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Fourth ventricle epidermoid cyst – a case report

Torbiel naskórkowa czwartej komory mózgu – opis przypadku

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Abstract

An epidermoid cyst is a slowly growing, typically benign tumour, which requires surgical treatment. It usually develops in the cerebellopontine angle or parasellar region, and only occasionally appears in the ventricular system. It is unusual for an epidermoid cyst to place itself in the fourth ventricle. There have only been few reports on this subject published in the literature in the recent years, and therefore, we would like to present a comprehensive description of a fourth ventricle epidermoid cyst diagnosed in two patients in a short period of time. Optimal recovery of the patient, with a minimal risk of tumour recurrence, is best accomplished by total microscopic removal of the mass.

Keywords: epidermoid cyst, fourth ventricle, cholesteatoma

Streszczenie

Torbiel naskórkowa to wolno rosnący, zazwyczaj niezłośliwy nowotwór, który powinien być leczony chirurgicznie. Zwykle zlokalizowany jest w kącie mostowo-mózdkowym lub w okolicy siodła tureckiego, sporadycznie występuje wewnątrz układu komorowego. Torbiel naskórkowa usytuowana w czwartej komorze mózgu stanowi niecodzienny przypadek, z uwagi na dość rzadkie występowanie w tej okolicy. Ze względu na niewielką liczbę opisanych przypadków tytułowego guza w ostatnich latach przedstawiamy interdyscyplinarny opis torbieli naskórkowej czwartej komory mózgu rozpoznanej u dwóch pacjentek w krótkim odstępie czasu. Pełne wyleczenie chorego, przy najmniejszym ryzyku odrostu guza, uzyskać można przez całkowite mikroskopowe usunięcie zmiany.

Słowa kluczowe: torbiel naskórkowa, czwarta komora mózgu, perlak

INTRODUCTION

An epidermoid cyst is a slowly growing, rare and benign developmental tumour of an ectodermal origin. It is usually found in the central nervous system, although it can also appear in other organs, such as the middle ear or the orbit. Epidermoid cysts constitute 0.2–1% of intracranial tumours and are most often (60%) located in the cerebellopontine angle (7% of all tumours in this area) and in the parasellar region. Intraventricular location is unusual. Epidermoid cysts developing in this region constitute only 5–18.5% of all epidermoid cysts that appear in the cranial cavity (Bhatoe et al., 2006; Tancredi et al., 2003; Tytus and Pennybacker, 1956). As there is only a small number of reported cases, we would like to present an interdisciplinary description of two epidermoid cysts that were diagnosed and treated in a short period of time in the same centre.

CASE REPORT

First case

A 37-year-old female was referred for magnetic resonance imaging (MRI) due to problems with eye fixation and impaired balance. MRI revealed a well-defined cystic lesion in the fourth ventricle with a diameter of $4.2 \times 4.5 \times 5.4$ cm. The lesion had a cerebrospinal fluid (CSF) signal in both T1- and T2-weighted images. There was no contrast enhancement, but diffusion weighted imaging (DWI) showed a high signal, which suggested an epidermoid cyst (Fig. 1 A, B). The tumour caused a distortion and enlargement of the fourth ventricle with penetration through both Luscka foramina. On admission to our department, the patient experienced horizontal nystagmus, upper limb ataxia with pronator drift and left upper extremity weakness. Surgical correction included monitoring of acoustic evoked potentials (AEP) and bilateral

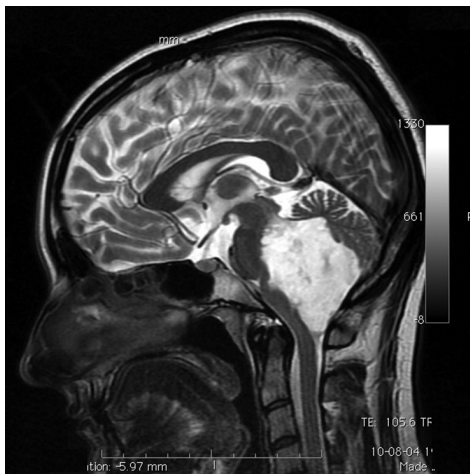


Fig. 1 A. Preoperative T2-weighted sagittal MRI of the first patient shows large lesion in the fourth ventricle

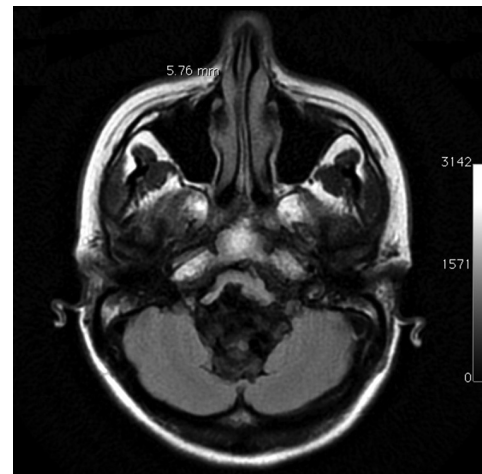


Fig. 1 B. Preoperative FLAIR axial MRI of the first patient shows heterogenous cystic lesion in the fourth ventricle

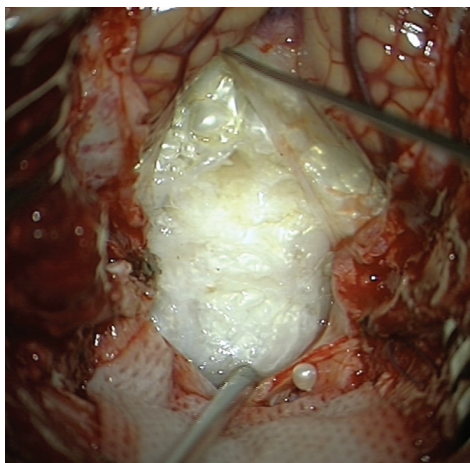


Fig. 2 A. Intraoperative view of the first pearly epidermoid cyst after opening its capsule

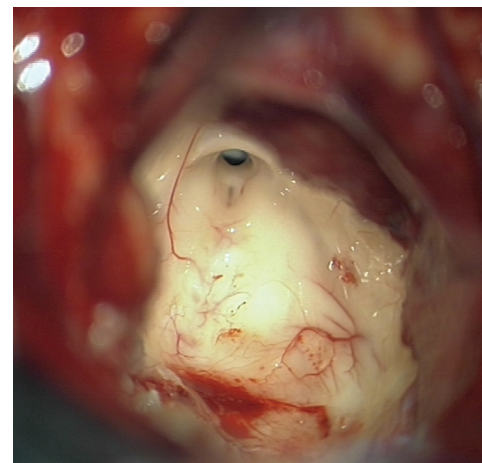


Fig. 2 B. Intraoperative view of the fourth ventricle and aqueduct after removal of the first tumour

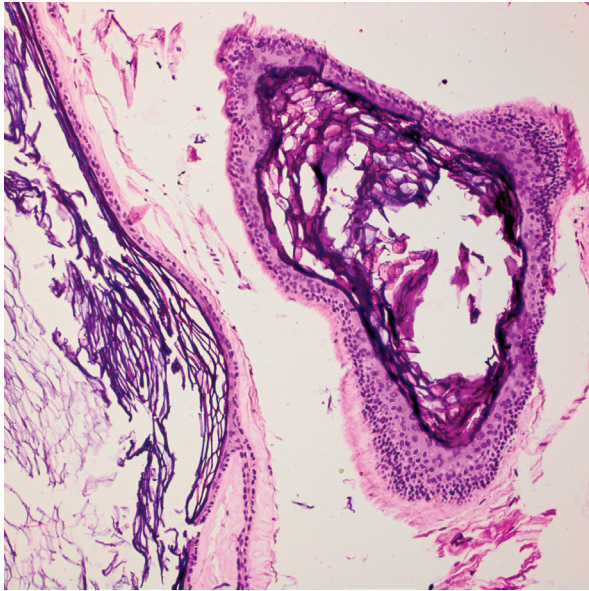


Fig. 3. Haematoxylin and eosin staining of an epidermoid cyst show epithelium and floating keratinized mass (the first case)

motor stimulation of V and VII cranial nerves. We proceeded with median suboccipital craniotomy. Pearly mass filling the great cistern was visible after opening the dura (Fig. 2 A). The tumour itself was easily removed with suction but the tumour capsule was strongly adherent to the floor of the fourth ventricle. It entered both Luscka foramina, penetrate to the cerebellopontine angles and displaced both vertebral arteries and posterior inferior cerebellar arteries. During separation of the cyst capsule from the floor of the fourth ventricle, increases in blood pressure up to 200 mm Hg were observed. The tumour was removed completely (Fig. 2 B). Pathological examination showed the presence of squamous epithelium and keratin masses inside the lesion, which confirmed the diagnosis of epidermoid cyst (Fig. 3). After surgery, we

observed worsening of imbalance, phonation problems, dysphagia and diplopia that resolved within a week. One month after the surgery, the patient recorded 100 points in the Karnofsky Performance Scale. MRI was performed 7 months after the surgery and revealed no residual tumour (Fig. 4 A, 4 B).

Second case

A 27-year-old female was referred to our department with an initial diagnosis of expansive arachnoid cyst, based on computed tomography (CT). On admission, the patient was neurologically intact, and she complained only about severe headaches for several years and suffered from refractory hypertension (systolic blood pressure up to 200 mm Hg). The MRI revealed a cystic lesion in the fourth ventricle measuring $2.7 \times 2.4 \times 3.0$ cm (Fig. 5 A, 5 B) that had a CSF signal in both T1- and T2-weighted images and no contrast enhancement. The cyst probably developed originally in the cerebellar vermis, causing enlargement of the fourth ventricle and compression of the medulla oblongata. Based on the DWI, an epidermoid cyst was suspected. The patient underwent surgery via median suboccipital craniotomy. After opening the dura, a pearly tumour was observed extending through the Magendie foramen (Fig. 6 A). First, the tumour was debulked and then the capsule was separated from the floor of the fourth ventricle, vermis and both cerebellar hemispheres. The tumour was removed completely (Fig. 6 B). Pathological examination revealed flattened squamous epithelium and keratin content and tumour capsule (Fig. 7), which confirmed the diagnosis of epidermoid cyst. After surgery, the patient experienced aseptic meningitis, which was effectively treated by the administration of corticosteroids. The postoperative CT showed no residual tumour (Fig. 8). The only symptom that the patient experienced for one month was vomiting.



Fig. 4 A. Postoperative T1-weighted contrast-enhanced sagittal MRI shows enlarged, empty fourth ventricle with no residual tumour (the first case)

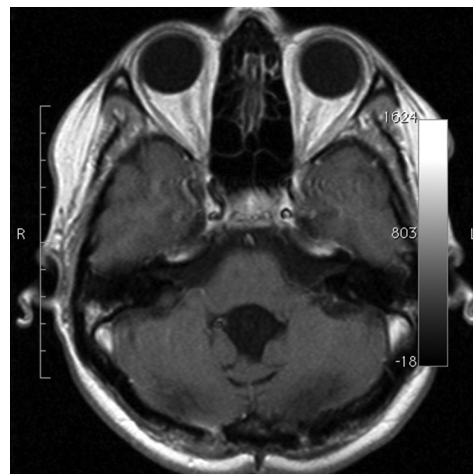


Fig. 4 B. Postoperative T1-weighted contrast-enhanced axial MRI shows enlarged, empty fourth ventricle with no residual tumour (the first case)



Fig. 5 A. Preoperative T2-weighted sagittal MRI of the second patient shows cystic lesion in the fourth ventricle

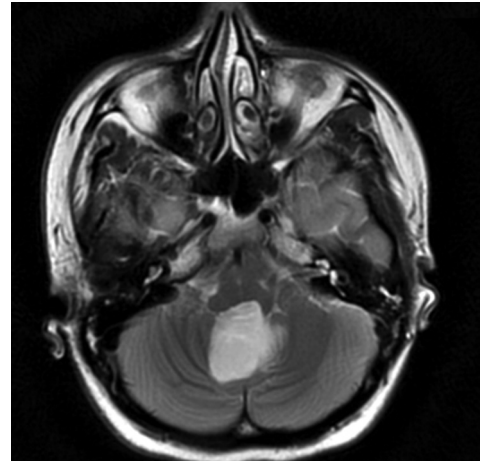


Fig. 5 B. Preoperative T2-weighted axial MRI of the second patient shows cystic lesion in the fourth ventricle

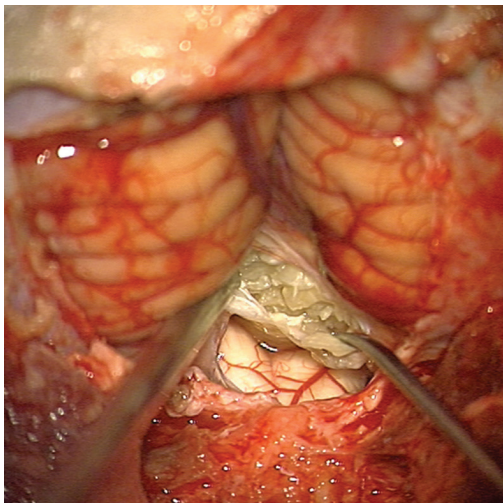


Fig. 6 A. An intraoperative view of the second tumour after opening its capsule

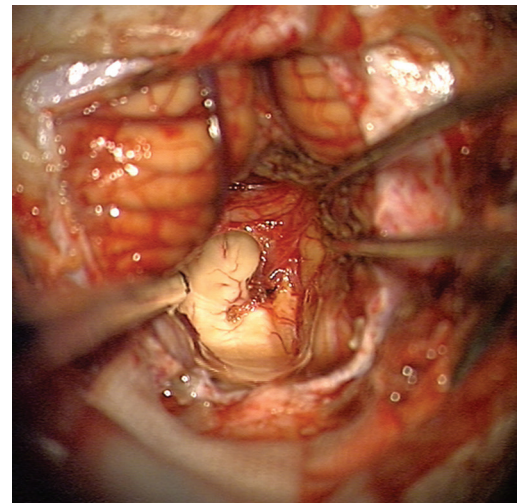


Fig. 6 B. An intraoperative view of the fourth ventricle after removal of the second tumour

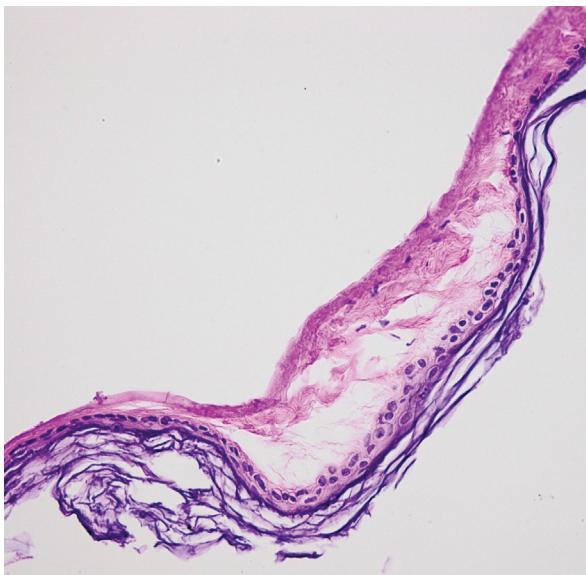


Fig. 7. Haematoxylin and eosin staining of the capsule of an epidermoid cyst (the second case)



Fig. 8. Postoperative CT scan shows no residual tumour and normal size and shape of the fourth ventricle (the second case)

DISCUSSION

Epidermoid cyst was described in 1807 by Pinson and in 1829 Cruveilhier, a French pathologist, introduced a term “pearly tumour” (fr. *tumeur perlée*), which depicted the cyst as “the most beautiful of all the tumours.” In 1836, Müller designated the tumour as a cholesteatoma, due to the presence of cholesterol deposits in it. A full pathological description of an epidermoid cyst was made by Bailey in 1920, and the first successful surgical removal of a tumour took place in 1865. The name “epidermoid cyst” was introduced by Critchley and Ferguson in 1928 in order to formally distinguish cholesteatoma of the middle ear from intracranial cholesteatoma, although both tumours are pathologically similar (Fox and South, 1965; Kuzeyli et al., 1996; Malorny and Wickboldt, 1987; Ziyal et al., 2005).

Epidermoid cysts can be congenital or acquired. The theory of the formation of an epidermoid cyst as a congenital anomaly was first reported by Remak in 1854 and has still been cited in the literature. According to this theory, an epidermoid cyst is formed between the 3rd and the 5th week of embryonic development due to improper closing of the neural tube. As for acquired epidermoid cysts, there are reports in the literature of iatrogenic and post-traumatic tumours, which resulted from epithelial cell implantation into the cranial cavity or spinal canal (Malorny and Wickboldt, 1987; Mermuys et al., 2008; Ziyal et al., 2005).

An epidermoid cyst is surrounded by a transparent and membranous capsule, which often fuses with neurovascular structures and the ventricular ependyma. The tumour has a milky surface, which resembles a shiny pearl. Inside the capsule, there is a basal membrane, on which the keratinising squamous cell epithelium grows into the tumour. The mass inside the tumour is brittle, soft and white, and has deposits of cholesterol and keratin fibres, although there are no sweat glands, sebaceous glands or hair outside the cyst, as in the case of dermoid cysts. Pericapsular calcification is visible in 10–25% of epidermoid cysts. An epidermoid cyst is described as a “mother-of-pearl-like,” producing “daughter pearls” (Fig. 2 A). The tumour is avascular and does not enhance after contrast administration in imaging studies. Sometimes, it is possible to observe sterile white milky fluid within the cyst, which can cause aseptic meningitis. The growth rate of the tumour is one cell generation per month, which corresponds to the growth rate of human skin (Forghani et al., 2007; Pepus et al., 1968; Ziyal et al., 2005).

Diagnostic imaging of brain tumours normally begins with a CT scan. In the case of an epidermoid cyst, CT may be inconclusive, because the tumour has a density similar to that of CSF, which may raise the possibility of the diagnosis of an arachnoid cyst. A preferred method for epidermoid cysts visualisation is MRI. In T1-weighted images, it is usually hypointense, while in T2-weighted images – hyperintense. A preferred diagnostic investigation is MRI in fluid light attenuation inversion recovery (FLAIR) and DWI.

In FLAIR images, the tumour is heterogeneous with a hyperintense centre, and in DWI it is hyperintense, which differentiates it from an arachnoid cyst. The differential diagnosis of epidermoid cysts located in the ventricular system should include intraventricular meningioma, ependymoma and choroid plexus papilloma (Çekiç et al., 2005; Franko et al., 2008; Mermuys et al., 2008; Ziyal et al., 2005).

The aim of a surgery is total resection of the tumour, because leaving a shred of a capsule may result in tumour recurrence. The beginning of removal of the tumour from its epithelial lodge is intracapsular, because it reduces the risk of penetration of the tumour content to the ventricular system (postoperative meningitis) and of damage to nervous structures due to stretching. In case of difficulty with total resection of the cyst, marsupialisation should be considered. The proportion of total cyst resection located in the fourth ventricle is under 30%. Recurrence of epidermoid or dermoid cyst within the posterior cranial fossa is between 0 and 54.5% (Akar et al., 2003; Pepus et al., 1968; Tancredi et al., 2003).

The most common complication of the surgery is aseptic meningitis, which is observed in almost 50% of cases, although a literature review by Chung et al. (2017) reported a 13.5% incidence. Other surgical complications are hydrocephalus, cranial nerve palsies and ventriculitis. Administration of steroids may effectively prevent inflammatory complications. Following incomplete resection, regrowth of the tumour should be considered as long-term undesirable situation – reported in the literature as occurring in up to 31% of patients. There are two significant prognostic factors: age and duration of symptoms. Patients with poorer outcomes were older and had longer duration of symptoms (Akar et al., 2003; Bhatoe et al., 2006; Chung et al., 2017).

Malignant transformation of an epidermoid cyst is a very rare phenomenon. The first researcher to describe a malignant cholesteatoma was Stromeyer (1910), as reported by Davidson and Small (1960), who also reported a malignant transformation of an epidermoid cyst. There are few reports of a benign epidermoid cyst developing into a squamous cell carcinoma (Akar et al., 2003). Fox and South (1965) described a case of a patient who developed a malignant cholesteatoma after the fourth regrowth of the tumour. Shah et al. (2010) presented a case of collision tumour, which consisted of an epidermoid cyst and a squamous cell carcinoma. Agarwal et al. (2007) reported a case of squamous cell carcinoma, which grew inside an epidermoid cyst. None of cysts presented in the above cited cases were located in the fourth ventricle.

CONCLUSION

A fourth ventricle epidermoid cyst is a rare lesion, which needs to be treated surgically, and has a high risk of postoperative aseptic meningitis. Optimal recovery of the patient, with a minimal risk of tumour recurrence, is best accomplished by total microscopic removal of the mass.

Conflict of interest

The authors declare that there is no conflict of interest.

References

- Agarwal S, Rishi A, Suri V et al.: Primary intracranial squamous cell carcinoma arising in an epidermoid cyst – a case report and review of literature. *Clin Neurol Neurosurg* 2007; 109: 888–891.
- Akar Z, Tanriover N, Tuzgen S et al.: Surgical treatment of intracranial epidermoid tumors. *Neurol Med Chir (Tokyo)* 2003; 43: 275–280; discussion 281.
- Bhatoe HS, Mukherji JD, Dutta V: Epidermoid tumour of the lateral ventricle. *Acta Neurochir (Wien)* 2006; 148: 339–342; discussion 342.
- Çekiç PY, Evlice AO, Sener RN: Epidermoid tumor in the fourth ventricle. *J Pediatr Neurol* 2005; 3: 195–196.
- Chung LK, Beckett JS, Ong V et al.: Predictors of outcomes in fourth ventricular epidermoid cysts: a case report and a review of literature. *World Neurosurg* 2017; 105: 689–696.
- Davidson SI, Small JM: Malignant change in an intracranial epidermoid. *J Neurol Neurosurg Psychiatry* 1960; 23: 176–178.
- Forghani R, Farb RI, Kiehl TR et al.: Fourth ventricle epidermoid tumor: radiologic, intraoperative, and pathologic findings. *Radiographics* 2007; 27: 1489–1494.
- Fox H, South EA: Squamous cell carcinoma developing in an intracranial epidermoid cyst (cholesteatoma). *J Neurol Neurosurg Psychiatry* 1965; 28: 276–281.
- Franko A, Holjar-Erlić I, Miletić D: Lateral ventricle epidermoid. *Radiol Oncol* 2008; 42: 66–68.
- Kuzeyli K, Duru S, Cakir E et al.: Epidermoid tumor of the occipital bone. *Neurosurg Rev* 1996; 19: 109–112.
- Malorny M, Wickboldt J: Epidermoid tumor of unusual expansion at the skull base. *Neurosurg Rev* 1987; 10: 299–303.
- Mermuys K, Wilms G, Demaerel P: Epidermoid cyst of the fourth ventricle: diffusion-weighted and FLAIR MR imaging findings. *JBR-BTR* 2008; 91: 58–60.
- Pepus M, Hutcheson JB, Tamayo JL: Pearly tumor of the fourth ventricle. *South Med J* 1968; 61: 487–490.
- Shah A, Goel A, Goel N: A case of cerebellopontine angle epidermoid tumor and brainstem squamous cell carcinoma presenting as collision tumor. *Acta Neurochir (Wien)* 2010; 152: 1087–1088.
- Tancredi A, Fiume D, Gazzeri G: Epidermoid cysts of the fourth ventricle: very long follow up in 9 cases and review of the literature. *Acta Neurochir (Wien)* 2003; 145: 905–911; discussion 910–911.
- Tytus SJ, Pennybacker J: Pearly tumours in relation to the central nervous system. *J Neurol Neurosurg Psychiatry* 1956; 19: 241–259.
- Ziyal IM, Bilginer B, Bozkurt G et al.: Epidermoid cyst of the brain stem symptomatic in childhood. *Childs Nerv Syst* 2005; 21: 1025–1029.