Neurenteric Cyst of the Craniocervical Junction
—Case Report—

Kazuhiro ABE, Kazutaka OYAMA, Kentaro MORI, Sumio ISHIMARU, Masanobu EGUCHI*, and Minoru MAEDA

Departments of Neurosurgery and *Pathology, Juntendo University, Izunagaoka Hospital, Shizuoka

Abstract

A 60-year-old female presented with occipital headache and limitation of neck movement. Neurological examination showed weakness of the right sternocleidomastoid muscle. Magnetic resonance imaging revealed a cystic lesion at the craniocervical junction and posterior compression of the brain stem. The lesion was totally removed through the transcondylar approach. The histological diagnosis was neurenteric cyst. The transcondylar approach provides a direct operative view of the clivus and anterior craniovertebral junction.

Key words: neurenteric cyst, craniocervical junction, transcondylar approach

Introduction

Neurenteric cysts are classified by the World Health Organization as structures lined by mucin-secreting epithelium resembling that of the gastrointestinal tract and occurring predominantly in intraspinal locations. Neurenteric cysts are rare and usually occur in the lower cervical to upper thoracic spine. Intracranial neurenteric cysts are very rare and usually occur in the posterior fossa, but may also be located in the fourth ventricle, medulla, pons, cerebellopontine angle, and craniocervical junction. We describe a case of neurenteric cyst at the craniocervical junction, which was successfully removed through the transcondylar approach.

Case Report

A 60-year-old female suffered onset of occipital headache and limitation of neck movement 3 weeks before admission. On admission, she had nystagmus on right lateral gaze and downbeat nystagmus. Neurological examination revealed only weakness and atrophy of the right sternocleidomastoid muscle.

Radiography of the skull and neck showed no evidence of bony abnormalities. Computed tomography (CT) showed a low density nonenhanced mass at the craniocervical junction. CT cisternography revealed a filling defect in this area. T1-weighted magnetic resonance (MR) imaging showed an area isointense to cerebrospinal fluid (CSF) at the craniocervical junction which was not enhanced by gadolinium administration (Fig. 1). T2-weighted MR imaging showed this area as a high intensity mass. The medulla and upper cervical cord were displaced posteriorly. Vertebral angiography demonstrated an avascular mass in front of the brain stem and the basilar artery shifted to the right. Single photon emission computed tomography with thallium-201 chloride showed no accumulation of radioisotope in the lesion.

The right transcondylar approach with C-1 half laminectomy was performed with the patient in the lateral position. The dura was opened to reveal a cystic mass with a thin wall at the ventral surface of the medulla (Fig. 2). The medulla was displaced posteriorly and the 9th, 10th, and 11th cranial nerves were attached to the cyst wall. Yellowish lipomatous tissue was also seen on the cyst wall. Milky fluid was aspirated from the cyst (Fig. 3 left). The cyst wall at the ventral surface of the medulla was totally removed.

Histological examination showed that the cyst wall consisted of fibrocollagen walls lined by par-
tially ciliated columnar epithelium (Fig. 3 center). The nuclei were round and basally situated. No mitosis was noted. The yellowish lipomatous tissue on the cyst wall was formed of accumulated fat-laden cells (histiocytes) and was verified as degenerative change. Many cells contained periodic acid-Schiff (PAS)-positive mucin. Immunocytochemical staining showed a positive reaction in the epithelial cells for carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), and cytokeratin, but no reaction for glial fibrillary acidic protein (GFAP) and S-100 protein. Electron microscopy showed microvilli at the luminal surface but the microvilli were not covered with electron-dense granular coating material (Fig. 3 right). The cilium contained a $9 + 2$ complex.

The postoperative course was uneventful and she was discharged without pain or neurological deficit. MR imaging performed one year after the operation showed no evidence of tumor recurrence.

**Discussion**

The pathogenic embryogenesis of neurenteric cysts is not completely understood. Neurenteric cysts are thought to be derived from malformations involving the embryonal rests of primitive endodermal cells. Dysgenesis of endodermal tissue during the third week of embryonic life is considered to be the most likely origin. Neurenteric cyst is a type of epithelial-lined cyst, but the presence of ciliated epithelium does not exclude an endodermal origin, since embryonic esophageal epithelium is ciliated at an early stage of development. Central nervous system epithelial cysts can be divided into three types according to the origin: ependymal cyst, neurenteric cyst (enterogenous cyst), and teratomatous cyst.

Abnormal adhesion between the notochord and endoderm during the third week of embryological development may be the common etiological event in the formation of enterogenous anomalies. The frequent association of neurenteric cysts with anomalies of the mesoderm (vertebral defects) and neuroectoderm (spina bifida and split cord syndrome) suggests that neurenteric cysts are a manifestation of a more complex malformation involving all three germ layers. Neurenteric cysts, together with various other malformations including combined spina bifida, split cord malformation, and certain intestinal malformations, are referred to as the “split notochord syndrome” or “ectoderm-endoderm adhesion syndrome.” Therefore, this clinical entity covers a wide range of diseases from isolated neurenteric cyst to split notochord syndrome. Also, there is no clear demarcation between neurenteric cyst and teratomatous cyst.

Table 1 summarizes the clinical, radiological, and histological findings of the previous 14 cases of neurenteric cysts located at the craniocervical junction and our present case. The patients were aged 7 to 60 years (mean 38 years) and there was no significant difference in sex distribution. The most frequent initial symptom was neck pain or occipital headache (10/15 patients), sometimes accompanied by stiff neck or limitation of
neck movement. Motor weakness and ataxia were the most frequent neurological findings (8/15 patients). Cranial nerve paresis was recognized in 4/15 patients. CT showed a low density non-enhanced mass at the craniocervical junction in all patients except one who had intracystic hemorrhage. T1-weighted MR imaging of 11 patients found an iso- to slightly hyperintensity mass located at the ventral to the neural axis in most patients. Three cysts appeared as high signal intensity. The MR imaging appearance of neurenteric cyst content may indicate fluid similar to CSF, with highly proteaceous fluid or keratin-containing and fat-like contents similar to epidermoid cysts. Most cysts contained white- to yellow-colored fluid, described as milky, turbid, or straw-colored fluid. The brain stem and upper cervical cord were displaced posteriorly, as seen with neurenteric cysts located in the spinal cord. Bony anomalies are associated with half of the neurenteric cysts in the spinal cord, but were found in only two patients with neurenteric cyst at the craniocervical junction. Bony anomaly is rarely associated with intracranial neurenteric cyst.

Immunohistochemical staining of the epithelial cells was positive for PAS, cytokeratin, EMA, and CEA, and negative for GFAP and S-100 protein. These results are completely identical to those for neurenteric cysts at other sites. EMA is a non-specific marker for cysts of epithelial origin (neurenteric cyst, ependymal cyst, choroid cyst, Rathke’s cleft cyst). CEA is useful for the diagnosis of neurenteric cyst. Positive staining for CEA reflects the fact that gastrointestinal epithelial cells contain CEA from the second to the sixth month of fetal

Fig. 2 Intraoperative photograph showing the thin-walled cystic lesion. The glossopharyngeal (IX), vagal (X), and accessory (XI) nerves are displaced superiorly. Yellowish lipomatous tissue is seen on the cyst wall (arrow).

Fig. 3 left: Photograph of the milky fluid from the cyst. center: Photomicrograph of the cyst specimen showing columnar epithelial cells with cilia. HE stain, ×200. right: Electron micrograph of the epithelial cells showing microvilli on the luminal surface. Bar = 1 µm.
Table 1 Cases of neurenteric cyst at the craniocervical junction

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/ Sex</th>
<th>Symptoms</th>
<th>CT</th>
<th>MR imaging</th>
<th>Cyst location to neuraxis</th>
<th>Bony anomaly</th>
<th>Operation</th>
<th>Cyst content</th>
<th>Adhesion</th>
<th>Cytochemical histology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Fabinyi and Adams (1979)²</td>
<td>54/F</td>
<td>neck pain, tetraparesis, sensory disturbance</td>
<td>LD</td>
<td>—</td>
<td>ventral</td>
<td>none</td>
<td>partial removal</td>
<td>gelatinous</td>
<td>anterior aspect of medulla</td>
<td>PAS (+)</td>
<td>good</td>
</tr>
<tr>
<td>2</td>
<td>Hirai et al. (1981)¹³</td>
<td>30/F</td>
<td>headache</td>
<td>LD</td>
<td>—</td>
<td>ventral</td>
<td>none</td>
<td>total removal</td>
<td>turbid fluid</td>
<td>none</td>
<td>PAS (+)</td>
<td>good</td>
</tr>
<tr>
<td>3</td>
<td>Hasegawa et al. (1988)¹¹</td>
<td>42/M</td>
<td>occipitalgia, hemiparesis, sensory disturbance</td>
<td>LD</td>
<td>slightly hyper-intense than CSF</td>
<td>ventral</td>
<td>ND</td>
<td>total removal (lateral)</td>
<td>milky fluid</td>
<td>anterior aspect of cervical cord</td>
<td>ND</td>
<td>good</td>
</tr>
<tr>
<td>4</td>
<td>Koksel et al. (1990)¹⁶</td>
<td>40/M</td>
<td>neck pain, nystagmus, tetraparesis, sensory disturbance</td>
<td>ND</td>
<td>CSF intensity</td>
<td>ventral</td>
<td>ND</td>
<td>total removal (transoral)</td>
<td>dark yellow fluid</td>
<td>ND</td>
<td>cytokeratin (+), CEA (+), GFAP (–)</td>
<td>good</td>
</tr>
<tr>
<td>5</td>
<td>Breeze et al. (1990)³</td>
<td>37/M</td>
<td>neck pain</td>
<td>LD</td>
<td>higher than CSF</td>
<td>ventral</td>
<td>ND</td>
<td>subtotal removal (suboccipital + laminectomy)</td>
<td>straw-colored fluid</td>
<td>anterior aspect of cervical cord</td>
<td>PAS (+), cytokeratin (+), GFAP (–)</td>
<td>good</td>
</tr>
<tr>
<td>6</td>
<td>Malcolm et al. (1991)²³</td>
<td>57/F</td>
<td>headache, vertigo, ataxia, VI paresis</td>
<td>LD</td>
<td>—</td>
<td>ventral</td>
<td>ND</td>
<td>partial removal (suboccipital + laminectomy)</td>
<td>viscous yellow material</td>
<td>lower cranial nerves</td>
<td>cytokeratin (+), EMA (+), GFAP (–)</td>
<td>recurrence (+)</td>
</tr>
<tr>
<td>7</td>
<td>Ito et al. (1992)¹⁴</td>
<td>31/M</td>
<td>III, V, and VI paresis, ataxia</td>
<td>HD (bleeding)</td>
<td>high intensity (bleeding)</td>
<td>ventral</td>
<td>ND</td>
<td>partial removal (suboccipital)</td>
<td>dark brown (hemorrhage)</td>
<td>brain stem</td>
<td>PAS (+), cytokeratin (+), EMA (+), GFAP (–)</td>
<td>good</td>
</tr>
<tr>
<td>8</td>
<td>Harris et al. (1991)¹⁰</td>
<td>30/F</td>
<td>occipitalgia, V and VI paresis, ataxia</td>
<td>LD</td>
<td>—</td>
<td>ventral</td>
<td>ND</td>
<td>total removal (suboccipital)</td>
<td>clear fluid</td>
<td>none</td>
<td>cytokeratin (+), S100 (–)</td>
<td>good</td>
</tr>
<tr>
<td>9</td>
<td>Menezes and Ryken (1995)²⁵</td>
<td>58/F</td>
<td>occipitalgia, vertigo, ataxia</td>
<td>ND</td>
<td>slightly hyper-intense than CSF</td>
<td>ventral</td>
<td>ND</td>
<td>total removal (suboccipital)</td>
<td>turbid fluid</td>
<td>none</td>
<td>cytokeratin (+), S100 (–)</td>
<td>good</td>
</tr>
<tr>
<td>10</td>
<td>Ito et al. (1992)¹⁴</td>
<td>24/M</td>
<td>hearing loss, nystagmus</td>
<td>LD</td>
<td>slightly hyper-intense than CSF</td>
<td>ventral</td>
<td>ND</td>
<td>total removal (suboccipital)</td>
<td>clear fluid</td>
<td>anterior aspect of medulla</td>
<td>PAS (+), GFAP (–)</td>
<td>good</td>
</tr>
<tr>
<td>11</td>
<td>Lazareff and Parra (1995)¹⁸</td>
<td>8/F</td>
<td>recurrent meningitis</td>
<td>LD</td>
<td>hyper-intense</td>
<td>ventral</td>
<td>bifid clivus</td>
<td>ND (suboccipital)</td>
<td>ND</td>
<td>ND</td>
<td>good</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Lazareff and Parra (1995)¹⁸</td>
<td>7/F</td>
<td>neck pain, torticollis, quadriparesis</td>
<td>ND</td>
<td>hyper-intense than CSF</td>
<td>ventral</td>
<td>bifid C-7 lamina</td>
<td>total removal (laminectomy)</td>
<td>xanthochromic fluid</td>
<td>none</td>
<td>cytokeratin (+), GFAP (–)</td>
<td>good</td>
</tr>
</tbody>
</table>

Contd.
Positive cytokeratin and negative GFAP staining can differentiate neurenteric cysts from ependymal cysts, which are also lined by columnar cells and are positive for mucin immunostaining.\(^8,14\)

Neurenteric cysts are basically located in front of the neural axis. The cyst walls frequently adhere to the anterior surface of the brain stem or upper cervical cord and lower cranial nerves, so complete removal is sometimes difficult. Neurenteric cysts may expand by cyst epithelial secretion or CSF accumulation by osmosis. Recurrence of the cyst may reflect osmosis after scarring and resealing of the remnant cyst wall.\(^25\) Therefore, the surgical treatment of intracranial neurenteric cysts should aim at total resection.\(^7,10\) Several approaches have been advocated to resect neurenteric cysts at the craniocervical junction, including the suboccipital approach with or without laminectomy and the transoral approach. The transoral approach is recommended because the cysts are located in the midline and attached to the anterior surface of the brain stem,\(^7\) but the transoral approach provides a limited operative field and carries the risk of postoperative infection. The suboccipital approach limits the operative view in the anterior surface of the brain stem. We chose the transcondylar approach and succeeded in complete removal of a neurenteric cyst located at the craniocervical junction. We recommend the transcondylar approach with partial removal of the occipital condyle and the jugular tubercle for surgical removal of neurenteric cyst at the craniocervical junction because this approach provides a direct operative view of the clivus and anterior craniovertebral junction.\(^9,20\)

### References

6. Fujita T, Kayama T, Saito S, Yamakawa M, Nakai O:


Address reprint requests to: K. Mori, M.D., Department of Neurosurgery, Juntendo University, Izunagaoka Hospital, 1129 Nagoaka, Izunagaoka-cho, Tagata-gun, Shizuoka 410-2295, Japan.