

Septum Pellucidum Cyst in Patient with Parkinson's Disease-minimally Invasive Staged Therapy

**K. Ghallab¹, Z. Novák¹, J. Chrastina*¹, M. Baláž²
and I. Říha¹**

*1 Department of Neurosurgery MF MU, St. Anne's Teaching Hospital Brno,
Czech Republic*

*2 First Department of Neurology MF MU, St. Anne's Teaching Hospital, Brno,
Czech Republic*

ABSTRACT

Background: The aim of the case report is to present the minimally invasive staged treatment of a patient with motor complications of Parkinson's disease with a large septum pellucidum cyst.

Material and Method: A two-stage surgical strategy was proposed by the movement disorders team: neuroendoscopic cyst fenestration with subsequent implantation of deep brain stimulation electrodes (subthalamic nucleus) depending on the effect of neuroendoscopic treatment.

Results: Despite the technical success of a neuroendoscopic cyst fenestration, as evidenced by a reduction in cyst volume, intraoperative ventriculocystography and postoperative CT showing free communication between the cyst and the ventricular system the motor symptoms caused by Parkinson's disease remained unchanged. A bilateral subthalamic deep brain stimulation system was implanted with good functional results as documented by her UPRDS III score in the off-medication state (52 points preoperative and 34 point after 6 years postoperative) and reduction of L-DOPA equivalent dose (60% of the pre-deep brain stimulation level).

Conclusion: Minimally invasive therapy-neuroendoscopic cyst fenestration, followed by a bilateral subthalamic stimulation implantation-led to the substantial and lasting improvement of the patient's symptoms.

Key words: septum pellucidum cyst, Parkinson's disease, deep brain stimulation, neuroendoscopy



*Correspondence: Jan Chrastina, M.D., Ph.D., Department of Neurosurgery MF MU, St. Anne's Teaching Hospital, Pekarska 53, 656 91 Brno, Czech Republic, phone: 00 420 543 182 697, fax: 00 420 543 182 687, e-mail: jan.chrastina@fnusa.cz

INTRODUCTION

The cavum septi pellucidi and cavum vergae are fluid collections between the leaflets of the septum pellucidum. The prevalence in children declines with age and in adults varies significantly depending on the criteria used for diagnosis as some remnant can be present in all patients. The prevalence of abnormal cavum septi pellucidi (cavum septi pellucidi contained on four or more 1,5 mm MRI slices) is 10,3 % for normal subjects [1]. The cavities may occasionally enlarge into cysts (by definition laterally bowing walls 10 mm apart or greater) and cause neurological symptoms. Although the causal relationship with neurological and psychiatric symptoms remains unclear, there are data reporting the association of septal cysts with some functional brain disorders (epilepsy, psychiatric problems) [2,3]. The aim of this paper is to present a case report of a patient referred for subthalamic deep brain stimulation (DBS) with a large midline cyst discovered by presurgical imaging studies. Minimally invasive therapy – neuroendoscopic cyst fenestration, followed by a bilateral subthalamic stimulation – led to the substantial and lasting improvement of the patient's symptoms.

MATERIAL AND METHODS

A 51-year-old female with late motor complications of Parkinson's disease (PD) was referred for deep brain stimulation. There was no family history of PD, and the patient did not complain of any previous neurological or psychiatric problems. She had been suffering from PD for 12 years, since the age of 39. L-DOPA therapy was started one year after the onset of symptoms. A positive therapeutic effect from this treatment lasted for 8 years, when "wearing-off" symptoms and disabling biphasic dyskinesias affecting the neck and upper and lower extremities appeared. The symptoms were uncontrollable by medication. No intellectual, cognitive, or memory dysfunctions were detected during the routine presurgical evaluation. The patient was scheduled for bilateral DBS subthalamic electrode implantation. A routine presurgical MRI revealed a large midline cyst with the bulging of the lateral walls and interventricular foramina narrowing (Fig.1). Although there were no symptoms indicating intracranial hypertension, a two-stage surgical strategy was proposed by the movement disorders team: neuroendoscopic cyst fenestration and subsequent implantation of DBS electrodes.

The first step (septum pellucidum cyst neuroendoscopic fenestration) was performed using frame based stereotactic system (ceramic frame Leibinger, 3D T1 WI, Zamorano Dujovny, Praezis Plus). The trajectory for a safe neuroendoscopic surgery was planned, with the entry site selected so as not to complicate the planned DBS implantation – anterior to the presumed point in front of the coronal suture (Fig. 2). Thinned and bulging walls suggesting expansive behaviour of the midline cysts together with slit like foramina of Monro were observed immediately after the endoscope entered the ventricular system (Fig. 3). After an uneventful fenestration (of both cyst walls), intraoperative ventriculocystography and postoperative CT showed free communication between the cyst and the ventricular system.

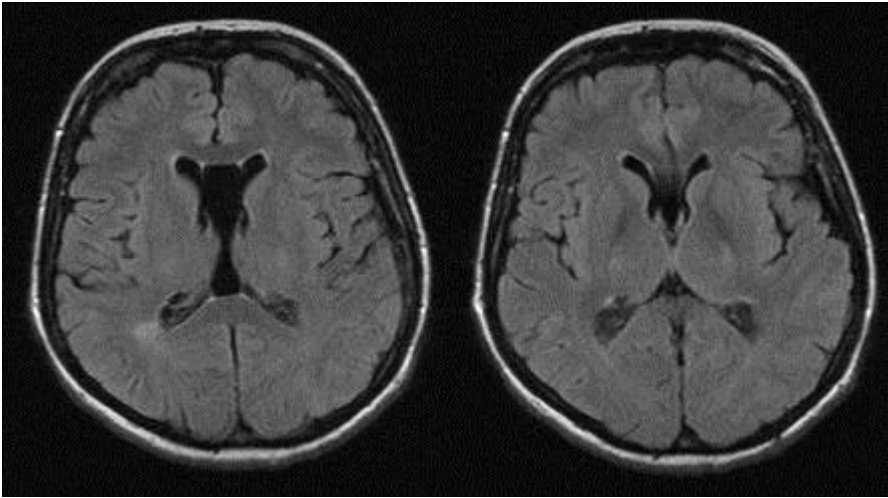


Figure (1): MRI findings - cyst wall bulging, the width of the cyst 16 mm



Figure (2): Presurgical planning of neuroendoscopic cyst fenestration



Figure (3): Endoscopic findings during surgery - bulging and thinning of the cyst walls

RESULTS

A volumetric MRI follow up (one month after surgery) also showed a reduction of cyst volume (7,8 cm³ before surgery and 6,4 cm³ after surgery, 17,9 % reduction of cyst volume). However, the patient's problems remained unchanged by the follow-up period 2 months after surgery. The patient then underwent an implantation of bilateral subthalamic DBS electrodes supported by intraoperative microrecording and macrostimulation. Clear subthalamic nucleus recordings were present on two (central and posterior) tracks of both sites out of four microelectrode trajectories. The central trajectories, with the tip of the electrode on the target or 1 mm below the targets, were selected for the definitive placement of DBS electrodes (Model 3389, Medtronic Inc, Minneapolis, MN, USA)(Fig. 4). The stimulator was implanted during the second stage of the surgery. After six years of stimulation, the patient significantly benefits from the procedure, as documented by her UPRDS III score in the off-medication state (52 points preoperative, 28 points after 3 years postoperative, and 34 point after 6 years postoperative). After six years, the patient's stimulation parameters are set at 3.4 V / 90 microsecond and 130 Hz. Medication (L-DOPA equivalent dose) is at 60% of the pre-DBS level.



Figure(4): Final position of the implanted electrodes

DISCUSSION

Three main issues were discussed when preparing the optimum treatment strategy for this individual patient:

- the relationship of clinical symptoms and cystic midline anomaly
- the indication for treatment of cystic lesion before DBS implantation
- the choice of appropriate surgical therapy

A search of the available literature did not find any paper suggesting a causal association between a midline cystic anomaly and parkinsonian symptoms. There was a frequent association between the cavum septi pellucidi and schizophrenia (30.4 %) and affective disorders (20 %) [2].

The main symptoms of midline cystic anomalies are intracranial hypertension, epileptic seizures and emotional instability [4]. A possible cause of the clinical symptoms associated with midline cysts is the compression of the interventricular foramen, the vascular structures, and the hypothalamoseptal triangle [3].

Only case reports or small clinical series of patients after neuroendoscopic surgeries for midline cystic anomalies have been reported. The main indication for neuroendoscopic cyst fenestration (cystoventriculostomy by fenestration of septal leaflet) is expansive behaviour of the cyst that may result in hydrocephalus caused by the obstruction or narrowing of the

interventricular foramen [5]. Meticulous attention should be paid to presurgical planning and ventricular entry to avoid injury to the fornices, thalamus, internal capsule, caudate nucleus, or the septal and thalamostriate veins. In a large surgical series of 10 patients (paediatric and adult cases) the main clinical symptoms were headaches, accompanied with dizziness, vomiting, and epileptic seizures; however, 2 of the patients presented with epileptic seizures only. Endoscopic septostomy (rigid endoscope via frontal approach) led to symptom relief and cyst size reduction in all patients, with no cyst recurrences [6].

From a group of 32 septostomies in 30 patients presented by Oertel, et al., 3 patients were operated on for septum pellucidum cyst. Long-standing cerebrospinal fluid pathway restoration was achieved by endoscopic septostomy [7].

Steerable endoscope was used for the treatment of a 6-year-old boy and a 42-year-old female presenting with headaches and syncopal episodes. Fenestration of the cyst walls allowed communication with both lateral ventricles and adhesions between the cyst wall and the interventricular foramen were lysed with endoscopic cautery [8]. Fenestration of both septal walls using ultrasound followed by cyst regression was reported in one patient, an 11-year-old boy [5].

Our indication for neuroendoscopic surgery was supported mainly by graphic findings suggesting expansive cyst behaviour (cyst walls bulging and foramen of Monro narrowing), because there were no clinical symptoms of intracranial hypertension. The manufacturer's policy contraindicates routine MRI follow up after DBS, therefore CT would be the only graphic modality for graphic cyst follow up. Moreover, artefacts from implanted electrodes may interfere with cyst volumetric analysis and anatomical study.

The literature data show that bilateral subthalamic nucleus deep brain stimulation has long-term beneficial effects in Parkinson's disease patients. A retrospective study performed by Kleiner-Fishman, et al., summarises the clinical evidence on the effectiveness of subthalamic nucleus deep brain stimulation (Medline and Ovid databases 1993-2004), with the identification of some prognostic factors for short and long term results [9]. Bilateral subthalamic nucleus stimulation is beneficial in the long term for Parkinson's disease motor symptoms, but does not prevent disease progression or the occurrence of axial L-DOPA unresponsive symptoms in some patients. There is a 50% improvement in the scales evaluating the quality of life in patients after deep brain stimulation [10] which corresponds with the final outcome of the reported patient.

CONCLUSION

Both neuroendoscopy and stereotactic neurosurgery are parts of the complex of minimally invasive neurosurgery. In this particular case of coincidence of an expansive midline cyst and Parkinson's disease, a combined treatment approach was utilised to maximise the functional outcome of the patient avoiding another surgery with DBS implant in situ.

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REFERENCES

- [1] Silbert P.L., Gubbay S.S. and Vaughan R.J. (1993) Cavum septum pellucidum and obstructive hydrocephalus. *J of Neurol Neurosurg Psychiatr* **56**: 820 - 822
- [2] Kwon J.S., Shenton M.E., Hirayasu Y., Salisbury D.F., Fischer I.A., Dickey C.C., Yurgelun-Todd D., Tohen M., Kikinis R., Jolesz F.A. and McCarley R.W. (1998). MRI study of cavum septi pellucidi in schizophrenia, affective disorder, and schizotypal personality disorder. *Am J Psychiatry* **155**: 509-15.
- [3] Fratzoglou M., Grunert P., Leite dos Santos A., Hwang P. and Fries G.(2003). Symptomatic cysts of the cavum septi pellucidi and cavum vergae: the role of endoscopic neurosurgery in the treatment of four consecutive cases. *Minim Invasive Neurosurg* **46**: 243-9.
- [4] Akiyama K., Sato M., Sora I., Otsuki S., Wake A., Fukui H., Takahashi Y., Yanagida K. and Sudara M.(1983). A study of incidence and symptoms in 71 patients with cavum septi pellucidi. *No To Shinkei* **35**: 575-81.
- [5] Weyerbrock A., Mainprize T., Rutka JT.(2006). Endoscopic fenestration of a symptomatic cavum septum pellucidum: technical case report. *Neurosurgery* **59**(Suppl 4): ONSE 491
- [6] Meng H., Feng H., Le F.and Lu JY.(2006). Neuroendoscopic management of symptomatic septum pellucidum cysts. *Neurosurgery* **59**: 278-83.
- [7] Oertel J.M., Schroeder H.W., Gaab M.R.(2009). Endoscopic stomy of the septum pellucidum: indications, technique, and results. *Neurosurgery* **64**: 482-91.
- [8] Lancon J.A., Haines D.E., Lewis A.I. and Parent A.D.(1999). Endoscopic treatment of symptomatic septum pellucidum cysts: with some preliminary observations on the ultrastructure of the cyst wall: two technical case reports. *Neurosurgery* **45**: 1251-7.
- [9] Kleiner-Fisman G., Herzog J., Fisman D.N., Tamma F., Lyons K.E., Pahwa R., Lang A.E. and Deuschl G.(2006). Subthalamic nucleus deep brain stimulation: summary and metaanalysis of outcomes. *Mov Disord* **21** (Suppl 14): S290-304.
- [10] Tarsy D., Vitek J.L., Starr P.A., Okun M.S.(eds), (2009), *DBS in Neurological and Psychiatric Disorders*, Humana Press, New York, pp, 3-32.

كيسة الحاجز الشفاف (Septum Pellucidum Cyst) في مريض يعاني من مرض الباركنسون (Parkinson's Disease) مراحل وطرق المعالجة

خالد غلاب، زدنك نوفاك، يان خراستينا . بلاج . ايفو ريهالك

- جمهورية التشيك- المستشفى التعليمي- مدينه برنو قسم جراحه المخ والاعصاب

E-mail: jan.chrastina@fnusa.cz, khaled.ghallab@fnusa.cz or drkhgh@hotmail.com.

ملخص

خلفية: الغرض من هذا التقرير لهذه الحالة المرضية هو تقديم العلاج المناسب والحديث لهذا المريض الذي كان بالإضافة للمشكلات الحركية التي سببها مرض باركنسون كان يعاني من وجود كيس كبير في الحاجز الشفاف. المواد والطريقة تمت على مرحلتين اقترح استراتيجية الجراحية من قبل فريق الاضطرابات الحركية بان تتم الجراحة على النحو التالي، يتم ثقب الكيس بواسطة التنظير الجراحي ومن ثم تزرع أقطاب كهربائية لاحقة للتخفيف الدماغي العميق الى نواة تحت المهاد اعتماد على تأثير معالجة التنظير.

النتائج: بعد النجاح التقني في ثقب الكيس وتوصيله بالنظام البطيني عن طريق التنظير الجراحي تم التأكد من صغر حجم الكيس واتصاله بالنظام البطيني اثنا العملية كذلك بعد العملية تم عمل اشعه مقطعيه حيث شوهد ان الاتصال بين الكيس و النظام البطين حر ، الأعراض الحركية لمرض باركنسن بقيت بدون تغيير، نظام التّخفيف الدماغي العميق لخلابا تحت المهادي التي تم زرعها من الجانبين عملت على التحسن الوظيفي للحركة بشكل جيد كما هي موقّعة من قبل يو بي آر دي إس الدرجة الثالثة في حالة خارج الدواء (52 نقطة ما قبل الجراحة و34 نقطة بعد 6 سنوات ما بعد الجراحة) و تخفيض جرعة L-DOPA الى (60% من مستواه قبل التّخفيف الدماغ العميق).

الخلاصة: العلاج الجراحي بواسطة التنظير الجراحي الدماغي لثقب الكيس الموجود في الحاجز الشفافي، والذي تلاها زرع التخفيف الدماغي العميق لتحت المهاد الثنائية - أدى إلى تحسن كبير ودائم من أعراض المريض.

الكلمات الدللية: كيسة الحاجز الشفاف، مرض باركنسون ، المخ، التنظير الدماغي.