

THERAPY

Endoscopic treatment of midline cerebral cysts

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Abstract

Axial cerebral cysts of various location and origin are relatively rare lesions. Endoscopic neurosurgery has made them amenable for safe intervention even in vital brain regions. Neuroendoscopic treatment was used by the author in 35 patients during the last 7 years without permanent morbidity or mortality. The aim of the surgery was to fenestrate the cyst or to perform cyst extirpation to relieve the pressure effect on the surrounding functioning neural tissue and to reestablish the patency of cerebrospinal fluid pathways, as well as histological verification of the cyst origin.

Contemporary imaging modalities are capable of diagnostic evaluation, but the detailed anatomical relationship to the surrounding structures can be visually analysed completely together with understanding of their functional effects only during endoscopic surgeries, supported by the results of imaging. Therefore the precision of endoscopic surgeries was substantially enhanced and during the surgery a large amount of knowledge enriching the contemporary information about the cerebrospinal fluid flow through the cerebral ventricular system and blood circulation in the deep cerebral venous system and the relationship of these veins to hydrocephalus was obtained (Tab. 1, Fig. 9, Ref. 11).

Key words: endoscopy, navigation, cyst.

Midline cerebral cysts, also known as axial cysts (1) are located within the confines of the narrow aqueductal area and non-paired cerebral ventricles. Generally speaking their formation is attributed to the maldevelopment of structures participating in the formation of the brain together with the meninges, especially when the subarachnoidal space is being formed in the tentorial incisura (2), which contains the brainstem structures.

In this difficult space, cysts related to the roof and floor of the third ventricle as well as the outlets of the fourth ventricle arise. Due to their location these cysts may interfere with cerebrospinal fluid circulation, but they are able to directly endanger cerebral function due to their intimate relationship to the underlying brain tissue. Their clinical manifestations can threaten directly the patient's life, and therefore surgical treatment is mandatory.

Even if some of these cysts can be treated by standard neurosurgical techniques (3), for instance fourth ventricular cysts and cysts located in the craniocervical junction, surgical interventions for some of them, especially third ventricular colloid cysts and aqueductal cysts are considered to be difficult with occasionally stormy postsurgical course (4). The surgical approach itself can be responsible for some degree of brain tissue damage.

Minimally invasive stereotactic technique gave rise to substantial progress in the treatment of midline cysts (5).

However the most profound change appeared just after the development of endoscopic neurosurgery (6). The surgical procedure itself is supported by stereotactic precision, especially when employing computerised navigation with virtual imaging. This progress led directly both to an improvement in surgical safety and success rate.

Group of patients and methods

During the last 5 years 35 patients with midline cysts (Tab. 1) underwent endoscopic treatment at the Neurosurgery Clinic, Faculty Hospital St. Ann's Brno, apart from postsurgical pseudocysts after tentorial meningioma surgery and posttraumatic

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Supported by IGA MZ ČR NR/7805-2.



Fig. 1. MRI imaging of velum interpositum cyst with internal cerebral veins stretched over its surface.

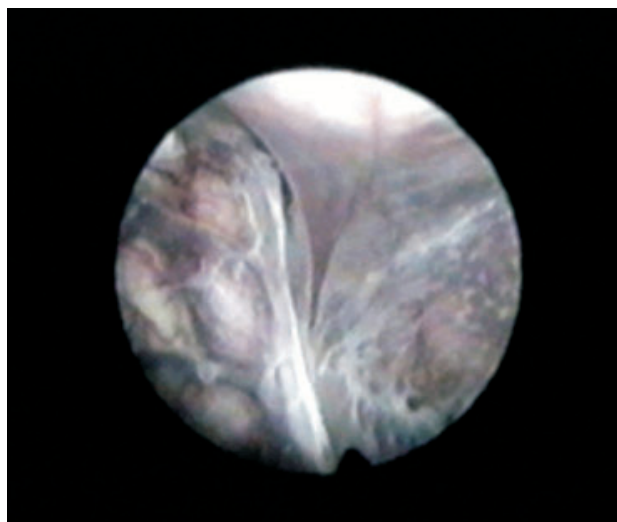


Fig. 2. Compression of internal cerebral vein.

pseudocyst after upper spinal cord injury. The age of clinical manifestation varied from 12 to 68 years with mean 38.75 years, with males being more frequently affected.

The history and symptoms of the individual patient depended on the cyst location. With the exception of suprasellar cysts, the key presentation was that of an obstacle blocking normal cerebrospinal fluid circulation, therefore various degrees of hydrocephalic dilatation of brain ventricles were present together with occasional incipient papilloedema.

The main complaints were headaches and unsteady gait, followed by problems related to oculomotor muscles dysfunction or convergence disturbances.

Focal findings were directly related to the cyst location and their clinical course can be better explained by short description of the clinical course together with the treatment result.

Septum pellucidum cyst is reported to be clinically silent, although some behavioral disturbances from the schizophrenic area are attributed to this lesion. Septostomy is then recommended to establish free communication between the cyst and ventricular cavity. However septostomy is more frequently indicated in

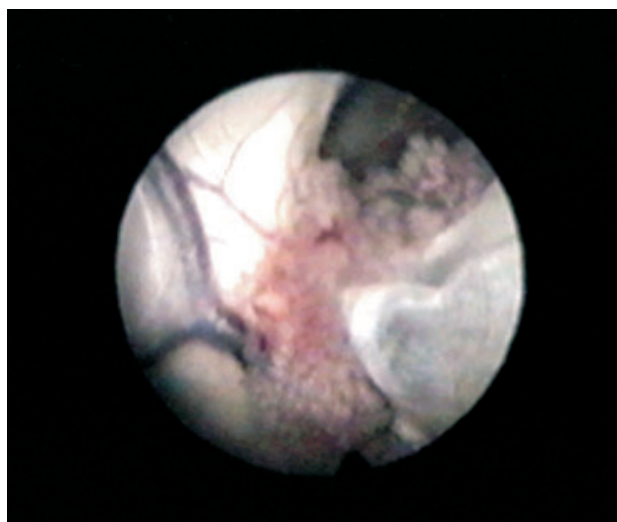


Fig. 3. Thalamostriate vein overfilled with stagnating blood.

Tab. 1. Axial cysts location.

Septum pellucidum	2
Cavum Vergle	2
Velum interpositum	3
Aqueductal area	3
Corpus pineale	2
IIIrd ventricle Rathke cleft cyst	2
colloid cyst	6
chorioid plexus cyst	3
IVth ventricle	2
Cisterna magna	8
Lamina terminalis cyst	1
Suprasellar cyst	3
Total	35

asymmetrical hydrocephalus cases, as confirmed also by our experience.

Cavum Vergae cyst situated over the anterior part of the third ventricle presented as facial tics and collapses, that were initially considered to be of epileptic origin due to EEG findings. After cyst fenestration to the right lateral ventricle the problems disappeared.

Cysts affecting the velum interpositum area over the posterior part of the third ventricle extending sometimes as far as the quadrigeminal plate (7) occasionally elicit headaches and their pathophysiological effects were explained by intraoperative findings during endoscopic surgery. In one case the cyst was found to compress the Galenic vein directly together with internal cerebral veins (Fig. 1, 2), therefore their tributaries, thalamostriate veins were distended by the stagnating blood inside them (Fig. 3). This fact may be considered to be the cause of communicating



Fig. 4 a, b. Posterior commissure floating freely above the aqueductal orifice.

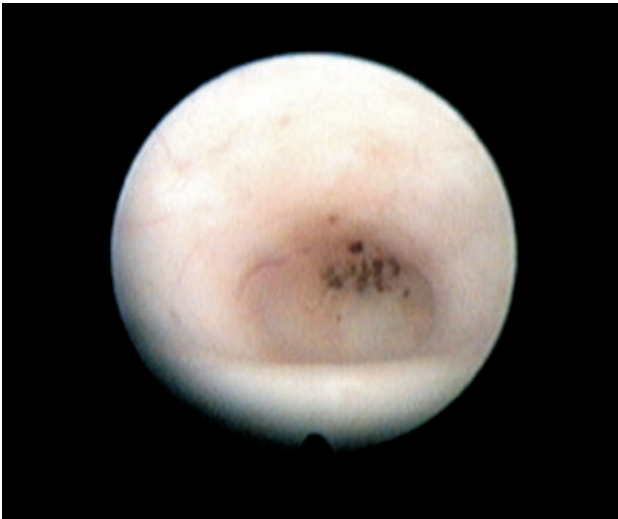


Fig. 5. Enormous dilatation of the aqueductal entry reaching as far as nigral nuclei.



Fig. 6. MRI appearance of third ventricle colloid cyst.

hydrocephalus, because the granulations found inside the deep cerebral vessels take part in cerebrospinal fluid resorption. After endoscopic surgery, the Galenic vein and internal cerebral veins became more markedly filled and the tributaries collapsed. In another case, the cyst compressed and elevated the posterior commissure and habenular area from below, so that these structures floated freely as an isolated transverse wall against the aqueductal orifice (Fig. 4 a, b). Probably due to the functional disturbance of the structures mentioned above the patient reported memory disturbances before surgery. The creation of a communication connecting the septum pellucidum cyst, cavum Vergae cyst and velum interpositum cyst with the lateral ventricle by means of neuroendoscopic technique is the procedure of choice.

Out of the three cysts located in the aqueductal area, one cyst was responsible for the dilatation of aqueductal orifice. The

aqueductal walls were deeply impressed into the midbrain tegmentum, therefore dark spots arising from substantia nigra became apparent (Fig. 5). The functional disturbance of the quadrigeminal plate was probably responsible for the reported feeling of shaking of the patient's visual field together with the damage of the secondary visual centers – upper collicular structures. From the cyst causing marked hydrocephalic dilatation of the ventricular system, only thin membrane remained and the membrane was amenable for endoscopic intervention. The second cyst with superficial vascular network caused hydrocephalus with papilloedema when acting as a curtain blocking the aqueductal orifice. Besides intractable headaches, the patient reported vertigo and unsteadiness due to visual impairment. The coagulation of the cyst wall together with cyst biopsy was enough to open the aqueduct. Histological investigation revealed the cyst sur-

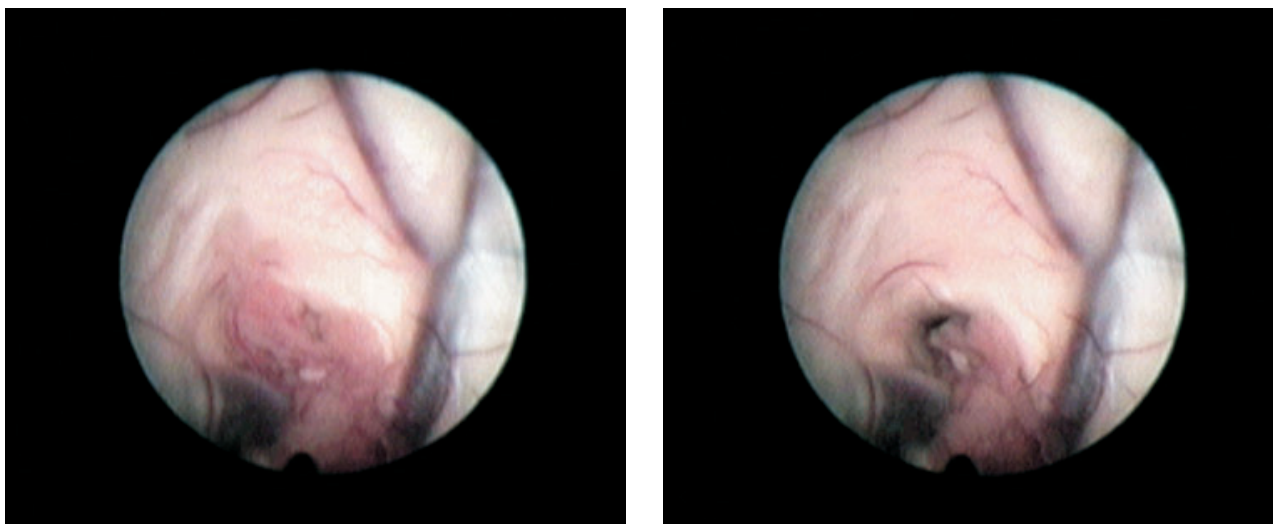


Fig. 7 a, b. Flat valve formed by choroidal plexus lobule alternatively closing and opening the foramen of Monro.

face covered by single layered hydropic epithelium, seated on an oxyphyllic membrane. In another patient, initially referred for surgery as normotensive hydrocephalus case, a cystic lesion with aqueductal compression and marked narrowing was treated during the surgical procedure.

Pineal cyst, discovered incidentally after head injury on MRI investigation, was considered to be asymptomatic. However hydrocephalic ventricular dilatation was present and biopsy specimen revealed the pathognomic finding of loose cellular tissue, with cells harboring differentiated round nuclei and intercellular fibrillary substance. All these cysts were approached endoscopically from the posterior part of the third ventricle.

In 6 colloid cyst cases the final diagnosis was established by their typical appearance (Fig. 6). Two of these patients suffered from epileptic seizures, in one case long lasting, in the other the diagnosis was established after short period of seizures. After surgery the patients became gradually seizure free.

In another two cases the final finding was a Rathke cleft cyst. The cyst was the underlying cause of long lasting gait unsteadiness and vertigo in two elderly patients, with symptom disappearance after cyst treatment. Histological investigation proved the cysts to be of hamartogenous endodermal origin, with layered lining, in some sites showing features of nonkeratinised squamous epithelium. Moreover, an inconstant layer of low basal cells was found with prominent secretion of acidic and neutral mucopolysaccharidic substance, which formed the content of the cyst. A choroid plexus cyst was found in an asymmetrical location inside the third ventricle in one case. In another case it nested directly inside the foramen Monro area, with features of a ball valve, periodically blocking and opening the interventricular foramen in rhythm with the heart beat (Fig. 7 a, b). The analysis of endoscopic surgery report has unequivocally proven that the flow of cerebrospinal fluid through the ventricular system is pulsatory with the lateral ventricles acting as a secondary heart. Univentricular hydrocephalus observed in both cases was

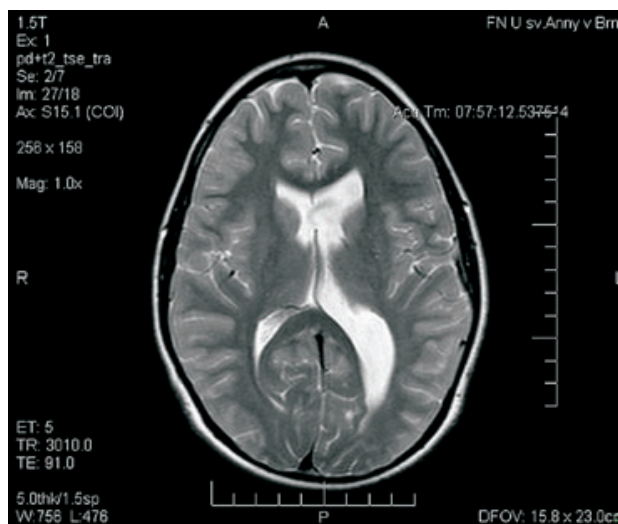


Fig. 8. MRI visualization of cerebrospinal fluid flow through the septostomy.

treated by direct intervention on the blocking lesion and septostomy was added as a safety measure to prevent symptom recurrence (Fig. 8).

Two patients with a cyst affecting the fourth ventricle area related to Dandy Walker syndrome were treated by cystoperitoneal shunting in the past. The malfunction of the drainage system presented with headaches and papilloedema. In one patient long lasting oculomotor problems were described before surgery and the second patient followed for infantile palsy had permanent epicystostomy inserted before surgery. A chronic urinary tract infection was responsible for temporary fever in the postoperative period.

Visual field narrowing was the main complaint of patients with suprasellar arachnoidal cyst. The trajectory was targeted to the floor of the third ventricle (Fig. 9).

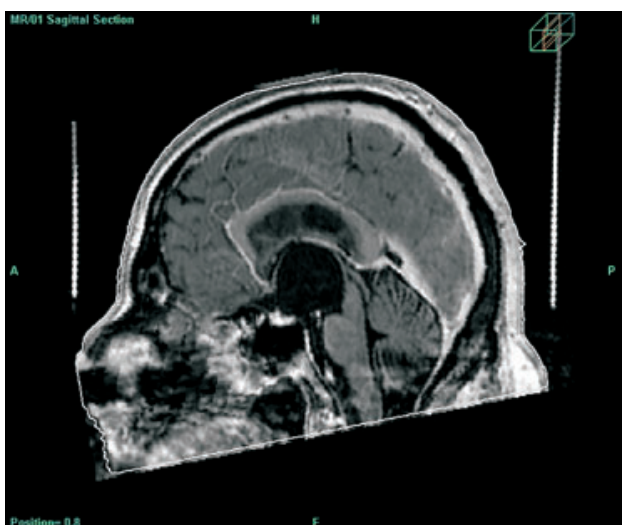


Fig. 9. MRI image of suprasellar arachnoid cyst.

All endoscopic surgeries on axial cysts were supported by stereotactic navigation and computer planning software – Praezis Plus with MRI as a primary source of navigation data. The insertion of Wolf endoscope along the pre-planned trajectory was safe. The surgeries were performed under general anaesthesia.

The aim of the surgery was the fenestration of the cyst wall, the creation of communication between the cyst and the cerebrospinal fluid circulation pathways, and reopening of cerebrospinal fluid circulation and radical cyst resection, when feasible. Based on the actual situation, the patency of cerebrospinal fluid circulation was secured by aqueductoplasty, endoscopic third ventriculostomy or septostomy. The free flow of cerebrospinal fluid to the basal cisterns and posterior fossa was repeatedly checked by intraoperative ventriculography. For subsequent follow-up non invasive MRI flow sequences were exclusively used. A sample for histological evaluation was always taken from the cyst wall and the entire surgery was recorded by videocamera.

Results

Because the surgical trajectory was always meticulously prepared preoperatively to establish the optimal direction of endoscope navigation, there were no surgical complications related to the approach. Temporary oculomotor dysfunction (difficulties with lateral view) appeared just after coagulation of the vascular network located on the cyst surface in case of aqueductal cyst. Transient nocturnal confusion after total removal of the Rathke cleft cyst intimately related to the third ventricle wall was probably related to the functional disturbance of thalamic structures located in close relationship to the lesion.

There was neither permanent surgical morbidity, nor mortality related to the procedure. On the other hand, the patients were always satisfied with the final outcome of surgery.

This is undoubtedly related to the fact that endoscopic visual evaluation of the local situation discovers conditions and circumstances, that cannot be identified preoperatively on CT or

MRI scans. During the same time, the true cause of problems is recognised, therefore causal and rational treatment procedure can be implemented.

Simple endoscopic inspection of the cyst interior with septations may explain the failure of cystoperitoneal shunt, which may be due to failure to drain multiple cyst compartments.

What is of much more importance is the discovery of the pathophysiological mechanism underlying the formation of univentricular hydrocephalus as a flat valve located in foramen of Monro. Between the affected lateral ventricle and third ventricle alternating pressure changes take place acting on both sides of the flap-like a valve dependent on heart rate. In mathematical terms this function was described by K.Kosorin M.Sc., Ph.D., from the Institute of Hydrology, Slovak Academy of Sciences by the differential equation:

$$\Delta V / \Delta t = \pm k \times (p_2 - p_1) \times S$$

The mechanism responsible for maintenance of pressure homeostasis in the cranial cavity is substantiated by keeping the steady rate of blood and cerebrospinal fluid exchange V during time unit t between two cardiac systoles, transmitted to the cerebral ventricles. This value is determined by the difference between pressures in the participating cerebral ventricles p_1 and p_2 and the size of communicating orifice S between them and the actual situation is determined by onstant k .

The communication between the participating cerebrospinal fluid spatial compartments is essential. The term critical size is a key to the proper understanding of the valve phenomenon in cerebrospinal fluid circulation pathways also when related to the formation of active hydrocephalus or cysts formation and development. With high probability this phenomenon can be also applied to aqueductal stenoses and to the previous endoscopic observations, in which the posterior commissure and habenular structures undermined by the cysts floated as a transverse wall against aqueductal orifice with intermittent blockage. Pressure relationships arising under such circumstances form the theoretical principles of the endoscopic neurosurgery and are contained in the research project mentioned below.

The compression of the Galenic vein and internal cerebral veins by velum interpositum cyst is of similar importance. The mechanism is responsible for blood stagnation inside the deep cerebral venous system and may be also be the cause of communicating hydrocephalus.

Discussion

There are various factors acting during embryogenesis responsible for the formation of axial cysts, for instance traction theory due to caudal neuropore fixation in hindbrain malformations, or hydrodynamic theory, in which cerebrospinal fluid is the main morphogenetic factor, giving rise to dural folds and sub-arachnoidal space. These theories were dealt with in detail by Hackel and Beneš (8). Hamartogenous changes are responsible for cyst formation inside the third ventricle or pineal region.

Clinical symptomatology varies due to different cyst location, but generally speaking these symptoms are related to the actual functional status of the cerebrospinal fluid space or surrounding neural tissue.

The diagnostic workup in arachnoidal cyst is relatively straightforward when employing modern imaging techniques, especially MRI and therefore these lesions become a subject of surgical treatment. Up to now drainage operations or microsurgical fenestration in favourably located cysts are the prevailing modes of treatment (9).

Stereotactic surgeries are a step towards further progress in the treatment possibilities. They substantially widened the amenability of axial cysts for surgical treatment, especially when causing the blockage of important venous structures (10).

Nowadays, endoscopic treatment of axial cerebral cysts is a new and evolving modality. The present observations express the disadvantage of prevailing therapeutic modes (11) which rely on drainage systems or direct surgical approach. Endoscopy as an up-to date modern, minimally invasive surgery is a progressive step not only in surgical safety, but especially when considering the outcome. New perspectives for the study of cerebrospinal fluid space functions are being discovered by this technique.

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Received December 12, 2005.

Accepted December 20, 2005.