Clinical Case Report

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Intracranial endodermal cyst presenting with nonobstructive hydrocephalus

A case report

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Abstract

Rationale: Endodermal cysts are rare benign lesions in the central nervous system. Their common symptoms include headache and neck pain caused by mass effect or inflammatory reaction. We report the case of an elderly woman with intracranial endodermal cyst who presented with nonobstructive hydrocephalus.

Patient concerns: A 78-year-old woman presented with acute deterioration of consciousness caused by acute hydrocephalus. She subsequently underwent ventriculoperitoneal shunt placement. Eighteen months after this operation, she developed numbness of extremities and gait and progressive cognitive disturbances.

Diagnosis: Initially, the endodermal cyst could not be recognized, but it became clinically evident in the craniocervical junction after ventriculoperitoneal shunt placement. MRI revealed multiple cystic lesions in the pontomedullary cistern. Postoperative pathology confirmed the diagnosis of endodermal cyst.

Interventions: Subtotal resection of the cystic lesion was performed. Intra-operatively, multiple cysts containing a milky white fluid were noted and the medullary surface including the median and lateral apertures of the fourth ventricle were covered by thick, cloudy arachnoid membrane.

Outcomes: Although the numbness of extremities improved after the surgery, she remained bedridden due to deterioration in cognitive function and generalized muscle weakness. The patient developed recurrence 2 months after the surgery; however, no additional surgery was performed owing to her poor general condition.

Lessons: Endodermal cysts rarely present with nonobstructive hydrocephalus caused by recurrent meningitis. In such cases, the lesions are often invisible on initial diagnostic imaging, and complete resection of the lesions is typically difficult because of strong adhesion between the cyst walls and contiguous vital structures.

Abbreviations: CSF = cerebrospinal fluid, MRI = magnetic resonance imaging, VP = ventriculoperitoneal.

Keywords: endodermal cyst, hydrocephalus, intracranial, posterior fossa

1. Introduction

Endodermal cysts, rare benign lesions in the central nervous system, typically develop in third and fourth decades of life.[1] Their histopathology is characterized by cyst wall lined by ciliated, mucous-filled, or simple columnar epithelium. The etiopathogenesis of endodermal cyst remains unknown. Endodermal cysts of the central nervous system primarily occur in the spinal subdural space but rarely in the intracranial space.[2] Intracranial endodermal cysts are commonly located anterior to the brainstem, and their common symptoms are headache, neck pain, and weakness of the extremities.[3,4] Their symptoms are typically caused by mass effect or inflammatory reaction to the cyst content, but they are rarely caused by nonobstructive hydrocephalus. Herein, we report the case of an elderly patient with endodermal cyst at the craniocervical junction who presented with nonobstructive acute hydrocephalus.

2. Case presentation

A 78-year-old woman presented with a 3-month history of headache and nausea following acute deterioration of consciousness. Her medical history was unremarkable. Computed tomography of her head showed ventricular enlargement with a particularly remarkable enlargement of the fourth ventricle. Temporary external ventricular drainage was performed due to severe disturbance of consciousness caused by acute hydrocephalus. Examination of cerebrospinal fluid (CSF) revealed normal levels of glucose and protein and absence of pleocytosis. No obstructive lesion was detected on gadolinium-enhanced magnetic resonance imaging (MRI); however, a small enhancing lesion was detected at the ventral side of the cervical spine (Fig. 1A). Accordingly, she underwent ventriculoperitoneal (VP)

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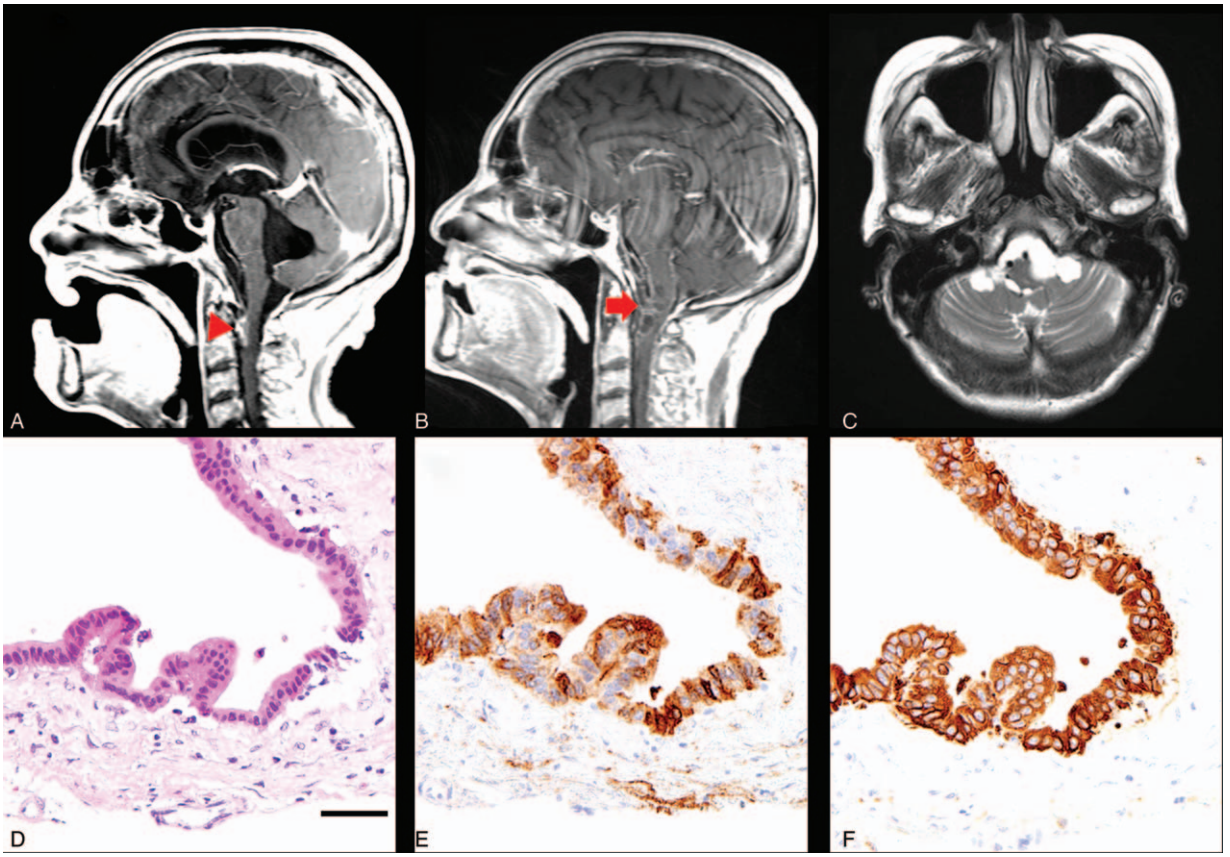

Figure 1. Magnetic resonance imaging (MRI; A–C) and micrographic findings (E–G). Gadolinium-enhanced MRI at the first admission showing hydrocephalus and small enhancing lesions in the ventral aspect of the cervical cord (A, arrowhead). Gadolinium-enhanced MRI 18 months after ventriculoperitoneal shunt placement shows multiple cystic lesions anterior to the brainstem (B, arrow), which homogeneously appear hyperintense on T2-weighted MRI (C). Micrograph showing the cyst wall lined by a single layer of columnar epithelial cells (HE, D). Epithelial cells showing high expression levels of EMA (E) and cytokeratin AE1/AE3 (F). The scale bar in (D) indicates 75 μm for D–F. EMA = epithelial membrane antigen, HE = hematoxylin and eosin, MRI = magnetic resonance imaging.

shunt placement. Her symptoms improved with reduction in the ventricular size. During the subsequent year, shunt revision surgery was performed for shunt tube obstruction on two occasions. Examination of CSF tapped from the shunt valve showed normal levels of protein and glucose. Eighteen months after the first operation, she developed numbness of extremities and gait and progressive cognitive disturbances. MRI revealed multiple cystic lesions in the pontomedullary cistern, and the cyst walls exhibited contrast enhancement with gadolinium (Fig. 1B). The cyst content was hyperintense on T2-weighted and hypointense on T1-weighted MRIs (Fig. 1C). Interestingly, CSF collected by lumbar puncture revealed remarkably high protein and normal glucose levels, whereas that collected from the shunt valve revealed normal protein and glucose levels. She underwent surgery via a far lateral transcondylar approach. Subtotal resection of the cyst capsule and cyst evacuation were performed to preserve the contiguous vital structures which were strongly adhered to the cyst walls. On intraoperative examination, multiple cysts containing a milky white fluid were noted; the medullary surface including the median and lateral apertures of the fourth ventricle were covered by thick, cloudy arachnoid membrane. Microscopic examination revealed a thin cyst wall lined by focally ciliated columnar epithelial cells (Fig. 1D). These cells showed intense immunopositivity for epithelial membrane antigen and cytokeratin AE1/AE3 (Fig. 1E and F). The pathological diagnosis was endodermal cyst. Although the numbness of extremities improved after the surgery, she has remained bedridden due to deterioration in cognitive function and generalized muscle weakness. Two months after the surgery, enlargement in the size of the cystic lesion was found on MRI; however, no additional surgery was performed owing to her poor general condition. The patient has provided informed consent for publication of the case.

3. Discussion

In a study by Bejjani et al, 20% of patients with intracranial endodermal cyst presented with aseptic meningitis caused by intermittent leakage of cyst content.[1] Previously reported cases of intracranial endodermal cyst were diagnosed as meningitis based on the clinical symptoms and findings of CSF examination and MRI. In our case, the CSF findings just before the surgery for endodermal cysts and the intraoperative findings were indicative of recurrent noninfectious meningitis, which was assumed to be the cause of nonobstructive hydrocephalus. The dissociation of the CSF findings between the lateral ventricle and spinal subarachnoid space and the focal thickening of arachnoid membrane suggested that the meningitis was restricted to the area around the endodermal cysts. The hydrocephalus characterized by significantly greater enlargement of the fourth ventricle may have been caused by inflammatory adhesion of the dorsal part of the cerebellomedullary arachnoid membrane, which prevented

the outflow of CSF not only from the fourth ventricle to the cerebellomedullary cistern but also from the cerebellomedullary cistern to the subarachnoid space on the inferior surface of the cerebellar hemisphere. Pressure gradient between the fourth ventricular surface and cerebellar surface in the posterior fossa may result in significant enlargement of the fourth ventricle. The decrease in infratentorial pressure due to drainage of CSF is believed to have led to the development of endodermal cysts. A similar case of enlargement of endodermal cysts following CSF drainage was previously reported.[1] Most cases of endodermal cysts with aseptic meningitis present with inflammation-associated headache. On reviewing relevant literature, we found a few cases which presented with obstructive hydrocephalus associated with mass effect of endodermal cyst; however, none of the reported cases showed nonobstructive hydrocephalus.[5–7] To the best of our knowledge, this is the first case of intracranial endodermal cyst which presented with nonobstructive hydrocephalus caused by recurrent noninfectious meningitis. Unnoticeable endodermal cysts should be considered as a cause of unexplained hydrocephalus.

Complete resection is the best treatment strategy for intracranial endodermal cysts, which prevents recurrence.[8–10] Recurrence rate after subtotal resection of endodermal cysts is higher than that after total resection. However, adhesion of the cyst walls to the contiguous vital structures often makes it difficult to achieve total resection. In particular, it is challenging in patients with extensive adhesions caused by recurrent meningitis, as observed in our patient. In a review of endodermal cysts, adhesion of cyst to the contiguous structures was observed in one-third of cases, and only subtotal resection was achieved in approximately 50% of cases.[1,11] In the present case, enlargement of the cystic lesion was observed only 2 months after the surgery, suggesting that incomplete excision and decrease in intracranial pressure via CSF drainage increase the risk of recurrence of endodermal cysts. In such cases, control of the intraventricular pressure is critical to prevent enlargement of endodermal cysts.

4. Conclusion

We report a clinically rare case of endodermal cyst. This case presented with nonobstructive hydrocephalus, and the cysts appeared after CSF drainage. The clinical examinations and intraoperative findings indicated the obstruction of CSF outflow caused by recurrent noninfectious meningitis. Complete resection of the lesions is typically difficult in such cases because of strong adhesion between the cyst walls and contiguous vital structures.

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