermal lesion of the central nervous system (CNS) that results from developmental failure during or shortly after the third week of embryogenesis (1-3). The NE cyst is typically lined by a mucin producing, columnar or...
cuboidal epithelium and the cyst may contain other cells originating from the endodermal cell layer. Although a NE cyst can occur at any level of the craniospinal neuraxis, it is most commonly found in the lower cervical and upper thoracic levels. An intracranial NE cyst is less common as compared with an intraspinal lesion.

To date, a number of cases of patients with intracranial or intraspinal NE cysts have been reported (1–13). The majority of the previous reports concerning MR imaging were case reports, describing various imaging features. The rarity and various MR imaging features of a craniospinal NE cyst make the preoperative diagnosis difficult. In this report, we evaluated MR imaging findings of six pathologically proven craniospinal NE cysts.

**Materials and Methods**

Of eight cases of NE cysts collected from the

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**Fig. 1.** A 25-year-old female presented with headache.

a. An axial T1-weighted image shows a well-defined lesion of hyperintensity in the left cerebellopontine angle containing a small nodular hypointensity (arrow).
b. On an axial T2-weighted image, the mass shows hypointensity
c. A gadolinium-enhanced axial T1-weighted image shows no enhancement in the hyperintensity area but a small focal enhancement within the hypointense area (arrow).
d & e. On a diffusion-weighted image (d) and ADC map (e), the lesion shows mild to moderate diffusion restriction.
f. A pre-contrast CT image shows diffuse hyperattenuation of the lesion.
pathological archives of our institution from January 1996 to May 2007, six cases of NE cyst were included in this study. Two cases were excluded, as preoperative MR images were not available. There were three male and three female patients and the mean patient age was 26 years (age range, 3–52 years). Patients preoperatively had nonspecific various neurological symptoms including dizziness, headache, motor weakness and back pain. All patients underwent surgical excision of the lesions. Pathological diagnosis was made on light microscopic findings and by additional immunohistochemical studies for two intracranial lesions. Also, two intracranial and spinal lesions with atypical MR imaging feature were performed imaging and pathologic correlation.

MR imaging was performed using 1.5-Tesla MR imaging systems (Sonata, Siemens, Erlangen, Germany, or Signa Horizon and Genesis Signa, GE Medical Systems, Milwaukee, WI USA). Spin-echo T1-weighted images (T1WI) and fast spin-echo T2-weighted images (T2WI) were obtained in all patients. Contrast-enhanced T1WI images were obtained in five patients after intravenous injection of gadopentetate dimeglumine (Magnevist, Schering, Berlin, Germany). Echo-planar diffusion-weighted images (DWI) with b = 1,000 s/mm² and apparent diffusion coefficient (ADC) maps were obtained in two patients with intracranial lesions. All MR images were retrospectively reviewed.

Fig. 1. g. Histopathological findings indicate that the cyst is lined by stratified squamous epithelium [a] but a pseudostratified ciliated columnar epithelium lined area is also present [b]. The lining epithelial cells are immunoreactive for pancytokeratin [c] and carcinoembryonic antigen [d], suggesting an endogenous origin of the epithelium.
in terms of the location, size, signal intensity and presence or absence of contrast enhancement. For two intracranial cases, CT attenuation was compared with the MR signal intensity.

**Results**

Table demonstrates the MR imaging features and locations of the 6 patients with NE cyst.

All cysts appeared as well demarcated, smoothly marginated round or ovoid masses. One intracranial lesion in the quadrigeminal cistern was a multiloculated cyst. The sizes of cysts varied from $0.8 \times 1.0 \times 1.5$ cm to $2.5 \times 3.9 \times 4.4$ cm (mean longest diameter: 3.17 cm). The intracranial cysts were larger than the intraspinal ones (mean 3.8 cm versus 2.9 cm).

The signal intensity of the cysts was diverse. Cysts were hyperintense on T1WI and hypointense to cerebrospinal fluid (CSF) on T2WI for two intracranial lesions (Figs. 1, 2) and one cervical lesion. The two intracranial lesions contained a small nodule isointense to the brain parenchyma on T1W2. For the remaining

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**Fig. 2.** A 52-year-old male presented with dizziness.

- **a.** An axial T1-weighted image shows a well-defined, multiloculated mass of hyperintensity with an isointense nodule (arrow) in the quadrigeminal cistern.
- **b.** On an axial T2-weighted image, the portion of T1 hyperintensity shows isointensity to the brain parenchyma and the nodule of T1 isointensity shows heterogeneous signal intensity.
- **c.** A gadolinium-enhanced axial T1-weighted image shows a focal enhancement within the isointense nodule (arrow).
- **d & e.** On a diffusion-weighted image (d) and ADC map (e), the main lesion shows low and high intensity, respectively, indicating high diffusivity.
- **f.** A pre-contrast CT image shows hyperattenuation of the main cystic mass and a tiny calcification in the solid nodular portion (arrow).
three spinal lesions, the signal intensity was nearly the same as the signal intensity of the CSF on both T1WI and T2WI (Figs. 3, 4). Contrast-enhanced T1WI studies which were undertaken in five patients showed a small focal nodular enhancement in two intracranial lesions (Fig. 1c, Fig. 2c) and a rim enhancement in one thoracic spinal lesion (Fig. 4c). The focal small nodular enhancing portion, as noted in the two intracranial lesions is likely to correspond to focal fibrous tissue and/or a xanthogranulomatous change in the histopathological findings. The other three spinal lesions showed no contrast enhancement. One cyst in

![Fig. 3. A 22-year-old male with progressive weakness of the lower extremities.](image)

- a. A sagittal T1-weighted image shows a well-defined, intramedullary mass of hypointensity in the spinal cord at the T4-T7 level.
- b. On a T2-weighted image, the expansile cystic mass is homogeneously hyperintense. There was no enhancement in the lesion on contrast-enhanced T1-weighted images [not presented].

![Fig. 4. A 38-year-old female with lower extremity weakness and pain.](image)

- a. A sagittal T1-weighted image shows a well-defined, intradural mass hypointense to the spinal cord at the T6-7 level, closely attached to the anterior surface of the spinal cord.
- b. A sagittal T2-weighted image shows the prominent hyperintensity of the cyst content with hypointense rim along the cyst-cord interface.
- c. On a gadolinium-enhanced T1-weighted image, the cyst has thick irregular peripheral rim enhancement. At surgery, the cyst was found to be intradural-extramedullary in location and firmly adhered to the spinal cord. Note kyphotic deformity and blocked vertebra of T6-T7.
the cerebellopontine angle showed isointensity to the brain parenchyma on DWI and slight hypointensity on ADC maps, indicating moderate diffusion restriction (Fig. 1d, e). The other lesion in the quadrigeminal cistern area was seen as isointense to the CSF on DWI and ADC map, indicating high diffusivity (Fig. 2d, e).

Pre-contrast CT scans obtained in two intracranial cases showed high attenuation corresponding to the T1 hyperintensity on MR. One case had a tiny calcification within the focal nodular enhancing portion (Fig. 2f). Two spinal lesions were associated with a vertebral kyphotic deformity. Table 1 summarizes the MR imaging findings.

### Discussion

Although the precise etiology of an NE cyst is unknown, an NE cyst is a congenital abnormality from an abnormal connection between the primitive endoderm and ectoderm during the third week of life. An NE cyst arises secondary to the persistence of the neurenteric canal, which is the temporary connection between the amniotic and yolk sacs. Persistent endoectodermal adhesions or a persistent adhesion between the notochord and the endoderm may produce notochodal dysgenesis and an NE cyst (14).

More than 80% of NE cysts are located in the spine, usually intradural-extramedullary, ventral to the spinal cord at the cervical and thoracic levels, whereas only 10% to 15% are intracranial. There are far fewer spinal intramedullary lesions than intradural-extramedullary lesions [1, 2, 5, 7, 8, 12, 18]. An intracranial lesion is most often found in the posterior fossa [8, 11, 18, 19]. It is typically midline, anterior to the brain stem [9] or in the cerebellopontine angle [10, 20, 21]. It can occur less commonly in the quadrigeminal cistern, as shown in our case 2 (Fig. 2). Supratentorial [2, 15, 22] and suprasellar [16] NE cysts are extremely rare. Since a NE cyst, along with a Rathke’s cleft cyst and colloid cyst, are endodermally derived lesions of the CNS, some investigators have suggested the possibility of the same origin for a Rathke’s cleft cyst, colloid cyst, and suprasellar NE cyst. There are no pathological or immunohistochemical criteria for a distinction between a Rathke’s cleft cyst, colloid cyst, and an NE cyst. These cysts can only be distinguished by their location. Rathke’s cleft cyst is located between anterior pituitary gland and posterior gland, and location of colloid cyst is the foramen of Monro (16).

An NE cyst can occur at any age. Although an intraspinal NE cyst has a slight male predominance (17) and an intracranial cyst has a slight female predominance (18), the actual prevalence of an NE cyst is likely to be independent of gender. The clinical presentation of an NE cyst is non-specific. It is frequently detected at adulthood, with compressive symptoms due to cyst growth with fluid accumulation, and it may cause an infectious focus of the CNS.

MR signal intensity characteristics of NE cysts vary depending upon the protein content of the cyst fluid. Most spinal NE cysts are isointense to the CSF on both T1WI and T2WI, indicating a low protein content. In our series, three of four spinal lesions showed the same signal intensity as that of the CSF on both T1WI and T2WI. Most intracranial NE cysts, however, appear to have a different signal intensity from that of most spinal NE cysts, suggesting a higher protein content in intracranial NE cysts. Preece et al. [2] reported that most intracranial NE cysts (for 16/18 cases) were proteinaceous with a hyperintense signal on T1WI and 11% of intracranial NE cysts were hypointense on T2WI. In our series, three of six (50%) NE cysts (two

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/gender</th>
<th>Location</th>
<th>Size [cm]</th>
<th>Signal intensity</th>
<th>Associated anomaly</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>T1WI T2WI DWI CE-T1WI</td>
<td></td>
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<tr>
<td>1</td>
<td>25/F</td>
<td>Rt.Cerebellopontine angle</td>
<td>2.5 × 2.1 × 3.2</td>
<td>High Low iso</td>
<td>Nodular No</td>
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<tr>
<td>2</td>
<td>52/M</td>
<td>Quadrigeminal cistern</td>
<td>4.4 × 3.9 × 2.5</td>
<td>High Low low</td>
<td>Nodular No</td>
</tr>
<tr>
<td>3</td>
<td>20/M</td>
<td>C1-2, IDEM</td>
<td>0.8 × 1.8</td>
<td>High Low NP</td>
<td>NP Kyphosis</td>
</tr>
<tr>
<td>4</td>
<td>3/F</td>
<td>C2-5, IDEM</td>
<td>2.0 × 1.3 × 0.9</td>
<td>Low High NP</td>
<td>No enhancement No</td>
</tr>
<tr>
<td>5</td>
<td>22/M</td>
<td>T4-7, IM</td>
<td>7.4 × 1.8 × 1.1</td>
<td>Low High NP</td>
<td>No enhancement No</td>
</tr>
<tr>
<td>6</td>
<td>38/F</td>
<td>T6-7, IDEM</td>
<td>1.4 × 1.3 × 1.9</td>
<td>Low High NP</td>
<td>Rim Kyphosis</td>
</tr>
</tbody>
</table>

Note: IDEM = intradural extramedullary, IM = intramedullary, NP = not performed

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Craniospinal Neurenteric Cysts

Table 1. Summary of the MR Imaging Findings in Six Patients with Craniospinal Neurenteric Cysts
intracranial cysts and one intraspinal cyst) were hyperintensity on T1WI and hypointensity on T2WI. The T1 hyperintensity of the cyst is suggestive of not only a highly proteinaceous content but also hemorrhage. However, at surgery, the cyst content was yellowish or light greenish, turbid, mucoid fluid and there was no hemorrhage within the cysts in the three cases of our series. On contrast-enhanced MR imaging, a NE cyst appears usually not enhanced or rarely enhanced. Enhancement rates have been reported from 0% to 35.7%. The enhancement pattern was usually partial or complete rim enhancement [1-4]. The focal small nodular enhancing portion, as noted in the two intracranial lesions (Figs. 1c, 2c), is likely to correspond to focal fibrous tissue and/or a xanthogranulomatous change in the histopathological findings. A xanthogranulomatous change is a nonspecific chronic cellular reaction with prominent lipid-laden histiocytes or macrophages, so-called foam or xanthoma cells. It can occur in benign, noninflammatory cystic or cystlike lesions including a neurenteric cyst, colloid cyst, epidermoid and choroids plexus cyst [21]. For the two intracranial lesions in the present study, microscopic histopathological findings showed that the cysts were composed of fibrous tissue and xanthogranulomatous reaction including foamy histiocytes, chronic inflammatory cells, multinucleated giant cells and cholesterol clefts. A rim enhancement pattern, as noted in case 6, is very rare even for intraspinal NE cysts. We found only two cases described in the literature [1, 12]. In case 6 of our series, microscopic histology showed a smooth muscle and dense collagenous tissue with chronic inflammatory cells within the cyst wall. The rim enhancement around the cyst might reflect some chronic inflammatory reaction of the spinal cord tightly adhered to the cyst. DWI signal intensity of an NE cyst has seldom been reported [2]. Preece et al. [2] reported that DWI obtained in the two intracranial cases showed no diffusion restriction in one case and showed mild diffusion restriction in the other case. DWI findings in the two cases in the present study are similar to those cases. As noted in case 2, a tiny calcification may be detected on the CT scan [22].

The differential diagnosis of intracranial NE cysts includes an epidermoid cyst, dermoid cyst, arachnoid cyst, parasitic cyst and cystic neoplasm. In the differential diagnosis of an intraspinal NE cyst, an ependymal cyst and focal syringomyelia would be additionally included. It usually is not difficult to differentiate from other lesions based on the MR imaging findings including the location, signal characteristics including DWI, size, shape, and contrast-enhancement pattern of the lesion. [2, 3, 26, 27]. It is most difficult or even not possible to differentiate between an NE cyst and "white" epidermoid cyst, when an extraxial cystic mass in the posterior fossa has T1 high intensity as seen on MR imaging. In that case, an immunohistochemical study may be helpful for the differential diagnosis between the two entities, as discussed below.

On a histopathological examination, an NE cyst is usually lined by a mucin-producing nonciliated epithelium that is simple or pseudostratified (columnar or cuboidal). The cyst can be ciliated or can have a mixture of gastrointestinal, pancreatic and/or squamous epithelium [1]. In our series, five NE cysts were lined with a ciliated columnar epithelium, stratified cuboidal to the transitional epithelium and/or squamous metaplasia. One case (case 2) exhibited the presence of dense collagenous tissue with focal loose myxoid fibrous tissue without a lining epithelium. In this case, the initial pathological diagnosis was a 'probable' epidermoid cyst. The patient underwent a second surgery for the recurrent cystic mass in the same area of the quadrigeminal cistern. At the first and second surgeries, the cyst content was light greenish, turbid, mucoid fluid, unlike the usual 'pearl-like' or 'soapflake-like' contents of an epidermoid cyst. The pathologist additionally performed an immunohistochemical study, resulting in the detection of a positive reaction to carcinoembryonic antigen (CEA) and cytokeratin. The final diagnosis was revised to a NE cyst. Immunohistochemical studies have an important role to differentiate endodermal lesions such as an NE cyst, Rathke's cyst and colloid cyst from ectodermal lesions such as an epidermoid cyst. An NE cyst usually demonstrates a positive reaction for CEA, whereas an epidermoid cyst is usually negative for the presence of CEA [16]. It could be assumed that a rare "white" epidermoid might have actually been an NE cyst, if an extraxial cyst in the posterior fossa had T1 high intensity on MR imaging, the cyst content was seen with turbid, mucoid fluid at surgery, and the pathological diagnosis was made by only light
microscopy. In that case, an immunohistochemical study may be desirable. An NE cyst may also show a positive reaction for epithelial membrane antigen (EMA) but will show a negative reaction to glial fibrillary acidic protein (GFAP), neuron-specific enolase (NSE) and vimentin (28).

The standard treatment for an NE cyst is surgical removal of the cyst wall. The location of a cyst determines the feasibility of surgery, and prognosis depends on the size and location of the cyst (23). Tight adherence of the cyst wall to the neural structures does not permit its safe total removal. According to previous reports, the recurrence rates are variable (11.9-37%) and recurrent intracranial hypertension might be caused by intermittent leakage of the cyst contents from a recurrent tumor. This may be an important sign of tumor recurrence (1, 19). Unusually hemorrhage or extensive craniospinal spread are possible (24, 25). In addition, three cases of malignant transformation of NE cysts have been reported (19).

**Conclusion**

Although most NE cysts have typical MR imaging findings, some NE cysts, particularly cysts in the intracranial cavity, may show various imaging findings including an unusual location in the cerebellopontine angle or quadrigeminal cistern, various signal intensities and small focal nodular or rim enhancement, as shown in the present case series. Craniospinal NE cysts have various MR imaging findings. Therefore, NE cysts can include differential diagnosis of cystic mass in the intracranial and intraspinal area, not only typical findings but also atypical MR findings.

**References**

뇌척수 신경장관 낭종: 다양한 자기공명영상소견

한성호1,3, 손철호1, 김은희1,4, 손규리1,5, 박상현2, 장기현1

목적: 신경장관 낭종은 중추신경계에 발생하는 비 종양성, 발생학적 낭종으로 다양한 자기공명 영상소견을 보인다. 이 연구의 목적은 저자들이 경험한 두개내와 척수내의 신경장관 낭종의 다양한 자기공명 영상 소견을 보이고 설명하고자 한다.

대상과 방법: 저자들의 병원에서 경험한 여섯 명의 신경장관 낭종환자를 대상으로 하였으며, 이 환자들의 자기공명 영상에서 병변의 해부학적 위치, 병변의 신호강도, 병변의 크기 및 조영증강형태에 대해 후향적으로 분석하였다.

결과: 두 개의 두개강 내 병변은 소뇌교각과 사구체조 부위에 축 외 낭종으로 보였다. 세 개의 척추 부위 병변은 경막내-척수외 낭종으로, 척수의 범위를 벗어나지 않았고, 한 개의 흉추 병변은 척수 내 낭종이었다. 두 개의 두개강내 낭종과 한 개의 경추부 낭종의 신호강도는 T1 강조영상에서 고 신호강도이고, T2 강조영상에서 뇌척수액과 같은 저 신호강도로 보였으며, 두개강내 병변은 모두 중등도 이상의 확산제한을 보였고, 두개강내 병변은 모두 중등도 이상의 확산제한을 보였으며, 두개강내 병변은 모두 중등도 이상의 확산제한을 보였고, 한 개의 흉추 병변은 가장자리에 환상의 조영증강을 보였다.

결론: 신경장관 낭종은 다양한 위치에서 발생할 수 있고 부분적인 결절성 또는 환상의 조영증강을 보일 수 있다. 그러므로, 비전형적인 자기공명영상소견을 보일 경우, 다른 비종양성, 종양성 낭종의 감별진단에 포함될 수 있다.

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