A rare presentation of a pediatric neurenteric cyst as an intra-axial pontine lesion: A case report with a 5-year follow-up

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ABSTRACT

Background: Intracranial neurenteric cysts are rare, benign, and slow-growing tumors. However, we encountered a pediatric case that the cyst expansion occurred in a short period of time resulting in rapid deterioration of the patient's symptoms.

Case Description: A previously healthy 7-year-old girl had a week history of dysarthric speech and diplopia along with headaches. Her magnetic resonance images (MRI) showed an abnormal cystic mass in her brainstem. Her symptoms were deteriorated for 1 month and her second MRI revealed an enlargement of the cystic lesion. The tumor biopsy and cyst drainage were carried out and histopathological examination of the cyst wall showed columnar epithelium containing ciliated cells. The final diagnosis of her tumor was neurenteric cyst.

Conclusion: We report a pediatric case of a neurenteric cyst in the brainstem, which expanded in a short period, and review this rare entity.

Keywords: Brainstem cyst, Neurenteric cyst, Pediatric brain tumor, Pons

INTRODUCTION

Neurenteric cysts are rare benign lesions that typically arise within the thoracic spine but can be encountered anywhere in the central nervous system from the cranium to the coccyx. Although over 140 cases of intracranial neurenteric cysts have been reported to date, brainstem neurenteric cysts are rare. Here, we report a case of a neurenteric cyst in the brainstem of a 7-year-old girl, which expanded in a short period. Neurenteric cysts are generally slow growing and the mechanisms underlying cyst enlargement are not fully understood. We describe this rare case and discuss it in the context of the current literature on neurenteric cysts.

CASE DESCRIPTION

A previously healthy 7-year-old girl was referred to our hospital for an abnormal cystic mass in her brainstem found using magnetic resonance imaging (MRI). She had a week history of dysarthric speech and diplopia along with headaches. Neurological examinations revealed...
facial and abducens nerve palsy on her left side. The MRI data showed a cystic mass measuring 22 mm × 21 mm × 18 mm, located in the pons with no connection to adjacent cisterns. The cystic component appeared as hypointense on the T1-weighted images, hyperintense on the T2-weighted images, and isointense on the diffusion-weighted images [Figure 1a-c]. One month later, after the deterioration of her symptoms, as suggested by hoarseness due to vocal cord paralysis, the patient underwent the second MRI, which revealed an enlargement of the cystic lesion to 24 mm × 27 mm × 20 mm. Although gadolinium administration showed no enhancement of the cyst contents, a small enhancing nodule was visualized [Figure 1d-f]. It was thought to be a cystic tumor, such as a pilocytic astrocytoma or hemangioblastoma.

Due to the patient’s progressive clinical course, tumor biopsy and cyst drainage were scheduled. The patient was placed in the prone position, and midline suboccipital craniotomy was performed. After dividing the arachnoid over the cisterna magna and cerebellomedullary fissure, the bilateral cerebellar hemispheres were gently retracted. The floor of the fourth ventricle was covered with the ependymal layer that appeared normal [Figure 2a]. The yellowish-white cyst was detected through the supracollicular safe zone [Figure 2b]. The cyst was filled with milky white viscous fluid. After removing the cyst component by aspiration, the cyst wall and mural nodule were partially removed [Figure 2c]. Histopathological examination of the cyst wall showed columnar epithelium containing ciliated cells. Some goblet cells were seen within the epithelium. These findings were responsible for the mucinous content and suggested endodermal origin cyst. Therefore, we diagnosed a neurenteric cyst [Figure 2d].

The postoperative course was uneventful, and complete resolution of symptoms was observed after surgery. At the last follow-up (5 years after surgery), she was asymptomatic, and there was no evidence of recurrence on follow-up MRI [Figure 3].

**DISCUSSION**

The first neurenteric cysts were described by Kubie and Fulton, in 1928, as teratomatous cysts and later named neurenteric cysts in 1954 by Holcomb and Matson. \[4\] Although most neurenteric cysts occur primarily in the spine, they can appear anywhere along the spinal axis from the cranium to the coccyx. Intracranial neurenteric cysts account for 0.01% of central nervous system tumors. \[3,9\] These cysts tend to be located anterior to the brainstem or within the fourth ventricle and commonly develop to extend into the cisterns or intraventricular space. To date, only four reports, to the best of our knowledge, have described brainstem neurenteric cysts.

Among the previously reported cases with brainstem neurenteric cysts, the cyst was located in the pons \[6\] in one case and in the medulla oblongata in the remaining cases.

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**Figure 1:** Preoperative magnetic resonance (MR) images: (a) T1-weighted image (T1WI) showing cyst contents as hypointense, (b and c) axial and sagittal T2-weighted images (T2WI) showing a hyperintense cyst in the pons, (d) contrast-enhanced T1WI showing a mural nodule in the cyst, (e and f) subsequent magnetic resonance images (after 1 month) showing cyst enlargement in axial and sagittal T2WI.
Table 1: Summary of previously reported brainstem neurenteric cysts.

| Case | References       | Age/Gender | Clinical presentation | Lesion location     | Lesion size (mm) | MRI features | Intraoperative findings |
|------|------------------|------------|-----------------------|--------------------|------------------|--------------|------------------------|
| 1    | Zalatnai et al., 1987 | 0/NA       | Died 70 min after birth | Medulla oblongata  | 4 mm            | NA          | NA                     |
| 2    | Lach et al., 1989  | 66/F       | Ataxia, vertigo, diplopia, hemianicranial pain | Medulla oblongata | NA              | Low         | NA                     |
| 3    | Cho et al., 2010  | 23/M       | Nausea, dizziness     | Medulla oblongata  | NA              | Low         | NA                     |
| 4    | Li et al., 2017   | 56/M       | Right V2 and V3 dysesthesia staggering gait Babinski sign Romberg sign | Pons | 30×25×25 | Low | NA | + | Transparent fluid |
| 5    | Present case      | 7/F        | Headache, Diplopia Dysarthria | Pons | 24×27×20 | Low | NA | + | Milky white viscous fluid |

T1WI: T1-weighted image, T2WI: T2-weighted image, F: Female, M: Male, CSF: Cerebrospinal fluid, MRI: Magnetic resonance imaging

Table 1] [2,5,11] Brainstem neurenteric cysts occur in patients of all ages, and clinical presentations vary from headaches to cranial nerve disorders depending on the lesion location. MRI reveals variability in signal intensity depending on cyst contents. Neurenteric cysts typically appear as isointense to slightly hyperintense relative to cerebrospinal fluid on T1- and T2-weighted images. Intracranial neurenteric cyst with enhanced mural nodules is extremely rare and only six cases have been reported to date. [10] Interestingly, three cases, including our case, showed nodule on MRI or intraoperatively.

Neurenteric cysts are considered to be benign and slow growing, with few studies reporting very gradual cyst expansion that occurs over several years. [7,8] In our case, however, cyst expansion occurred in a short period of time resulting in rapid deterioration of the patient’s symptoms. Cyst enlargement was detected immediately because the lesion was located in the brainstem and caused symptomatic deterioration. Potential mechanisms underlying this rarely observed rapid cyst enlargement could include increased secretion, hemorrhage, and inflammation. Hemorrhage seems an unlikely cause for the expansion due to a lack of evidence of vascularity within the lesion. Although inflammation could be a possible cause, we did not observe any microscopic evidence such as histiocyte accumulation. Secretion by the columnar epithelial cells of neurenteric cysts has been shown to be responsible for the expansion of intraparenchymal cysts. Furthermore, secretions of the

Figure 2: Intraoperative imaging. (a) Intraoperative images through the microscope showing bilateral cerebellar hemispheres retraction exposing the intact fourth ventricle floor, (b and c) intraoperative images showing the presence of cyst containing milky white viscous fluid and mural nodule. (d) Histopathological examination of the cyst wall showed columnar epithelium containing ciliated cells.

Figure 3: Postoperative imaging after 5 years. (a-c) Magnetic resonance imaging showing small residual cyst in the brainstem on T1WI and T2WI.
cyst epithelium may also result in an osmotic response contributing to cyst enlargement. Recently, Yamamoto et al. suggested that melanin produced from ciliated columnar cells induce inflammation and may lead to cyst expansion.\[10\]

Complete surgical resection is the gold standard for the treatment of neurenteric cysts and is associated with the most favorable outcomes.\[3\] Chen et al. analyzed correlations between clinical characteristics and recurrence.\[1\] They reported three risk factors of recurrence: (i) age under 30 years old, (ii) subtotal resection in the first operation, and (iii) size of supratentorial neurenteric cysts over 30 ml.\[1\] However, total resection is not always achievable due to the adhesion of the cyst wall to surrounding critical neural structures such as brainstem in our case.\[9\] Considering the age of the patient and subtotal resection in our case, follow-up MRI is necessary to detect recurrence and close surveillance is also required for detecting symptoms of regrowth of the lesion.

**CONCLUSION**

We reported a case of a neurenteric cyst in the brainstem of a 7-year-old girl, which expanded in a short period, and review this rare entity. Neurenteric cysts in the brainstem should be considered to be distinct from other brainstem cystic lesions and need close observation.

**Acknowledgments**

The authors thank the cooperation of this patient involved. The present study was supported by Juntendo.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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