Gradually Progressive Symptoms of Normal Pressure Hydrocephalus Caused by an Arachnoid Cyst in the Fourth Ventricle: a Case Report

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Key words
- Arachnoid cyst
- Direct fenestration
- Fourth ventricle
- Normal pressure hydrocephalus

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
CT: Computed tomography
MRI: Magnetic resonance imaging

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BACKGROUND: **Arachnoid cysts in the fourth ventricle are extremely rare, with only 13 cases having been described in the literature. Especially, only 1 case of a patient older than 70 years has been reported. Arachnoid cysts in the fourth ventricle may cause obstructive hydrocephalus. Here, we report the case of a 72-year-old man who presented with an arachnoid cyst in the fourth ventricle that caused gradually progressive symptoms of normal pressure hydrocephalus.**

METHODS: **A 72-year-old man complaining of persistent dizziness and gait difficulty was admitted to our hospital due to a gradual worsening of his symptoms and apparent cognitive impairment. Computed tomography scan of the head showed symmetrically dilated third, fourth, and lateral ventricles.**

RESULT: **Though we performed a ventriculoperitoneal shunt operation, his trunk ataxia persisted. We finally diagnosed an arachnoid cyst in the fourth ventricle by direct ventricular infusion of enhanced material. We performed direct surgical fenestration of the cyst and achieved a good outcome.**

CONCLUSION: **Arachnoid cysts of the fourth ventricle are exceedingly rare, but it is important to recognize them because they cause normal pressure hydrocephalus symptoms and cerebellar or brainstem deficit. We propose detailed neurologic and radiologic examinations of patients with normal pressure hydrocephalus symptoms to avoid unnecessary shunt.**

Arachnoid cysts develop at various sites in the central nervous system, such as in the Sylvian fissure, or middle cranial or posterior cranial fossa. These types of cysts account for 1.1–1.7% of cysts found in healthy individuals,[1–3] and although the majority of these lesions are asymptomatic, they can occasionally cause various symptoms such as intracranial hypertension or convulsive seizure.

Arachnoid cysts within the posterior cranial fossa occur among infants and youths, but arachnoid cysts in the fourth ventricle are exceedingly rare with only 13 cases having been reported in the current literature.[3–12]

Here, we describe the case of an elderly individual who presented with an arachnoid cyst located in the fourth ventricle that gradually caused symptoms of progressive normal pressure hydrocephalus.

CASE REPORT

History and Examination

A 72-year-old man complaining of persistent dizziness and gait difficulty was admitted to our hospital due to a gradual worsening of his symptoms and apparent cognitive impairment from one year ago. The neurological examination on admission did not reveal any motor or sensory deficits, and we found his deep tendon reflexes to be normal. Furthermore, we found that cranial nerve functions were normal. The patient had difficulty in walking independently, showed severe dementia with 6/30 on the Hasegawa Dementia Scale-Revised test, and exhibited bladder and rectal dysfunction. He had no history of head trauma or any infections.

Noncontrast computed tomographic (CT) scan of the head showed symmetrically dilated third, fourth, and lateral ventricles (Figure 1). The analysis of the cerebrospinal fluid was unremarkable, and intracranial pressure was determined to be 70 mm H_2O with a lumbar puncture. CT cisternography revealed normal cerebrospinal fluid (CSF) flow pattern without any reflux into the ventricular system. Magnetic resonance image (MRI) performed with contrast material demonstrated dilation of the ventricular system with anterior compression of the brainstem and cerebellar tonsil descending into the foramen magnum (Figure 2). There was no evidence of any neoplastic lesion.

Operation

Although we suspected the presence of a cystic lesion in the fourth ventricle based on the MRI findings and normal CSF flow pattern, we performed a ventriculoperitoneal shunt because of the patient's triad of symptoms that included gait disturbance, dementia, and bladder dysfunction. During the shunting procedure, we also infused 5 mL of contrast material into the lateral ventricle in order to check the contents of the fourth ventricle.

Two weeks after the operation, although the patient's gait improved and his cognitive capacity recovered, as was revealed by an improved score (21/30) on the Hasegawa Dementia Scale-Revised

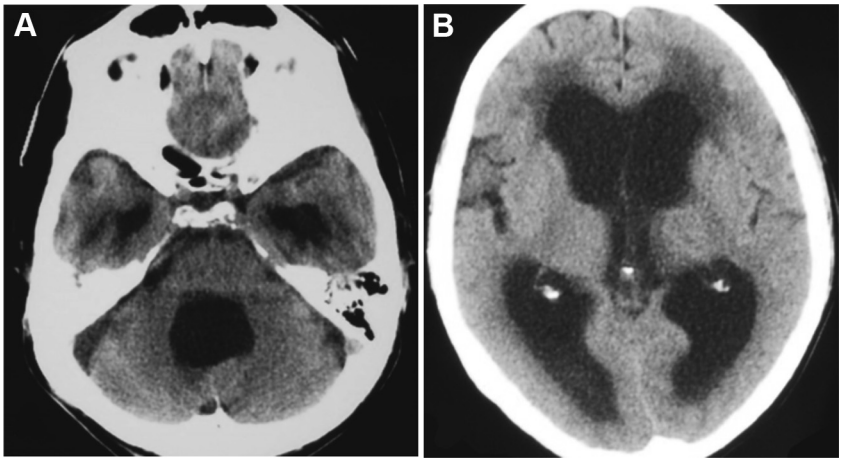

Figure 1. A computed tomography scan on admission documenting symmetrically dilated fourth (**A**), lateral and third (**B**) ventricles.

test, the patient's trunk ataxia persisted. Furthermore, a postoperative CT scan revealed that the size of the lateral and third ventricle had decreased, but that the size of the fourth ventricle remained unchanged. Moreover, it was determined that the contrast material had not flown into the fourth ventricle (Figure 3).

Pathologic Findings and Follow-up

We diagnosed the patient as having a cystic lesion in his fourth ventricle, which was responsible for causing noncommunicating hydrocephalus. Since the patient's trunk ataxia persisted postperatively, we performed cyst fenestration via a median suboccipital approach. After opening the dura, the tonsil and biventer lobes were laterally compressed. We were then able to visualize the cystic mass, which comprised a smooth thin wall in the fourth ventricle. After the cyst wall was resected, the CSF filling the mass was aspirated. The restored communication between the fourth ventricle and aqueduct was also confirmed (Figure 4). Histologic examination of the cyst wall revealed a single epithelial layer of arachnoid cells and no signs of inflammation or trauma; therefore we diagnosed this lesion as an arachnoid cyst. A postoperative CT scan revealed diminution in the size of the fourth ventricle. Furthermore, the patient's truncal ataxia improved remarkably, and he was discharged from the hospital without any neurologic deficits. A postoperative MRI 1 year after fenestration showed normal ventricular size (Figure 5).

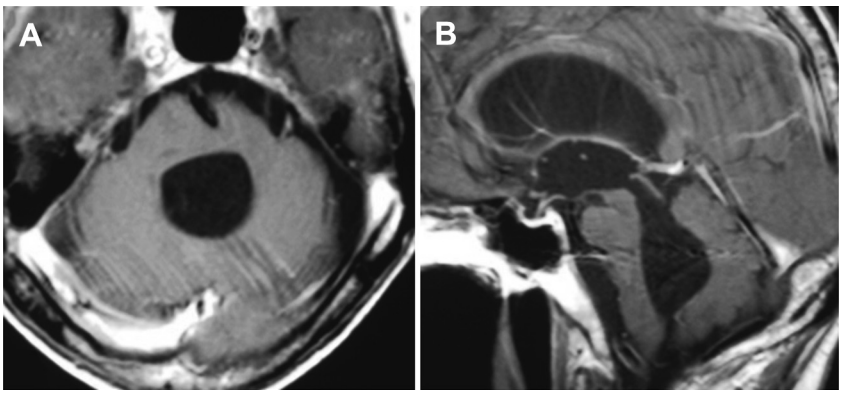

Figure 2. A magnetic resonance image with contrast material demonstrating dilation of the ventricular system with compression of the brainstem and cerebellar tonsil descending into the foramen magnum. Neoplasm was not detected. (**A**) Axial image. (**B**) Sagittal image.

DISCUSSION

Although arachnoid cysts can develop in several brain regions, it has been suggested that approximately 66.2% of these are located in the temporal fossa, 14% in the frontal convexity, 12% in the posterior fossa, and 7.7% in other locations of the neurocranium.[13] Many cases of arachnoid cysts within the posterior fossa develop in the cerebellopontine angle[14]; however, arachnoid cysts of the fourth ventricle are exceedingly rare, and only 13 cases have been reported in the literature.

In 1979, Di Rocco reported the first case of a 7-year-old boy who suffered from hydrocephalus caused by an arachnoid cyst located in his fourth ventricle.[6] In the previously reported cases, ventriculoperitoneal shunts were performed, similar to our case, to relieve symptoms of headache, dementia, and gait disturbance caused by hydrocephalus.[5–7,9] In the case reported here, we diagnosed the fourth ventricle arachnoid cyst only after an initial shunt operation with injection of contrast material into the lateral ventricle. However, if we had evaluated the MRI or CT cisternography more carefully, we would have been able to make the correct initial diagnosis. In this case, differential diagnoses would have included dermoid cysts, epidermoid cysts, ependymal cysts, cystic astrocytoma, or dilatation of the fourth ventricle due to obstruction of the foramen of Majandie or Lushka. Typically, arachnoid cysts can be distinguished from neoplasmatic cysts on the basis of MRI or CT scans; however, it is often difficult to differentiate an arachnoid cyst from non-neoplastic cysts such as ependymal or choroid plexus cysts.[3]

Bonde et al. described that ependymal cyst cannot be differentiated from arachnoid cyst on radiologic analysis, and they can only be identified by histopathologic studies after excision. They also described