Frontal Neurenteric Cysts Combined with Anterior Skull Base Osteoma and Spontaneous Intracranial Pneumatosis: A Case Report and Literature Review

Yuekui Wu  
Tsinghua University

Xin Pan  
Tsinghua University

Pengxiang Yan (yanpengxiang@bjtth.org)  
Capital Medical University

Yuqi Zhang  
Tsinghua University

Yanhui Sun  
Capital Medical University

Jian Cui  
Capital Medical University

Case report

Keywords: Neurenteric cysts, skull base, osteoma, intracranial pneumatosis

Posted Date: November 8th, 2021

DOI: https://doi.org/10.21203/rs.3.rs-1017276/v1

License: This work is licensed under a Creative Commons Attribution 4.0 International License. Read Full License
Abstract

Background: Neurenteric cysts (NCs) are rarely that arise from incomplete dissolution of the neurenteric canal, most are found in the spinal canal, intracranial neurenteric cysts occur rarely, no cases of neurenteric cysts combined with anterior skull base osteoma have been reported.

Case presentation: A 23-year-old man presented with intermittent headache and vomiting for more than 20 days after running. The computed tomography (CT) and magnetic resonance imaging (MRI) showed a cystic mass and bone tumor in the left frontal lobe, accompanied by intracranial pneumatosis in the left ventricle and left frontal lobe, then he underwent a bicoronal scalp incision and frontal craniotomy to perform resection of the lesion. Postoperative cranial MRI showed no residual tumor. The patient recovered well after surgery, without ambulation or neurological complications.

Conclusions: Based on existing literature and this case, we suggest that surgical resection of intracranial neurenteric cysts should be the first choice, and the accompanying intracranial osteoma is removed at the same time. The treatment of choice remains complete surgical excision, and prognosis is good.

Background

Neurenteric cysts are rarely that arise from incomplete dissolution of the neurenteric canal during the third week of embryonic life. Most are found in the spinal canal, Intracranial neurenteric cysts occur rarely and there are only a small series of patients published in literature worldwide. We report a case of frontal neurenteric cysts combined with anterior skull base osteoma and spontaneous intracranial pneumatosis, which is the first case of this kind to be reported. This report presents a case of a 23-year-old young man with frontal confirmed pathologically proven neurenteric cysts combined with anterior skull base osteoma and spontaneous intracranial pneumatosis, who underwent a bicoronal scalp incision and frontal craniotomy to perform resection of left frontal lesion, after which there was improvement in symptoms.

The first report of of neurenteric cysts of CNS was published by Kuba and Fulton in 1928 and were first described in detail in the spinal region by Puusepp in 1934, the embryopathogenesis of neurenteric cysts is still unclear and various hypotheses have been proposed. The most common hypothesis is that neurenteric cysts arise from failure of dissolution of the neurenteric canal. Intracranial locations occur predominantly anterior to the brainstem, at the cerebellopontine angle and to a lesser extent within the fourth ventricle. Clinical symptoms are due to mass effect, and presentation is therefore variable and related to lesion size and location, the most common clinical manifestations are headache, vertigo, nausea, vomiting, and various cranial nerve deficits depending on lesion location.

Neurenteric cysts are benign, while rare, malignant transformation may occur, and the recurrence interval is gradually shortened, neurenteric cyst combined with anterior skull base osteoma has not been reported in the literature. To our knowledge, we report the first case of neurenteric cyst combined with an anterior skull base osteoma and spontaneous intracranial pneumatosis which was detected accidentally by
diagnostic imaging in a 23-year-old man. We also discuss the results of a brief literature review and the pathological findings, imaging spectrum, evaluation and management of neurenteric cysts.

Case Report

The 23-year-old man presented with intermittent headache and vomiting for more than 20 days after running. The CT and MRI showed a 8.0 × 6.0×6.7cm large cystic mass in the left frontal lobe, local brain parenchyma was compressed, accompanied by intracranial pneumatosis in the left ventricle and left frontal lobe. The signal intensity of the cystic lesion was similar to intensity of white matter on T1-weighted images (T1WI) and high-signal intensity on T2-weighted images (T2WI), enhanced MRI did not demonstrate enhancement. Head CT showed that the bone tumor communicated with the left anterior ethmoid sinus and protruded into the ethmoid sinus.

After the patient was admitted to hospital, the relevant preoperative examinations were completed, surgical management was the recommended course of the treatment, and then the left frontal lobe lesion was excised through the left approach of the coronal incision under general anesthesia on 2021-07-30. After the dura was opened during the operation, multiple cystic lesions in the left frontal lobe were exposed. The cyst wall was thin but intact, the cyst fluid was yellow and transparent, which was viscous. The cyst membrane was punctured and gelatinous fluid evacuated, the only attachment point of the cyst wall was located on the surface of the bony tumor at the frontal base. After complete resection, the lateral ventricle, interventricular foramen and skeletal plexus were clearly visible, the ventricular wall was well protected.

A cauliflower-shaped osteoma was seen in the left skull base, which was white in color and hard in nature. The osteoma was bitten off with a biting forceps, the skull base was reconstructed filled with its own muscle. Finally the lesion was completely resected. Postoperative cranial MRI showed no residual tumor. The patient recovered well after surgery, without ambulation and neurological complications.

Discussion

Neurenteric cysts can occur anywhere throughout the neuraxis and have various synonyms; they are typically located ventral to the cervical or thoracic spinal cord, these benign lesions rarely occur intracranially, embryopathogenesis of neurenteric cysts is still unclear and various hypotheses have been proposed. Clinically, neurenteric cysts may present with symptoms related to mass effect, or they can be asymptomatic and incidentally discovered[4, 5].

Radiological characteristics of intracranial neurenteric cysts are quite variable depending on protein content of the cystic fluid. The CT appearance is variable with neurenteric cysts previously reported as hypodense or hyperdense depending on protein content. As with CT, the MRI appearance is variable and depends on the protein content within the cyst. Typically, neurenteric cysts with low protein content are isointense to slightly hyperintense relative to cerebral spinal fluid on T1WI, while cysts with higher protein
content are more hyperintense on T1WI. T2WI and FLAIR signal intensities can be variable, but are more commonly hyperintense in signals. Typically, the cysts rarely show rim enhancement by contrast medium. These findings highlight the variability in imaging characteristics displayed by neurenteric cysts, which is likely a reflection of the range in protein level of the cyst contents.

Skull osteomas are benign bone-forming neoplasms that can occur in any part of the skull. Most osteomas are asymptomatic, and some are accidentally found in imaging examinations. Only 4%-10% of osteomas may cause clinical symptoms, especially when the lesion penetrates skull base, which may cause headache, frontal sinusitis or chronic sinusitis. If the osteoma produces compression effect or penetrates into the skull to form pneumocephalus and mucous cysts, surgical treatment is necessary. Ongoing advancements in endoscopic surgery have allowed less invasive surgical approaches to be adopted for removal than open surgery. If the time is long enough, the osteomas can recur, and the tumor grows faster after incomplete resection. There is no report of malignant transformation of skull base osteoma.

No cases of intracranial neurenteric cysts accompanied by skull base osteoma have been reported, and the relationship between these two lesions is unclear, which may be a coincidence, the cause for the formation of pneumocephalus is considered to be that the osteoma penetrates the dura, which makes the dura mater lose its integrity, and the air from the sinuses can then enters the skull.

The first-line treatment for neurenteric cysts and osteomas is complete surgical resection, as both of these diseases are not sensitive to radiotherapy or chemotherapy. If the mass is large or is causing symptoms, not only radical but also subtotal resection can lead to good control of disease opening on to good postoperative course and outcome. Total resection of the cyst wall can cure the disease. If the cyst wall can not be completely removed during the operation, the cyst wall should be removed as much as possible, which can reduce the recurrence rate that may occur several years after surgery. Therefore, a long-term follow-up lasting even more than ten years after surgery is recommended. During the operation, the contents of cyst can be sucked out in advance to achieve the purpose of volume reduction, avoiding the overflow of the contents caused by the rupture of the capsule cavity, which may help reduce the occurrence of aseptic meningitis.

Conclusions

Based on existing literature and this case, we suggest that surgical resection of intracranial neurenteric cysts should be the first choice, and the accompanying intracranial osteoma is removed at the same time. The treatment of choice remains complete surgical excision, and the prognosis is good.

Abbreviations

NCs: Neurenteric cysts; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; CNS: Central Nervous System; T1WI: T1-Weighted Images; T2WI: T2-Weighted Images; GFAP: Glial Fibrillary Acidic
Protein; IHC: Immunohistochemistry; FLAIR: Fluid Attenuated Inversion Recovery.

Declarations

Acknowledgements

None.

Authors’ contributions

Yuekui Wu, Xin Pan, Pengxiang Yan performed the operation and drafted the manuscript. Yuqi Zhang, Yanhui Sun, Jian Cui participated in its coordination, supervision, and revision of the manuscript. All authors read and approved the final manuscript.

Funding:

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Availability of data and materials

Not applicable.

Ethics approval and consent to participate

Not applicable.

Consent for publication

The patient received written informed consent before undergoing surgery and related clinical data collection.

Competing interests:

The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Author details

1Department of Neurosurgery, YuQuan Hospital· Tsinghua University, Beijing, China

2Department of Neurosurgery, Beijing Tiantan Hospital, Capital Medical University, Beijing, China

References


Figures
Figure 1

Figures A and B show preoperative head CT, with cystic space occupying in the left frontal lobe and osteoma in the skull base are clearly visible. Figure C shows preoperative head-enhanced MRI; Figures D and E are intraoperative images, showing multiple cystic solid lesions in the left frontal area with yellow cystic fluid, and cauliflower-shaped bony bulge in the anterior skull base. Figures F and G show postoperative enhanced MRI, suggesting complete tumor resection; figure H and I show postoperative HE
staining (original magnification 40× and 10×). Pathology report: Neurenteric cysts in left frontal lobe, complicated with angiolympoma. IHC showed CK (epithelial +), EMA (epithelial +), D2-40 (lymphatic +), CD31 (vascular +), GFAP(-), fibrous tissue and mature bone tissue in the left frontal floor.