Case Report

Pineal region neuroenteric cyst in a 13-year-old girl: A rare localization with postoperative recurrence and local dissemination

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INTRODUCTION

Endodermal (neurenteric) cysts are rare benign neoplasms of the central nervous system most often located in the spinal cord and rarely, intracranially. The nonspecific neuroimaging features make management planning potentially challenging. We present a case of a radiologically misdiagnosed neurenteric cyst with a complicated course.

BACKGROUND: Neuroenteric cysts are rare cystic benign neoplasms of the central nervous system most often located in the spinal cord and rarely, intracranially. The nonspecific neuroimaging features make management planning potentially challenging. We present a case of a radiologically misdiagnosed neuroenteric cyst with a complicated course.

CASE DESCRIPTION: A 13-year-old girl presented with a 6-month history of headache, tinnitus, and dizziness. Initial magnetic resonance images (MRIs) were indistinguishable from a pineal arachnoid cyst with aqueductal stenosis and hydrocephalus. Cyst fenestration was performed through an infratentorial supracerebellar approach. Histology revealed a neuroenteric cyst. On day 10 postoperatively, she deteriorated with acute hydrocephalus and cyst enlargement. An external ventricular drain was inserted. She remained asymptomatic thereafter. At 1-year postoperative, the patient remains asymptomatic despite the MRI showing cyst enlargement and local dissemination in the form of multiple cystic lesions in the cerebellum along the operative corridor. The patient was managed conservatively considering adhesion noted intraoperatively.

CONCLUSION: Neuroimaging features of brain cystic lesions may be nonspecific. Special attention should be awarded to posterior fossa and paramedian cystic lesions. Rarer lesions like neuroenteric cysts should also be considered. When in doubt, we recommend using the following methods to prevent the free outflow of the cyst contents into the subarachnoid space: lining the cyst and operative corridor with cotton wool and puncture opening and suctioning of fluid. However, the “gold standard” remains surgical treatment with radical excision of the endodermal cyst capsule. It is necessary to preassess the possibility of total or subtotal resection.

Keywords: Arachnoid cyst, Brain embryology, Cyst recurrence, Endodermal cyst, Neurenteric cyst, Pineal region cyst
fossa, that is, the ventral surface of the brain stem and cerebellopontine angle. Supratentorial location is less common. In other cases, they have been reported in the craniovertebral junction region and the cervicothoracic part of the spinal cord. The ratio of spinal and intracranial enterogenic cysts is 3:1.\textsuperscript{2, 3, 12, 14, 16, 22} Among the currently available methods of neuroimaging for differentiation of intracranial cysts, MRI is optimal.\textsuperscript{4} However, there is no consensus on the MRI characteristics of enterogenic cysts. Several publications indicate that most often the contents of enterogenic cysts have a slightly hyperintense or isointense signal to the cerebrospinal fluid (CSF) in the T1 mode; in T2 and FLAIR mode, the signal is usually hyperintense to CSF. The cyst walls do not typically enhance with contrast. However, in some cases, weak post contrast signal amplification may be observed.\textsuperscript{1, 5, 14, 18}

This paper presents an initially misdiagnosed and surgically treated intracranial endodermal cyst, leading to recurrence and local dissemination in a child.

**CASE DESCRIPTION**

A 13-year-old girl presented with a 6-month history of headaches, tinnitus, and dizziness. The symptoms were partly relieved by nonsteroidal anti-inflammatory drugs. The attending neurologist requested a magnetic resonance imaging (MRI) of the brain which showed noncommunicating hydrocephalus secondary to a large pineal region cyst compressing the cerebellum and the cerebral aqueduct. Ophthalmology examination revealed features of papilledema. The child was fully conscious without focal or meningeal signs and symptoms. A brain MRI with contrast enhancement showed an irregular noncontract enhancing cystic lesion of the pineal region [Figure 1].

The cyst pushed the quadrigeminal plate and the aqueduct anteriorly and the cerebellum downward. Its lower-ventral surface adjoined the fourth ventricle, superiorly the walls of the cyst bordered on the bodies of moderately dilated lateral ventricles. Periventricular edema was noted. A diagnosis of arachnoid cyst of the pineal region with sub-compensated occlusive hydrocephalus was made. After multidisciplinary consultation, the patient was scheduled for microsurgical fenestration of the cyst walls and allows communication with the subarachnoid spaces of the posterior fossa to eliminate the mass effect and restore adequate CSF circulation.

The cyst was approached through a midline infratentorial-supracerebellar corridor. On dural opening, a bulging cerebellum compressed by the cyst was noted with many arachnoid adhesions with the surrounding structures. These adhesions were carefully released and the whitish-yellow cyst was visualized [Figure 2].

The cystic fluid was partly cloudy and colorless. A biopsy of the capsule was taken. Postoperative computed tomography (CT) showed decreased cyst size with hyperdense content (interpreted as normal postoperative changes), mild pneumocephalus, and a patent cerebral aqueduct [Figure 3].

Pathohistological results indicated that the lesion was an endodermal cyst. The tissues showed dense and loose fibrous connective tissue lined by a single-layered and pseudostratified ciliated epithelium. Epithelial cells expressed cytokeratin (CK) 7, pan-keratin, and epithelial membrane antigen and were negative for glial fibrillary acidic protein, S100, and CK 20 [Figure 4].

Day 6 postoperative MRI showed a decrease in the cyst size to 10–20% of the preoperative size with a symmetrical reduction in lateral ventricle size [Figure 5].

On day 10 postoperatively, the patient deteriorated with severe headache, vomiting, and developed a CSF leak. Control brain CT showed features of noncommunicating hydrocephalus and a $\times$ 2.5 increase in the cyst size compressing the cerebral aqueduct [Figure 6]. An urgent endoscopic third ventriculostomy was performed. Postoperative MRI showed regression of the hydrocephalus and increase of the endodermal cyst to about 80% of the original size [Figure 7]. The MRI features of the cyst contents were also of interest. On initial presentation, it was similar

![Figure 1: MRI of the brain on admission showing the cyst isointense to cerebrospinal fluid (blue arrow) in T1, T2, and flair, dilated ventricles (orange arrow), and compressed cerebral aqueduct (white arrow).](image-url)
to CSF on T2, T1, and FLAIR modes. However, the last two images showed a clear hyperintense signal.

On day 4 after the second operation, she was discharged with no neurological deficit. At 1 and 3 months after discharge, the patient was still asymptomatic. Control brain MRIs were essential without change. However, 1-year routine control MRI showed significant changes compared to previous images [Figure 8].

On the superior surface of the cerebellum along the operative corridor of the supracerebellar approach, multiple noncommunicating cysts of various sizes were visualized. The size of the original pineal region cyst had slightly increased. Given the unusual progression of the disease, an enhanced MRI of the entire central nervous system and a brain positron emission tomography-CT with methionine was performed and normal. The patient was managed conservatively with a follow-up MRI done after 1 year and 20 months [Figures 7 and 9]. Of note, there was an increase in the number of new smaller cysts in the parenchyma of the cerebellum, as well as a slow progressive increase in their size with no perceptible mass effect. The pineal region cyst also showed a gradual increase, especially caudally. There are still no secondary changes in the spinal cord.

From the time of discharge, the patient has been asymptomatic and active with no focal neurological deficits. However, the authors are watchfully waiting and following up the patient clinically and radiologically. Should the cysts present with significant mass effect to cause clinical symptoms in the near future, surgical management with a goal of radical resection will be needed.

DISCUSSION

Endodermal cysts (neurenteric cysts and enterogenic cysts) are a rare malformation resulting from disorders of embryogenesis, leading to the formation of cystic cavities in the central nervous system filled with high-protein secretion.\[3,12]\ It is believed that neurenteric cysts form at about 3 weeks of gestation. At this stage, the yolk and amniotic sacs are connected by a temporary neurenteric canal, which subsequently closes as the notochord develops.\[2,12]\ Several mechanisms, leading to the development of neurenteric cysts, have been described: the separation of the notochord by endodermal tissue or diverticulum; the fusion of the notochord and endodermal layer; persistence of the neurenteric canal due to its incomplete obliteration; incomplete separation of the notochord and endoderm; and
improper formation of the notochord as a consequence of the incomplete separation of the ectoderm and endoderm.\(^2\)

At the moment, there are no accurate data describing the time when cyst enlargement begins, the rate of growth, and when the patients present with clinical features. In the pediatric population, the average age of diagnosis is around 6.4 years.\(^4\) The endodermal cysts manifest mainly by compression of surrounding tissues as they gradually grow. The features depend on the cyst location and structures being compressed, that is, nonspecific symptoms with increased intracranial pressure due to the large cyst size or the development of secondary occlusive hydrocephalus, focal cranial nerve deficit due to direct compression at the brain stem, and convulsive syndrome when located in the cerebral cortex.\(^15,16\)

The treatment method of choice of neurenteric cysts is neurosurgical excision, which has shown favorable outcomes and prognosis. Total excision of the cyst and its capsule is the standard of surgical treatment and reduces the risk of recurrence of the enterogenic cyst.\(^8\) However, depending on the localization, it is not always possible to achieve a total resection cyst capsule due to extensive adhesions with important anatomical structures.\(^15\) Incomplete excision of the cyst increases the risk of recurrence, which may warrant additional surgical interventions in the future. Takahashi et al. in his work proposed the option of cyst puncture using MR navigation,\(^20\) while other authors proposed the installation of a cysto-subarachnoid shunt.\(^21\) However, these methods are not preferable due to the risk of aseptic meningitis, which is a frequent complication seen when the contents of an enterogenic cyst pour into the subarachnoid space.\(^11\) Another frequent complication of surgical treatment of neurenteric cysts includes recurrence, commonly resulting from incomplete excision.\(^4\) According to the described cases of surgical treatment of endodermal cysts, the probability of recurrence ranges from 11.9% to 37%.\(^3\) Very few isolated cases of local and craniospinal dissemination of enterogenic cysts after surgical treatment are described.\(^9,22\)

In some cases, malignant transformation occurs, and in all sources in the literature, the histological diagnosis was adenocarcinoma.\(^7,10,13,17,19\) In the clinical case presented, the endodermal cyst was not initially suspected due to the lack of convincing neuroimaging features on MRI. Parasitic invasion was also not considered due to uncharacteristic neuroimaging features. The cyst contents were isointense with CSF on T1, T2, and FLAIR modes. There were no solid component and no contrast enhancement; therefore, the diagnosis of “arachnoid cyst” was initially made. By the time of cyst recurrence and postoperative deterioration, the histological diagnosis was already known. Repeat surgical intervention was not considered because intraoperatively, there were extensive adhesions between the cyst wall and the surrounding deep veins and cerebellar tissue. Despite the isolated cases of neurenteric cyst dissemination described in the literature, an extremely unexpected presentation was the formation of secondary cysts along the surgical corridor in the late postoperative period which was not present in the early control MR images.

Given the satisfactory condition of the child at the time of discharge, there was no clinical indication for additional surgical intervention. It is impossible to predict the future appearance of additional “daughter” cysts, including in the spinal subarachnoid space, despite the absence of convincing
signs of communication between the endodermal cyst and the foramen Magno cistern in postoperative images. It seems quite logical to have possible dissemination of the enterogenic cysts in the subarachnoid space when their contents are poured out during surgery. In this regard, an individualized surgical approach to avoid opening the contents of the cysts into the subarachnoid may be ideal. It is worth assessing the possibility of radical cyst excision in open surgical intervention and considering other management options for symptomatic relief if radical excision is impossible. Nonsurgical methods seem appropriate, given reports of spontaneous regression of the neurenteric cyst and neurological symptoms without surgical intervention. In hindsight, in the presented case, we believe the best management plan was to manage intracranial hypertension and observe the cyst size with a regular MRI.

Figure 7: MRI 1 year after surgery showed cystic lesions in the operative corridor (blue arrow) and an enlarging pineal region cyst with its contents slightly more hyperintense to CSF on T1 (white arrow).

Figure 8: MRI of the brain, 11th day after surgery.

Figure 9: MRI at 20-month postoperative showing increased number of cysts (blue arrow) and progressive increase in pineal cyst size (white arrow).
The possibility of total cyst excision should be discussed during preoperative planning.

CONCLUSION

Neuroimaging features of brain cystic lesions may be nonspecific. Special attention should be awarded to posterior fossa and paramedian cystic lesions. Rarer lesions like neurenteric cysts should also be considered. When in doubt, we recommend using the following methods to prevent the free outflow of the cyst contents into the subarachnoid space: lining the cyst and operative corridor with cotton wool and puncture opening and suctioning of fluid. However, the "gold standard" remains surgical treatment with radical excision of the endodermal cyst capsule. It is necessary to reassess the possibility of total or subtotal resection.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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