

Ventricular subependymoma with intratumoral haemorrhage mimicking haemocephalus due to aneurysm rupture

Intraventrikulární subependymom s intratumorálním prokrvácením imitující hemocefalus při aneuryzmatickém subarachnoidálním krvácení

Dear editor,

Subependymoma is a tumour which originates from a subependymal cell plate. It most commonly occurs in the area of the fourth ventricle (50–60%), and less often in the lateral ventricles (30–40%). It can also occur in the septum pellucidum and the central spinal canal [1,2]. This tumour is benign and slow growing. The World Health Organisation classifies it as grade I [3]. Subependymomas are rare and can be an incidental finding during autopsies (0.4%). They make up approximately 0.7% of all resected symptomatic intracranial tumours. If the subependymoma is symptomatic, it usually manifests with symptoms of obstructive hydrocephalus or mass effect [4,5]. This tumour is often misdiagnosed as a glioma, ependymoma, meningioma, cavernoma, etc. [6]. We present a rare case of symptomatic lateral ventricle subependymoma, which was misdiagnosed as an intraventricular haematoma caused by a ruptured middle cerebral artery aneurysm.

A 71-year-old woman was admitted to our department with a sudden onset headache and meningeal signs. Her neurological condition was graded as Hunt Hess 1 and World Federation of Neurological Societies (WFNS) grade 1. Subarachnoid haemorrhage in the area of the right Sylvian fissure and haemocephalus in the left lateral ventricle were evident on the initial CT, therefore the finding was evaluated as Fisher grade 4 (Fig. 1A, B). The subsequent CTA showed a middle cerebral artery aneurysm on the right side, which was suitable for clipping. Surgery was performed on the same day without complications. The subsequent postoperative

course was without any adverse events. The patient was transferred to her local hospital on the 14th postoperative day. Three months later, the patient was referred to our department, due to a newly developed gait disorder. She could only walk with the aid of two crutches and her gait was unstable with tendencies to fall. She also suffered from memory impairment and incontinence. CT showed symmetrical dilatation of the ventricular system and a persisting hyperdense structure in the left lateral ventricle. Compared to the previous CT, the structure density remained identical. This excluded our previous assumption that it was an intraventricular blood clot (Fig. 1C). The patient was admitted for further examination. An MRI was performed, which showed an exophytic tumour arising from the area of the left thalamus with contrast enhancement. Foramen of Monro was not occluded, thus there were no signs of obstructive hydrocephalus (Fig. 2A). Consequently, a lumbar infusion test was performed with a positive result (resistance to outflow 15.3 mmHg/ml/min). Analysis of cerebrospinal fluid (CSF) showed a mild non-infectious inflammatory response. The patient preferred active treatment and endoscopic resection of the tumour was indicated; however, she was informed that the risk of subsequent ventriculoperitoneal shunt placement is high. Endoscopic resection was performed without complications with no residual tumour on the postoperative MRI (Fig. 2B). Histological analysis was suggestive of a subependymoma with inner haemorrhagic transformation (Fig. 2C). The patient was discharged on the 4th postoperative day. On subse-

The Editorial Board declares that the manuscript met the ICMJE "uniform requirements" for biomedical papers.

Redakční rada potvrzuje, že rukopis práce splnil ICMJE kritéria pro publikace zasílané do biomedicínských časopisů.

T. Radovnický, K. Pištěk, M. Sameš

Department of Neurosurgery,
J. E. Purkyně University, Masaryk Hospital
in Ústí nad Labem, Krajská zdravotní, a.s.,
Czech Republic



Tomáš Radovnický, MD, PhD
Department of Neurosurgery
J. E. Purkyně University
Masaryk Hospital
Sociální péče 3316/12A
403 40 Ústí nad Labem
Czech Republic
e-mail: tomas.radovnický@kzcr.eu

Přijato k recenzi: 8. 2. 2022

Přijato do tisku: 10. 3. 2022

quent postoperative follow-up examinations, her neurological status was getting better. The patient was able to walk without aids, did not fall and her memory and continence improved. After a 12-month follow-up period, she still does not require shunt implantation.

In our case report, we present a case of a subependymoma misdiagnosed as intraventricular haemorrhage caused by rupture of a middle cerebral artery aneurysm. The reason for this was that the tumour demon-

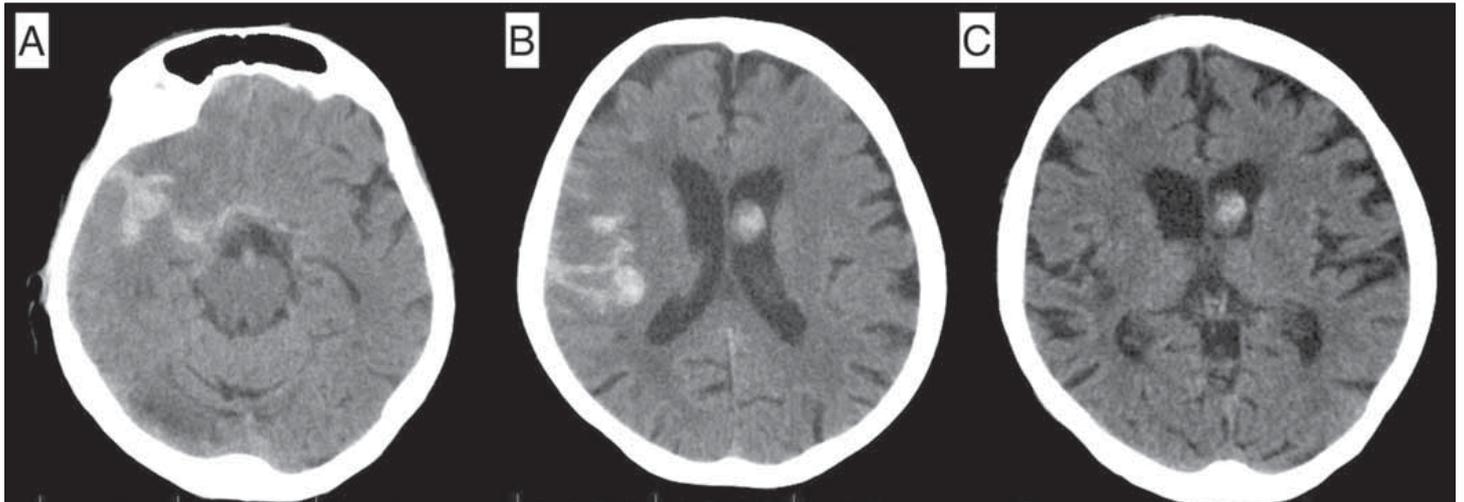


Fig. 1. Initial non-contrast brain CT (A) with subarachnoid haemorrhage dominantly in the right Sylvian fissure; (B) with a hyperdense formation in the left lateral ventricle and (C) after 3 months with a persistent hyperdensity in the left lateral ventricle.

Obr. 1. Vstupní nativní CT mozku s (A) subarachnoidálním krvácením, zejména v pravé Sylviově rýze; (B) s hyperdenzním útvarom v levé postranní komoře a (C) po 3 měsících s přetrvávající hyperdenzitou v levé postranní komoře.

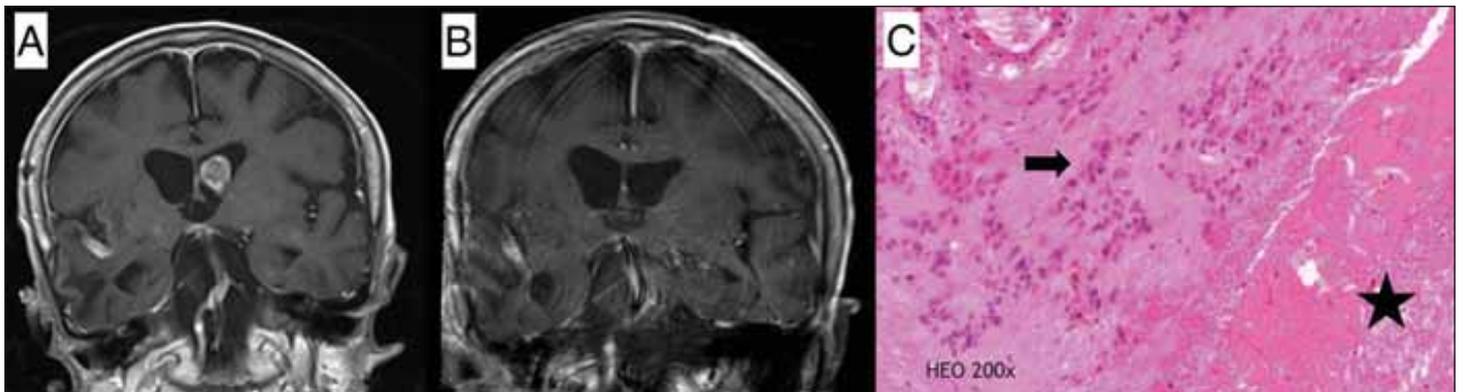


Fig. 2. (A) MRI (postcontrast T1 weighted image) with an exophytic enhancing tumour in the left lateral ventricle. (B) Postoperative MRI with no tumour remnant. (C) Histological examination (haematoxylin eosin dye). Arrow sign points at tumour, star sign at intratumoral haemorrhage.

Obr. 2. (A) MR (T1 vážený obraz, postkontrastní) s exofytickým tumorem v levé postranní komoře s postkontrastním syčením. (B) Pooperační MR bez viditelného rezidua tumoru. (C) Histologické vyšetření (barvení hematoxylin eosin). Šipka označuje tumor, hvězda pak intratumorální prokrvácení.

strated similar density as an intraventricular blood clot on the initial non-contrast CT. The hyperdense characteristic of these lesions is rare; most commonly they are isodense to the brain tissue [7]. Histological analysis revealed an intratumoral haemorrhage, which may be the reason for its high density. Intratumoral haemorrhage is relatively rare, with only a few cases described in the literature [8]. It can even represent the cause of intraventricular bleeding [9,10]. A special feature was identical tumour density on the initial and follow-up CT scans. Thus, haemorrhage within the tumour most likely occurred repeatedly. A follow-up CT was performed after 3 months for symptoms which corresponded to secondary normal pressure hydrocephalus (sNPH). On this examination, the

structure in the left lateral ventricle persisted with a constant density, which ruled out the diagnosis of an intraventricular blood clot. An MRI was indicated and revealed an exophytic tumour in the left lateral ventricle arising from the left thalamus. However, the question remained whether the tumour was merely an incidental finding or whether it was responsible for the symptoms. Subependymoma most often manifests with symptoms of obstructive hydrocephalus or mass effect [6]. In our case, the cerebrospinal pathways were without obstruction. The tumour was also not large enough to compress the periventricular tissue of the brain. Rather, the symptoms indicated post-haemorrhagic sNPH. For this reason, a lumbar infusion test with normal CSF basal pressure and high re-

sistance to outflow was performed, which confirmed our hypothesis. There were two options for further treatment. The first was implantation of a ventriculo-peritoneal (VP) shunt followed by tumour growth monitoring. The second was tumour resection with a high risk of subsequent VP shunt implantation. According to the patient's request, the second option was chosen – endoscopic resection of the tumour. This procedure had the advantage of obtaining a tumour sample for histological analysis.

Surprisingly, the symptoms of sNPH disappeared after the surgery and implantation of the VP shunt was not necessary. The reason for improvement is not clear. Obstructive hydrocephalus was not present on preoperative MRI, therefore a type

of sNPH must have been present. Our literature search did not reveal any similar cases. Analysis of CSF showed a non-inflammatory serous reaction; however, the interval from subarachnoid haemorrhage was 3 months, therefore it could not have been a consequence of this bleeding episode. A possible explanation is that it could have been a reaction to recurring intratumoral haemorrhage. This non-inflammatory reaction could be one possible pathophysiological mechanism of hydrocephalus development.

Acknowledgement

We thank Aeskulab Pathology Laboratory Prague for providing the histology images.

References

1. Varma A, Giraldo D, Mills S et al. Surgical management and long-term outcome of intracranial subependymoma. *Acta Neurochir (Wien)* 2018; 160(9): 1793–1799. doi: 10.1007/s00701-018-3570-4.
2. Nishio S, Morioka T, Mihara F et al. Subependymoma of the lateral ventricles. *Neurosurg Rev* 2000; 23(2): 98–103. doi: 10.1007/pl00021701.
3. Louis DN, Perry A, Reifenberger G et al. The 2016 World Health Organization Classification of tumors of the central nervous system: a summary. *Acta Neuropathol (Berl)* 2016; 131(6): 803–820. doi: 10.1007/s00401-016-1545-1.
4. Matsumura A, Ahyai A, Hori A et al. Intracerebral subependymomas. Clinical and neuropathological analyses with special reference to the possible existence of a less benign variant. *Acta Neurochir (Wien)* 1989; 96(1–2): 15–25. doi: 10.1007/BF01403490.
5. Rath TJ, Sundgren PC, Brahma B et al. Massive symptomatic subependymoma of the lateral ventricles: case report and review of the literature. *Neuroradiology* 2005; 47(3): 183–188. doi: 10.1007/s00234-005-1342-3.
6. Bi Z, Ren X, Zhang J et al. Clinical, radiological, and pathological features in 43 cases of intracranial subependymoma. *J Neurosurg* 2015; 122(1): 49–60. doi: 10.3171/2014.9.JNS14155.
7. Chiechi MV, Smirniotopoulos JG, Jones RV. Intracranial subependymomas: CT and MR imaging features in 24 cases. *AJR Am J Roentgenol* 1995; 165(5): 1245–1250. doi: 10.2214/ajr.165.5.7572512.
8. Zhang Q, Xie SN, Wang K et al. Intratumoral hemorrhage as an unusual manifestation of intracranial subependymoma. *World Neurosurg* 2018; 114: e647–e653. doi: 10.1016/j.wneu.2018.03.045.
9. Akamatsu Y, Utsunomiya A, Suzuki S et al. Subependymoma in the lateral ventricle manifesting as intraventricular hemorrhage. *Neurol Med Chir (Tokyo)* 2010; 50(11): 1020–1023. doi: 10.2176/nmc.50.1020.
10. Alserehi M, Turkistani F, Alghamdi F et al. Apoplexy of a collision tumour composed of subependymoma and cavernous-like malformation in the lateral ventricle: a case report. *Br J Neurosurg* 2019; 33(5): 581–583. doi: 10.1080/02688697.2017.1390063.

Impakt faktor časopisu Česká a slovenská neurologie a neurochirurgie pro rok 2020 činí **0,35**.