Case Report

Arachnoid cyst of the velum interpositum originating from tela choroidea

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Abstract

Background: Arachnoid cysts originating from the velum interpositum are very rare, and their existence as a clinicopathologic entity remains controversial. We report a case of a patient with an arachnoid cyst of the velum interpositum presenting with memory disturbance, focusing on the anatomical origin of the lesion and the physiological mechanisms causing memory disturbance.

Case Description: A 65-year-old man with a large cystic lesion in the velum interpositum experienced progressive memory disturbance and enlargement of the lesion 6 months before referral to our institution. Neuropsychological evaluation on admission demonstrated severe memory disturbance. Radiological examination did not reveal hydrocephalus, but the bilateral fornices and thalami were compressed by the cyst. The patient underwent endoscopic cystoventriculostomy via the frontal horn of the right lateral ventricle through a frontal burr hole. Histopathology of the sample was consistent with that of an arachnoid cyst, and the endoscopic findings suggested that the cyst originated from the tela choroidea, which covers the velum interpositum. The symptoms resolved after surgery with significant improvement in neuropsychological test scores.

Conclusion: Arachnoid cysts of the velum interpositum are rare but distinct clinicopathologic entities that originate from the tela choroidea. The lesions can cause memory disturbance without hydrocephalus due to compression of the fornices and thalami, but this can be reversed by surgery.

Key Words: Arachnoid cyst, neuroendoscopy, velum interpositum



INTRODUCTION

Velum interpositum is the space between the two layers of the tela choroidea in the roof of the third ventricle.^[15] Pathological abnormalities that can originate from the velum interpositum include arachnoid cysts and meningiomas.^[3-7,16] However, arachnoid cysts of the velum interpositum are a controversial and not well-defined clinicopathologic entity. Reports of arachnoid cysts in this location are very rare and there have only been four reports to date.^[3,4,6,16] The pathogenic mechanism of such lesions remains unclear

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because the velum interpositum does not have the apparent arachnoid membrane.

We report a case of a patient with a cyst of the velum interpositum successfully treated by neuroendoscopy. We discuss the anatomical origin of the lesion, which was identified from both endoscopic findings and histopathology, as well as physiological mechanisms causing memory disturbance.

CASE REPORT

A 65-year-old male visited a medical clinic for memory disturbance. Magnetic resonance imaging (MRI) of the head revealed a cystic lesion around the velum interpositum. The symptoms worsened with enlargement of the lesion, and the patient was referred to our hospital for further examination and treatment. A neurological examination on admission revealed mild cognitive impairment with a Mini-Mental State Examination score of 22. There was no papilledema or focal neurological deficit. Further neuropsychological evaluation of memory using the Wechsler Memory Scale-Revised test^[19] revealed significant memory disturbance including severe impairment of recall (general memory index, 75; delayed recall index, <50). MRI on admission revealed a large cystic lesion between the third and lateral ventricles, inside of which had the intensity equal to that of the cerebrospinal fluid [Figure 1a]. The cyst compressed the fornix superiorly and the bilateral thalami laterally, whereas it did not involve the quadrigeminal cistern and displaced the internal cerebral veins inferiorly [Figure 2]. A computed tomography (CT) cisternography revealed delayed accumulation of a tiny amount of contrast medium in the cyst. As radiological examination did not reveal hydrocephalus, the cerebral glucose uptake was studied using fluoro-2-deoxy-d-glucose positron emission tomography (FDG-PET) for further examination on memory disturbance. FDG-PET revealed decreases in glucose metabolism in the bilateral thalami and medial aspects of the frontal and parietal lobes corresponding to the bilateral cingulate gyri [Figure 3a].

The patient underwent an endoscopic cystoventriculostomy via the lateral ventricle [Figure 4]. A 4-mm rigid endoscope was inserted into the frontal horn of the right lateral ventricle through a frontal burr hole. The foramen of Monro, anterior septal vein, and thalamostriate vein were preserved. A large protuberance was observed on the septum pellucidum, and the body of the lateral ventricle was occupied by the protuberance. A small hole was created on the protuberance using monopolar radiofrequency coagulation. The hole was enlarged using a 4-French Fogarty catheter with an inflatable balloon. A small section of the tissue was resected using microscissors and microforceps for histologic examination. The true cyst wall, which resembled a normal arachnoid membrane, was clearly recognizable under the thin ependymal membrane. The cyst was explored with a flexible endoscope inserted through the hole; the internal cerebral veins were observed through the semi-transparent inferior cyst wall. The veins were shifted inferiorly due to compression by the cyst. No apparent communication was seen between the cyst and quadrigeminal cistern. After cystoventriculostomy, the cyst collapsed and the body of the lateral ventricle became clearly visible. Histologic examination revealed that the cyst wall consisted of a thick collagen layer including arachnoid cells, which is a typical finding for an arachnoid cyst wall.^[14]

The symptoms resolved immediately after surgery. MRI after surgery confirmed a significant decrease in the size of the cyst [Figure 1b]. FDG-PET performed 20 days after surgery revealed improvement in glucose metabolism in the cingulate gyri and thalami [Figure 3b]. A year after surgery, the patient had no further symptoms and was able to perform activities of daily living independently, and the Wechsler Memory Scale-Revised test demonstrated significant memory improvement (general memory index, 104; delayed recall index, 100).



Figure 1:T2-weighted axial magnetic resonance imaging before (a) and after (b) surgery



Figure 2: T2-weighted sagittal magnetic resonance imaging before surgery. The internal cerebral veins are located at the inferior margin of the cyst (arrow), and the fornix is compressed superiorly by the cyst (arrowhead)



Figure 3: (a) Fluoro-2-deoxy-d-glucose positron emission tomography (FDG-PET) before surgery revealing reduction in glucose metabolism of the medial aspects of the cerebrum (arrows) and bilateral thalami (arrowheads). (b) FDG-PET after surgery demonstrating improvement in glucose metabolism at the regions indicated on the preoperative image

DISCUSSION

The velum interpositum is the space situated on the medial side of the body portion of the choroidal fissure in the roof of the third ventricle below the body of the fornix and between the superomedial surfaces of the thalami.^[15] Anatomically, the velum interpositum is defined as the potential space between the two layers of the tela choroidea; the upper layer of the tela choroidea is attached to the lower surface of the fornix and hippocampal commissure, and its lower layer is attached to the paired parallel strands of the choroid plexus in the roof of the third ventricle [Figure 5a]. The velum interpositum contains the internal cerebral veins and medial posterior choroidal arteries. It is usually a closed space that tapers to a narrow apex just behind the foramen of Monro, but it may infrequently communicate with the quadrigeminal cistern to form the velum interpositum cistern.^[15]

In the present case, neuroendoscopic examination revealed that the cyst wall was covered with ependymal tissue and involved the internal cerebral veins at its inferior margin, which indicates the cyst was located in the velum interpositum. Furthermore, the locational relationship between the cyst and the internal cerebral



Figure 4: Endoscopic views. (a) View from the frontal horn of the right lateral ventricle showing a round protuberance on the septum pellucidum. The foramen of Monro, anterior septal, and thalamostriate vein are intact. (b) Small fenestration on the protuberance. (c) Semi-transparent true cyst wall under a thin ependymal membrane; the true cyst wall is held up with microforceps. (d) View of the interior of the cyst. The internal cerebral veins inferior to the cyst are seen through the semi-transparent cyst wall (arrows). Ant., anterior; For., foramen; Sept., septal; Str., striate; V., vein

veins suggests that the cyst originated from the upper layer of the tela choroidea [Figure 5b].

Arachnoid cysts in the ventricular system, where no arachnoid tissue exists, are a relatively well-known pathologic entity.^[9,10,17,21,22] To the best of our knowledge, however, only four cases of arachnoid cysts have been reported in the velum interpositum.^[3,4,6,16] Although the pathogenic mechanism of such lesions has not been discussed well, Spiegel *et al.* speculated that the tela choroidea, which developed as the fusion of two layers of "pia arachnoid," was a possible origin of the arachnoid cyst.^[16] Some authors also considered the tela choroidea as a source of the meningioma, which is derived from the arachnoid cap cells.^[5,7] Our interpretation of the endoscopic findings in the present case is consistent with their speculation.

Two entities are sometimes confused with arachnoid cysts of the velum interpositum. One is the cavum veli interpositi, a physiological dilatation of the velum interpositum cistern. It is a normal variant observed in premature and newborn infants, similar to the cavum septi pellucidi and cavum vergae.^[8,13] Although the cavum veli interpositi usually disappears as the brain matures, it is rarely seen in adults as a persistent primitive structure.^[13] The major characteristic of the cavum veli



Figure 5: Schematic illustrations showing a coronal plane of the cerebrum. Orange lines indicate the ependymal layer, while green lines indicate the embryonic pial layer. Illustration presenting the anatomic layers around the velum interpositum. (a) The velum interpositum is the space between the two layers of the tela choroidea. A small cyst originates from the upper layer of the tela choroidea. Enlargement of the cyst. (b) The cyst wall is covered with the ependymal layer, and the internal cerebral veins are shifted inferiorly due to compression by the cyst. Chor., choroid; Interposit., interpositum; Lat., lateral; Plex., plexus; Vent., ventricle

interpositi is that it has free communication with the quadrigeminal cistern,^[13] which was not observed in the present case according to both CT cisternography and surgical findings. Another confusing abnormality that should be differentiated from an arachnoid cyst of the velum interpositum is the one originating from the quadrigeminal cistern. Park et al. described a case of an arachnoid cyst of the lateral ventricle extending from the quadrigeminal cistern.^[12] An arachnoid cyst of the quadrigeminal cistern can mimic that of the velum interpositum if it extends in the superoanterior direction. The cyst of the quadrigeminal cistern, however, would displace the internal cerebral veins upward because the arachnoid membrane in the quadrigeminal cistern is topographically below the internal cerebral veins; this was not observed in the present case.^[1] Differentiation of an arachnoid cyst of the velum interpositum from a cavum veli interpositi and arachnoid cysts of the quadrigeminal cistern has been somewhat arbitrary in previous reports, which may explain in part why reports of arachnoid cysts of the velum interpositum are so rare.

Although memory deficit is a representative cognitive symptom caused by intracranial arachnoid cysts,^[20] we could not find any cases of a velum interpositum arachnoid cyst causing memory deficit in the literature.^[3,4,6,16] According to a review of 17 velum interpositum meningiomas,^[7] the other pathology originating from the velum interpositum, the most common symptoms included headache, papilledema, gait disturbances, and altered mental status.^[7] In their review, memory disturbance was attributable mainly to hydrocephalus: 10 of the 17 cases (59%) had hydrocephalus, with 4 of them suffering from dementia or memory disturbance. Our patient, however, suffered from severe memory disturbance without hydrocephalus. Reduced glucose metabolism in the bilateral cingulate gyri and thalami may be a key mechanism of memory impairment observed in the present case. Generally, an intracranial arachnoid cyst can reduce perfusion and metabolism in the surrounding cortical regions through its mass. These changes can cause mental impairments including memory impairment and are reversible once the cyst has been decompressed.^[18,20] In the present case, the cyst did not directly compress the cingulate gyri. However, the compression of the bilateral fornices and thalami may have affected the electric activity of a limbic circuit known as the Papez circuit,^[11] which may have resulted in memory deficit and secondary metabolic reduction of the cingulate gyrus as revealed by FDG-PET. This speculation is consistent with the report of Clarke et al.,^[2] who demonstrated secondary metabolic reduction of the cingulate gyrus after damage to the thalamus. Memory deficit may therefore be one of the major and treatable symptoms caused by an arachnoid cyst of the velum interpositum even if patients do not suffer from hydrocephalus.

CONCLUSION

Histologic and endoscopic findings of the present case suggest that arachnoid cysts of the velum interpositum are rare but distinct clinicopathologic entities that originate from the tela choroidea covering the velum interpositum. The lesions can cause memory disturbance without hydrocephalus due to compression of the fornices and thalami, but this can be reversed by surgery.

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