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Case Report

Late-onset aqueductal membranous occlusion treated neuroendoscopic procedure and consideration of its pathological findings: A case report

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ABSTRACT

Background: Aqueduct of Sylvius stenosis/obstruction interferes with cerebrospinal fluid (CSF) flow and leads to the non-communicating hydrocephalus. Acquired non-neoplastic causes of aqueduct of Sylvius stenosis/obstruction include simple stenosis, gliosis, slit-like stenosis, and septal formation, but the detailed mechanisms are not clear. In the present study, we experienced a case of late-onset aqueductal membranous occlusion (LAMO) successfully treated by neuroendoscopic procedure, which allowed us to examine the pathology of the membranous structures of the aqueduct of Sylvius occlusion.

Case Description: A 66-year-old woman presented with gradually progressive gait disturbance, cognitive dysfunction, and urinary incontinenc. Brain magnetic resonance imaging (MRI) showed enlargement of the bilateral lateral ventricles and the third ventricle without dilatation of fourth ventricle, and heavily T2-weighted images showed an enlarged aqueduct of Sylvius and a membranous structure at its caudal end. Gadolinium contrast-enhanced T1-weighted images showed no neoplastic lesions. We diagnosed this case that the hydrocephalus due to late-onset idiopathic aqueductal stenosis or LAMO and the patient underwent endoscopic third ventriculostomy and endoscopic aqueduct oplasty. Membranous tissue specimens were obtained from the occluded aqueduct of Sylvius at the time of treatment. Histopathological examination revealed gliosis, and inside the gliosis, there were cell clusters that appeared to be ependymal cells and were corpora amylacea. We confirmed CSF flow at the site of obstruction of the aqueduct of Sylvius and the stoma of the third ventricle floor by MRI images. Her symptoms were improved immediately.

Conclusion: We experienced a case of LAMO successfully treated by neuroendoscopic procedure, which allowed us to examine the pathology of the membranous structure of the aqueduct of Sylvius. The pathological study of LAMO is rare, and we report it, including a review of the literature.

Keywords: Hydrocephalus, Late-onset aqueductal membranous occlusion (LAMO), Neuroendoscopic surgery, Pathology

INTRODUCTION

The aqueduct of Sylvius is one of the narrowest pathways of cerebrospinal fluid (CSF), and its obstruction can be a cause of hydrocephalus. Late-onset aqueductal membranous occlusion (LAMO) is known as a non-neoplastic and acquired disorder of the aqueduct of Sylvius.[7] There

have been few reports of LAMO. Non-neoplastic causes of aqueduct of Sylvius stenosis/obstruction include simple stenosis, gliosis, slit-like stenosis, and septal formation. However, the detailed mechanisms are not clear. There have been no reports on the pathology of acquired membranous structure occluding the aqueduct of Sylvius that we could find. In this report, we describe a case of LAMO that was successfully treated by neuroendoscopic procedure, and its pathological considerations of the membranous structure.

CASE REPORT

A 66-year-old woman presented with progressively worsening gait disturbance and urinary incontinence over a year. She was diagnosed with hydrocephalus and was admitted to our department for further examinations and treatment.

She had a history of hypertension, hyperlipidemia, and cholecystitis, and no history of head trauma, encephalitis, or meningitis and other hemorrhagic events that would cause hydrocephalus.

Neurological findings revealed no consciousness disturbance, but slow overall movement, and a wide-base open-leg gait. Timed up and go test was 18.9 s. The preoperative mini-mental state examination (MMSE) score was 20/30 and the frontal assessment battery (FAB) score was 10/18. There was no motosensory disturbance, and rigidity, tremor, or head enlargement.

CSF study meant color: Watery clear, initial pressure: 15 cm/H_2O, cell count: <1/uL, PH: 7.4, protein:30 (10–40) mg/dL, sugar:73 (50–75) md/dL, and chloride:113 (110–125) mEq/L. Blood analysis showed no inflammatory findings.

Brain magnetic resonance imaging (MRI) showed enlarged lateral and third ventricles without the dilatation of fourth ventricle. Narrowing of the subarachnoid space in the parietal region (high cranial area and longitudinal fissure) and heterogeneous enlargement of the basilar and Sylvian fissures indicated idiopathic normal pressure hydrocephalus was not observed [Figures 1a-c]. Heavily T2-weighted images showed an enlarged aqueduct of Sylvius and a membranous structure at its caudal end and the bulging of the floor of the third ventricle [Figure 1d]. T1-weighted images with gadolinium showed no enhanced lesion [Figure 1e]. Phase-contrast cine MRI confirmed the cessation of CSF flow in the aqueduct of Sylvius.

We diagnosed with hydrocephalus caused stenosis or obstruction of aqueduct of Sylvius.

Preoperatively, endoscopic third ventriculostomy (ETV) alone was considered, but MRI showed a funnel-shaped enlargement of the entrance to the aqueduct of Sylvius with an obstructive mechanism at its base, and intraoperatively, it was recognized that the membrane structure in the obstructed area was very thin, so we decided to perform aqueduct plasty as well. When performing the procedure, we took great care to avoid mesencephalic injury using small forceps. Our endoscopic surgical corridor planned preoperatively from the anterior horn of the right lateral ventricle to the third ventricle floor and the aqueduct of Sylvius was presented [Figures 1a-c].

Hence, we performed ETV, and endoscopic aqueduct plasty (EAP) as below. Postoperative MRI showed a reduction in ventricular size [Figure 2a], the corridor used in the endoscopic surgery is shown in [Figures 2b-d]. Moreover, heavily T2-weighted images showed the opening of the aqueduct of Sylvius [Figure 2e]. Phase-contrast cine MRI showed CSF flow at the floor of the third ventricle [Figure 2f]. Gait speed improved to 10.9 s in the timed up and go test. Cognitive function improved to MMSE score 30/30 and FAB score 14/18. Urinary incontinence improved. On postoperative day 37, she was discharged without complications.

Intraoperative findings

The flexible endoscope (Olympus VEF type V, Olympus, Tokyo, Japan) was inserted through anterior horn of the right lateral ventricle through a transparent sheath (Neuroport mini, Hakko, Tokyo, Japan). An enlarged lateral ventricle, foramen of Monro, third ventricle, and stretched interthalamic adhesion were observed, and the septum pellucidum was partially perforated [Figure 3a]. The enlarged aqueduct of Sylvius was occluded by a white thin membranous structure. EAP was performed and get some specimens of membranous structure. Patency of CSF was observed in the perforated area [Figures 3b-d]. Moreover, ETV was performed on the thinning base of the third ventricle after EPA [Figure 3e]. CSF flow between the third ventricle and prepontine cistern through a stoma was observed [Figure 3e]. The Lilliquist's membrane was thinned out [Figure 3f].

Histopathological findings

Histopathologically, we examined the thin membranous structure that occluded the aqueduct of Sylvius. Hematoxylin Eosin: Staining revealed a mild increase of glia cells in the neuropil, reflecting gliosis. Within the cellular matrix, cell aggregates with an island-like distribution and loss of continuity which were observed [Figures 4a and b]. These cells, which formed island-like cell clusters, were positive for glial fibrillary acidic protein: and epithelial membrane antigen: Immunostaining [Figures 4c and d]. These findings were suggesting that they have the character of ependymal cells. Corpora amylacea that stained mild basophilic were also observed in the neuropil [Figure 4e].

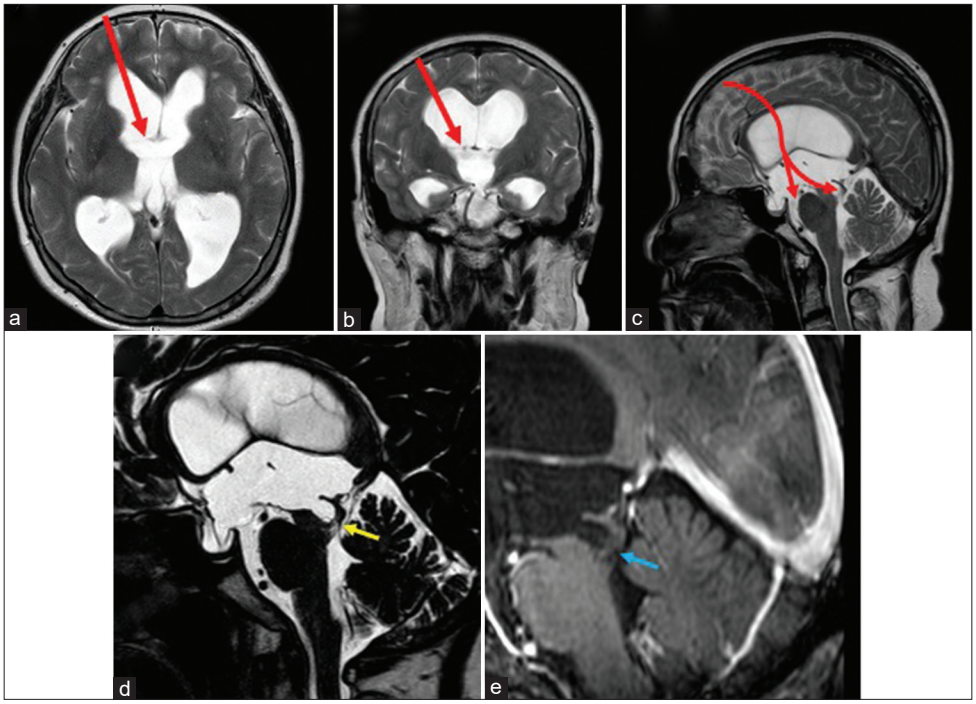

Figure 1: Magnetic resonance imaging pretreatment findings: (a-c) T2WI showed ventricular dilatation of the lateral and the third ventricles but not of the fourth ventricle. Preoperative planning of the endoscopic corridor was shown as red arrow. (d) Heavily T2WI showed an enlarged aqueduct of Sylvius and a membranous structure at its caudal end (yellow arrow). (e) T1WI with gadolinium did not demonstrate any enhanced lesion (blue arrow).

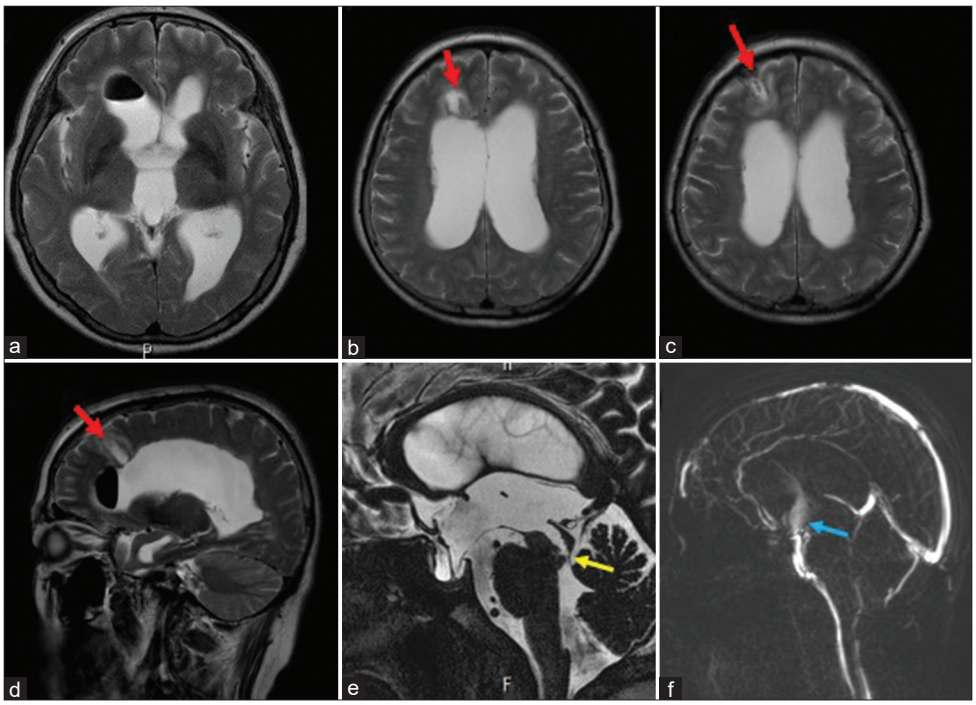

Figure 2: Magnetic resonance imaging (MRI) posttreatment findings: (a-d) T2WI axial and saggitalimages showed mild ventricular shrinkage and was seen surgical collider at the right frontal lobe (red arrow). (e) Heavily T2WI showed the opening of the aqueduct of Sylvius (yellow arrow). (f) Phase contrast cine MRI showed cerebrospinal fluid flow at the stoma of the third ventricle floor (blue arrow).

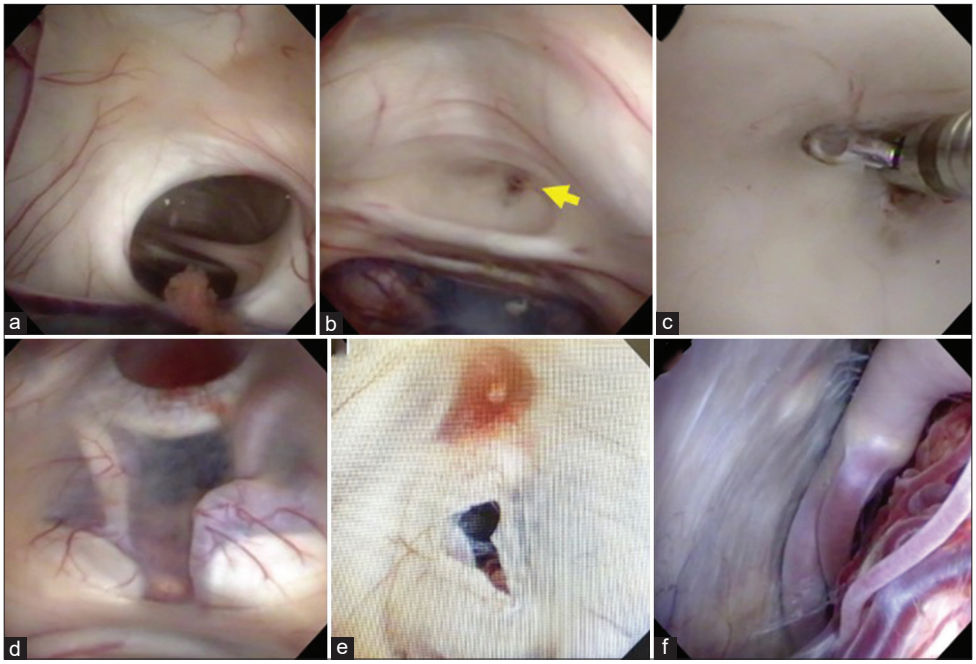

Figure 3: Intraoperative findings (a) foramen of Monro was dilated and observed stretched interthalamic adhesion. (b) Membranous structure (yellow arrow) occluded the aqueduct of Sylvius. (c) Membranous structure was removed with forceps. (d) The floor of the third ventricle was thinning (e) A stoma was created at the floor of the third ventricle. (f) The Lilliquist's membrane was thinning and defects in prepontine cistern.

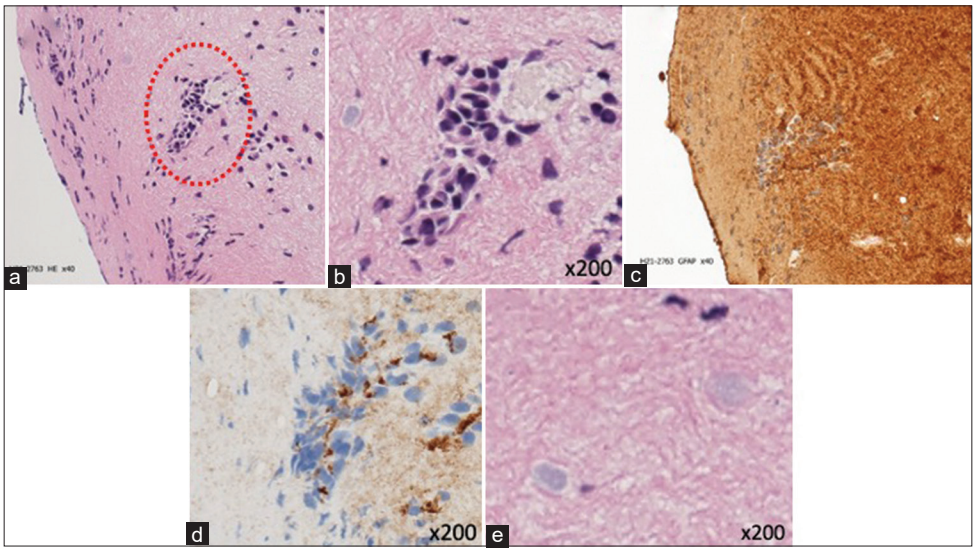

Figure 4: Pathological findings (a and b) island-like cell aggregates with loss of continuity were seen in the neuropil on staining (red dotted circle). (c and d) Cells forming island-like aggregates were glial fibrillary acidic protein (d) positive and epithelial membrane antigen (d) positive. (e) Corpora amylacea were scattered in neuropil on staining.

DISCUSSION

LAMO is known as a disease that causes hydrocephalus due to non-neoplastic aqueduct of Sylvius occlusion. The disease concept was first proposed by Matsuda *et al*. in 2011,[7] and Terada *et al*. defined it in 2020.[10] They defined LAMO as: (1) The presence of a membranous structure in the aqueduct of Sylvius and no CSF flow in the aqueduct

of Sylvius; (2) no other occlusive lesion, and no recent subarachnoid hemorrhage, intraventricular hemorrhage or meningitis that could cause hydrocephalus; (3) ventricular dilatation of the lateral and third ventricles but not of the fourth ventricle; and (4) not diagnosed in childhood. A similar disease concept called late-onset idiopathic aqueductal stenosis: LIAS is also known. Fukuhara and Luciano[5] defined LIAS as follows: (1) no lesions other than hydrocephalus in the brain, (2) small size of the fourth ventricle compared to the lateral and third ventricles, (3) no history of cerebral hemorrhage or meningitis, (4) no history of surgical treatment for hydrocephalus, and (5) no diagnosis in childhood. The exclusion of complicated cases of intracranial lesions is emphasized. In LIAS, the aqueduct of Sylvius shows stenosis, whereas in LAMO, a septum can be seen in the aqueduct of Sylvius, resulting in enlargement of the mesencephalic aqueduct proximal to the septum.

The common clinical presentation of these disease groups is often headache in young-onset cases and symptoms similar to the clinical presentation of normal pressure hydrocephalus in the elderly. The previous reports have reported the usefulness of sagittal and coronal sections of heavily T2-weighted images for imaging evaluation of membranous structures.[2,9]

Fifteen cases of LAMO have been reported[1,3,4,6,7,8,10,11] [Table 1] and surgical treatment was performed by ETV or EAP, or both. When EAP is performed, preoperative and intraoperative findings should be evaluated and adequate attention should be paid to mesencephalic and aqueduct injury. In this case, the intraoperative findings showed that the membranous structure of the aqueduct was very thin and their tips were transparent. Moreover, we confirmed that the entrance to the aqueduct of Sylvius was enlarged and that adequate endoscopic manipulation could be performed, so we performed EAP carefully. The flexible videoscope is a useful tool in this case due to its ease of use and high image quality.

As far as we could find, there were no reports of detailed pathological examination about the membranous structures. The exact mechanism of formation of the septa that occlude the aqueduct of Sylvius is unknown. In this case, we were able to histopathologically examine the membranous structure that formed in the aqueduct of Sylvius.

Histopathologically, membranous structure in the aqueduct of Sylvius showed increased glia cells, reflecting gliosis, and cell aggregates of ependymal cells. In addition, Corpora amylacea, which are clumps of polyglucosamine that accumulate at the tips of astrocyte projections, were found in the neuropil. In the previous reports, Del Biggio[3] and Cavanagh[1] have reviewed the pathological findings of the damaged ventricular wall. They have examined the distribution of ependymal cells and the accumulation of Corpora amylacea in the neuropil. They mention the possibility that ependymal cell clusters may persist in the gliosis as a result of some accidental ependymal injury and that ependymal regeneration, but with a failure of the repair mechanism, may promote gliosis with the accumulation of Corpora amylacea in periventricular astrocytes. Marta *et al.*[6] also mentioned that Corpora amylacea arise during aging and tissue repair processes. In the present case, some kind of accidental ependymal injury would have added chronic stress to the periventricular astrocytes, resulting in repeated gliosis

Table 1: Previously reported LAMO cases.

S. No.	References	Age	Sex	Symptom	Operation	Outcome
1.	Chen *et al.*[2]	9	F	Headache, vomiting	EAP	Imoroved
2.	Steel *et al.*[9]	10	M	Headache, vomiting	Direct surgery	Imoroved
3.	Chen *et al.*[2]	20	M	Headache, vomiting	EAP	Imoroved
4.	Chen *et al.*[2]	24	F	Headache, vomiting	EAP	Imoroved
5.	Chen *et al.*[2]	26	M	Headache, vomiting	EAP	Imoroved
6.	Chen *et al.*[2]	28	F	Headache, vomiting	EAP	Imoroved
7.	Schroeder and Gaab[8]	31	F	Headache, vomiting, seizure	EAP	Imoroved
8.	Chen *et al.*[2]	33	F	Headache, vomiting	EAP	Imoroved
9.	Terada *et al.*[10]	36	M	Headache, loss of consciousness	EAP+ETV	Imoroved, Diplopia
10.	Schroeder and Gaab[8]	46	F	Headache, cognitive disfunnction	EAP	Not improved
11.	Matsuda *et al.*[7]	57	M	Gait disturbance	EAP+ETV	Improved
12.	Schroeder and Gaab[8]	66	M	Gait disturbance, cognitive disfunction, urinary disfunction	EAP	Not improved
13.	Schroeder and Gaab[8]	66	F	Headache, gait disturbance, cognitive disfunction, urinary disfunction	EAP+ETV	No data
14.	Chen *et al.*[2]	67	F	Blurred vision, loss of consciousness	EAP	Imoroved
15.	Utsuki *et al.*[11]	71	M	Gait disturbance, cognitive disfunction	ETV	Improved
16.	Our case	66	F	Gait disturbance, cognitive disfunction, urinary disfunction	EAP+ETV	Imoroved

LAMO: Late-onset aqueductal membranous occlusion, EA: Endoscopic aqueductoplasty, ETV: Endoscopic third ventriculostomy, F: Female, M: Male

formation and ependymal regeneration failure. Reflecting these findings, histopathology would have confirmed gliosis with clusters of ependymal cells and Corpora amylacea. As a result of these ependymal repair failures, membranous structures formed in the mesencephalic aqueduct, and resulting non-communicating hydrocephalus.

CONCLUSION

We experienced a case of LAMO successfully treated by neuroendoscopic procedure, which allowed us to examine the pathology of the membranous structure of the aqueduct of Sylvius. The histopathological findings suggested that the result of ependymal repair failure may have contributed to the obstruction of the aqueduct of Sylvius.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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