Case Report

Rare Recurrent Enterogenous Cyst Located in Different Areas of Cerebellar Hemisphere

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Summary

Enterogenous cysts are rare congenital anomalies. The great majority of them are located in the spinal canal, mediastinum and abdominal cavity, but it is extremely rare locating in cerebellar hemisphere. To our knowledge, there had been only a few cases located in this area reported in the literature, but it has not been reported about recurrent ones in cerebellar hemisphere, especially in different areas before and after recurrence. We herein presented a quite rare case of recurrent enterogenous cyst of cerebellar hemisphere. Considering the harm of cysts in cerebellar hemisphere, we should pay more attention to them, especially to recurrent ones.

Key words: Enterogenous cyst; cerebellar hemisphere; congenital; recurrent

INTRODUCTION

Enterogenous cysts are rare benign congenital lesions of presumed endodermal derivation resulting from a dysembryogenetic error that occurs in the third or fourth week of intrauterine life. They are generally located in the mediastinum, the abdominal cavity, skull, or within the spinal canal, especially in the cervical or thoracic segments of the spine. An intracranial lesion of enterogenous cyst located in cerebellar hemisphere is relatively quite rare. To date, there had been only a few recurrent cases reported in the literature to date. But it has not been reported about recurrent ones in cerebellar hemisphere. In the paper, we described a unique case of a recurrent cyst of cerebellar hemisphere. The lesion reappeared in different locations of cerebellar hemisphere 4 years after its first
CASE PRESENTATION

A 21-year-old male who presented with headache of nearly one month duration was first admitted to hospital in May 2007, accompanied with no other abnormal symptoms. The magnetic resonance imaging (MRI) of the head revealed a large cystic mass located in the right posterior fossa, with a separation in it (Fig. 1A, B). Thereafter, he underwent the excision of cyst under general anesthesia. At surgery, we found the contents of the cyst were turbid liquid with a gelatinous consistency. The final histological diagnosis was enterogenous cyst. The postoperative symptom of headache disappeared, but the patient had no sensory discomfort at that time. Thereafter, the brain MRI was made twice, separately in 2 years and 3.3 years after the operation, but there had not been much change compared to the postoperative images and the patient still had no exceptional symptoms.

The patient was admitted into hospital four years after the first operation, because of a headache, vomiting and dizziness of approximately two weeks duration. No abnormal findings were revealed throughout physical examination. The laboratory test result of cerebrospinal fluid (CSF) revealed no abnormal result except slightly increasing of protein. The MRI of the head revealed an intracranial space occupying lesion in the right cerebellar hemisphere. The T1-weighted axial MRI images showing a large hypointense signal cystic lesion in the right cerebellar hemisphere, while the T2-weighted axial MRI images presenting a hyperintense signal (Fig. 2A, B). The patient received resection of cyst of cerebellar hemisphere again. The final histological diagnosis was still enterogenous cyst (Fig. 3). During the three-month follow-up, the clinical symptoms of the patient disappeared. A long follow-up period is currently in progress.

Fig 1: Preoperative brain MRI without contrast, T1-weighted (A) and T2-weighted transversal projections (B) reveals a large cystic mass located in the right posterior fossa, with a separation in it. The obvious mass effect it creates can be observed
DISCUSSION

Enterogenous cysts are denied as congenital cysts lined by mucin secreting epithelium resembling that of the gastrointestinal tract according to the World Health Organization. The first enterogenous cyst was described by Puusep in 1934(1) and was located in the spinal cord, but the first descriptions of intracranial enteric cysts were described by Small(11) and Giombini et al.(5).

Enterogenous cysts are extremely rare, and occupy approximately 0.01% of all tumors of the central nervous system(8), but the exact incidence has not been known. They are found mostly in the spinal, especially in the lower cervical and upper thoracic segments. Enterogenous cysts are found more often at the level of the spinal canal than intracranially, and usually present most often axially.

Fig 2: The T1-weighted axial MRI images showed a large hypointense signal cystic lesion arising from the right cerebellar hemisphere (A), while the T2-weighted axial MRI images presenting a hyperintense signal (B)

Fig 3: The final histological diagnosis was enterogenous cyst (HE staining, ×100)
Enterogenous cysts were once named neuroenteric cysts, enteric cysts, endodermic cysts, gastrogenic cysts, etc. The most widely accepted hypothesis was that enterogenous cysts usually derived from incompletely separation between the endoderm and notochord during the third or fourth week of the embryonic development. When they develop in an intracranial location, these cysts are mainly located along the midline of the posterior cranial fossa, anterior to the brainstem or in the fourth ventricle. Differential clinical diagnosis should be made from the lipoma, neuroepithelial cysts and epidermal cysts. The clinical presentation was mostly because of the oppression of lump caused by enterogenous cysts, such as headache, nausea, vomiting, vertigo, etc.

In our case, although both the original cyst and the recurrent cyst were located in the cerebellar hemisphere, the locations where the enterogenous cysts developed before and after recurrence were different. Two last answers of postoperative histopathology were both diagnosed as enterogenous cyst.

In view of the benign nature of the enterogenous cysts, they are amenable to conservative treatment firstly without abnormal symptoms. If not, the treatment of choice for enterogenous cysts is total surgical resection of the cyst wall. In our operations, we could find the contents of the cyst were turbid, milky liquid with a gelatinous or mucous consistency, the same as the results of our first operation and the reports in the literature.

To date, it has not been reported about the recurrence of the enterogenous cysts in cerebellar hemisphere. It was reported that the estimated time for a foreseeable recurrence after partial resection from 2 months to 14 years. Therefore, a long-term clinical and neuroradiological monitoring of the operated patients is highly recommended.

Although the exact mechanism of enterogenous cyst located in different positions of cerebellar hemisphere is still unknown, we suggest that the migration of foregut cells must play a very important role. To the ectopic enterogenous cyst in cerebellar hemisphere, as in our case, more focus is needed, especially with respect to their development. Also, the further understanding of the embryology and genetics of enterogenous cyst in cerebellar hemisphere is desired.

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REFERENCES
3. Bejjani G, Wright DC, Schessel D, Sekhar LN. Endodermal cysts of the posterior fossa: report of