

EPIDERMOID CYST OF THE CRANIOVERTEBRAL JUNCTION – A CASE REPORT

**Tomasz Siwek¹, Łukasz Grabarczyk², Wojciech Kloc²,
Beata Szopa³, Ewa Iżycka-Świeszewska⁴, Beata Zwiernik¹,
Zbigniew Cebulski¹, Jacek Zwiernik⁵**

¹ Department of Neurology, University Hospital, University of Warmia and Mazury in Olsztyn, Poland

² Department of Neurology and Neurosurgery, Faculty of Medical Sciences, University of Warmia and Mazury in Olsztyn, Poland

³ Department of Neurosurgery, The Nicolaus Copernicus Regional Traumatology Center in Gdańsk, Poland

⁴ Chair and Division of Pathomorphology, Medical University of Gdańsk, Poland

⁵ Department of Neurology, Provincial Specialist Hospital in Olsztyn, Poland

ABSTRACT

Introduction. An epidermoid cyst (EC) is a congenital, benign, and noninfiltrating tumor which originates from the central nervous system (CNS) as a result of neural tube defects (NTD). The presence of lesion may cause CNS symptoms by means of mass-effect, when compressing the adjacent structures, or rupture and secondary aseptic inflammatory process. Within the CNS, EC is most frequently located in: pontocerebellar angle, suprasellar region, posterior cranial cavity, and spinal canal. The clinical symptoms can be general, like headache or dependent on the lesion location: dizziness, nystagmus, damage to the cranial nerves within the pontocerebellar angle and brain stem, disturbed fields of vision, and epileptic seizures. Magnetic resonance imaging (MRI) allows for visualizing the lesion which is shown as a heterogenic iso- or hypointense mass in relation to the cerebrospinal fluid. Using diffusion weighted images (DWI) is a helpful tool in differentiating tumor mass from cerebrospinal fluid. In computed tomography (CT) scans, EC has the same or almost the same density as the cerebrospinal fluid which makes the image difficult to differentiate from an arachnoid cyst. Treatment of choice is to remove the lesion in a possibly complete spectrum. In asymptomatic patients or patients exhibiting a mild intensity of symptoms, it is possible to wait and observe which solution could be chosen.

Corresponding address: Tomasz Siwek, Oddział Neurologii Szpitala Uniwersyteckiego z Poradnią SPZOZ UW-M w Olsztynie, ul. Warszawska 30, 10-082 Olsztyn, Poland; phone: +48 728 323 385, fax: +48 89 524 53 82, e-mail: tswk@wp.pl

Received 22.12.2010, accepted 7.02.2011

Aim. The aim of this paper was to present a case report concerning treatment of an EC located in the craniovertebral junction.

Materials and methods. A clinical management concerning a patient diagnosed and treated both in an outpatient clinic and in hospital at the departments of neurology and neurosurgery has been described.

Case study. A 69-year old female patient came to the Neurology Outpatient Clinic complaining of headaches, pain in her neck, and balance disturbances. These symptoms had intensified for 2 years. An MRI scan of the cervical spine revealed a large lesion in the posterior cranial cavity. An MRI scan of the head showed a multicystic tumor located in the posterior fossa in the projection of cerebellomedullary cistern, which did not undergo contrast intensification. The patient was treated surgically. Following surgery, headaches, pain in the neck as well as neurological symptoms subsided.

Results and discussion. Microsurgical removal of the lesion is the treatment of choice for EC. A total removal should be strived for, preferably en block, although leaving part of the cyst is not a mistake. Postoperative mortality may reach 8.9%, whereas recurrences amount to 4.5% during an 8-year long observation period. EC untreated surgically can, through dermal fistulas, cause bacterial meningitis. Thus, treatment limited to the observation of the lesions may pose a threat, and surgical treatment should be decided upon quickly.

Conclusions. Surgical treatment with the use of microscopic technique aided with neuromonitoring allows for removing tumors of the cholesteatoma type safely and radically.

Key words: epidermoid cyst (EC), brain tumor, craniovertebral junction

INTRODUCTION

An epidermoid cyst (EC) was first reported in 1807, when Pinson, an artist in the School of Medicine in Paris, made a wax model of a small lesion located in the cerebellopontine angle. In 1829, Cruveilhier, a French pathologist, first recognized EC as a specific entity and called it “cholesteatoma” [3].

EC is a result of ectodermal structures being misplaced and appearing within the neural tube when it is being formed in the 3rd–5th week of fetal development [10]. This lesion accounts for 0.2–1.8% of all intracranial tumors and less than 1% of intracanal tumors, with the incidence being similar in both sexes [10]. The first symptoms most often occur in the 4th decade of life. Frequently, however, such tumors are found during routine autopsies carried out due to other reasons [6].

Symptoms depend on tumor location, and involve: dizziness, nystagmus, features of damage to the cranial nerves (facial and vestibulocochlear), disturbed fields of vision, and epileptic seizures [2, 8, 9]. The symptoms usually progress slowly, but sudden and intermittent courses have been also reported [6]. Spontaneous, iatrogenic, or post-

traumatic damage to the cyst can lead to lymphocytic cerebrospinal meningitis, as a result of the cyst content penetrating into the subarachnoid space and inducing aseptic inflammatory reaction [8]. Imaging diagnostics involves computed tomography (CT) and magnetic resonance imaging (MRI). In a CT scan, EC is shown as isodense lesions in relation to the cerebrospinal fluid, which do not undergo contrast intensification. Consequently, the image is difficult to differentiate from that of an arachnoid cyst. MRI is recommended because of its being more significant in differential diagnosis of arachnoid cyst [2, 3]. MRI shows EC as a heterogenic hypointense lesion (similar to the intensity of the cerebrospinal fluid) on a T1-weighted image, which does not undergo contrast intensification, and is usually of a hyperintense signal on a T2-weighted image. Diffusion weighted imaging sequence in which EC is hyperintense can be helpful in differentiating the tumor from the cerebrospinal fluid surrounding it and from non-specific CNS lesions [1, 9]. In rare cases metrizamide cisternography is also used since it allows for differentiating a tumor capsule [3, 5].

Treatment of choice involves surgical removal of the lesion. It must be performed very carefully so that the content of the cyst does not penetrate into the subarachnoid space and brain tissue [3]. In asymptomatic patients or patients with single, stable symptoms, observation is recommended in order to detect the appearance or development of clinical symptoms, and their possible intensification, before deciding on surgical treatment [4]. This paper describes the rare location of a tumor in the craniovertebral junction, which is pressing against the brain stem and cerebellum.

AIM

The aim of this paper is to present a case report concerning treatment of an EC located in the craniovertebral junction.

MATERIALS AND METHODS

A clinical management concerning a patient diagnosed and treated both in an outpatient clinic and in hospital at the departments of neurology and neurosurgery has been described.

CASE STUDY

A 69-year old female patient came to the Neurology Outpatient Clinic complaining of headaches, pain in her neck, and balance disturbances. X-ray of the cervical spine revealed degenerative changes. Neurological examination confirmed a stiff neck, disturbances in alternating movement, upper limbs dysmetria, positive Romberg's sign with closed eyes.

An MRI scan of the cervical spine revealed a large lesion in the posterior cranial cavity (Fig. 1, 2), whereas an MRI scan of the head showed a multicystic tumor, measuring $39 \times 30 \times 24$ mm, located in the posterior fossa in the projection of cerebellomedullary cistern. Its signal was similar to that of the cerebrospinal fluid and did not undergo contrast intensification. The tumor compressed the medulla and the initial section of the

cervical spinal cord, and pressed them against the dens of the epistropheus. From the bottom, it also pressed against cerebellar hemispheres, especially the left one. Features of infiltration and brainstem or medulla oblongata edema were not detected (Fig. 3, 4).

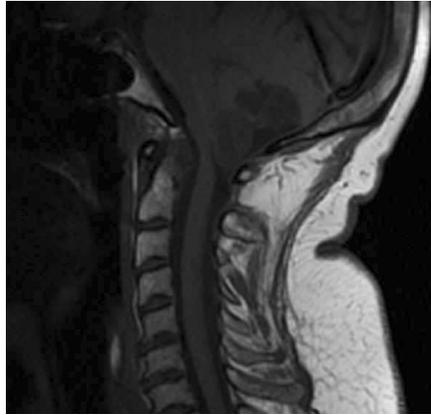


Fig. 1. Sagittal T1-weighted MRI scan of the cervical spine showing a cyst at the craniovertebral junction

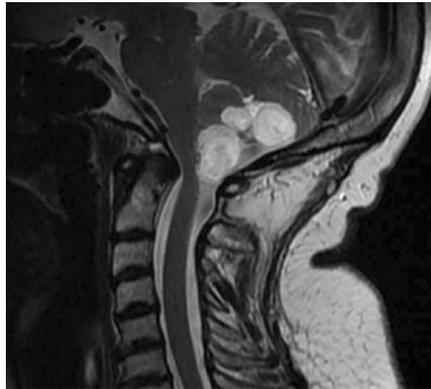


Fig. 2. Sagittal T2-weighted MRI scan of the cervical spine showing a cyst at the craniovertebral junction

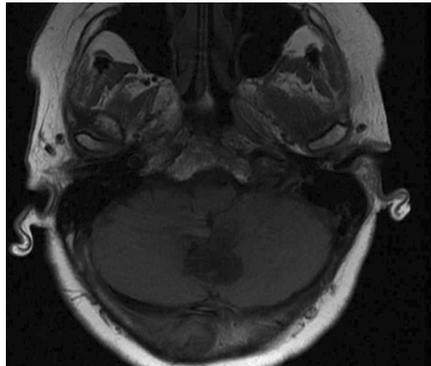


Fig. 3. Axial T1-weighted MRI scan of the head showing a cyst

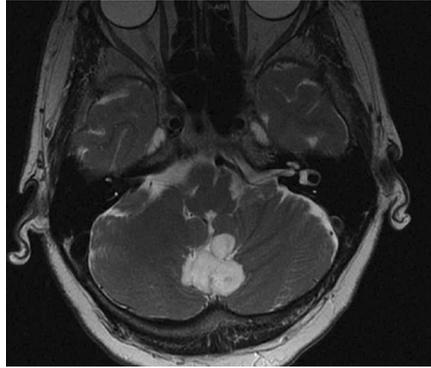


Fig. 4. Axial T2-weighted MRI scan of the head showing a cyst

The patient was qualified for surgical treatment. Suboccipital craniectomy was performed in the sitting position, with the use of neuronavigation and with the aid of intraoperative monitoring of: somatosensory evoked potentials and motor evoked potentials of the pyramidal tracts, cranial nerve nuclei, and auditory evoked potentials. No complications were observed postoperatively. In a follow-up CT scan of the head no features of the tumor were detected (Fig. 5). The histopathological test detected *cholesteatoma* (Fig. 6). The patient was discharged on the 7th day following surgery, in an improved condition. During a control examination performed 1 month following surgery, regression of all the previous symptoms: headaches, pain in the neck and neurological symptoms, was observed.

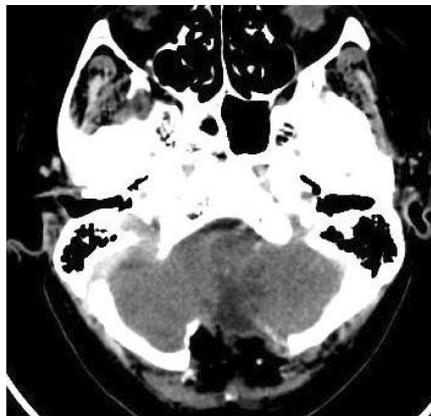


Fig. 5. CT scan of the head after the operation, 1st day, showing no features of a cyst

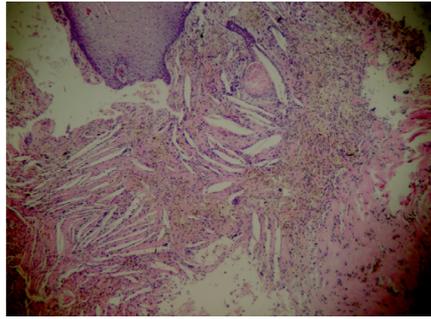


Fig. 6. Microscopic image of *cholesteatoma* (EC) – wall lined with keratinized stratified squamous epithelium, in the center keratinized mass, cholesterol crystals and elements of granulation tissue [HE, magn. 40×]

RESULTS AND DISCUSSION

EC are generally found extracerebrally and show a strong tendency toward a cranial base location [1]. They are most frequently observed in the regions of the craniovertebral junction and hypophysis [1, 2, 4, 6–8, 10]. The cyst's wall is lined with stratified squamous epithelium embedded in collagen, whereas the content consists of desquamated epithelium rich in cholesterol crystals, and macroscopically looking like a pearl mass (hence EC is called *cholesteatoma*) [1, 3]. In 10% of intracranial epidermal cysts, calcifications are observed resulting from the saponification of the cyst content [1].

Tumors show a tendency for focal hyperplasia and require surgical treatment. Microsurgical removal of the lesion is the treatment of choice for EC. A total removal should be strived for, preferably en block, although leaving part of the cyst is not a mistake [3, 5, 7]. Postoperative mortality may reach 8.9%, whereas recurrences amount to 4.5% during an 8-year long observation period [3]. Aseptic meningitis is detected in approximately 20% of patients. EC untreated surgically can, through dermal fistulas, cause bacterial meningitis. Thus, treatment limited to the observation of the lesions may pose a threat, and surgical treatment should be decided upon quickly [4, 7, 10].

CONCLUSIONS

With respect to EC, modern neurosurgical techniques, such as microsurgical techniques aided with a surgical microscope and intraoperative neuromonitoring, enable physicians to remove large lesions of adverse locations safely, irrespective of the patient's age.

REFERENCES

1. Aribandi M., Wilson N. J.: *CT and MR imaging features of intracerebral epidermoid – a rare lesion*. Br. J. Radiol., 2008; 81: 97–99.
2. Chih Kai C., Ham-Min T., Yi-Ho Y.: *Clinical presentation of posterior fossa epidermoid cysts*. Euro. Arch. Otorhino-laryngol., 2006; 263 (6): 548–551.
3. Imieliński B. L., Kloc W.: *Śródczaszkowe torbiele naskórkowe (perlaki prawdziwe, epidermoidy) [Intracranial epidermal cysts (true cholesteatomas, epidermoids)]*. Neurol. Neurochir. Pol., 1993; 27 (6): 859–867.
4. Lopes M., Capelle L., Duffau H., Kujas M., Sichez J. P., van Effenterre R., Failott T., Bitar A., Fohanno D.: *Prise en charge chirurgicale des kystes epidermoides intracraniens. A propos de 44 patients et revue de la litterature [Surgery of intracranial epidermoid cysts. Report of 44 patients and review of the literature]*. Neurochirurgie, 2002; 48 (1): 5–13.
5. Lunardi P., Fortuna A., Cantore G., Missori P.: *Long-term evaluation of asymptomatic patients operated on for intracranial epidermoid cysts. Comparison of the diagnostic value of magnetic resonance imaging and computer-assisted cisternography for detection of cholesterol fragments*. Acta Neurochir., 1994; 128 (1–4): 122–125.
6. Lunardi P., Missori P., Gagliardi F. M., Fortuna A.: *Dermatoid and epidermoid cysts of the midline in the posterior cranial fossa*. Neurosurg. Rev., 2009; 15 (3): 171–175.
7. Mohanty A., Venkatrama S. K., Rao B. R., Chandramouli B. A., Jayakumar P. N., Das B. S.: *Experience with cerebellopontine angle epidermoids*. Neurosurgery, 1997; 40 (1): 24–30.
8. Muzammil S., Leong K. S.: *A case of cerebellopontine angle epidermoid cyst presenting as trigeminal neuropathy*. South. Med. J., 2009; 102 (5): 534–536.
9. Ozgen T., Oge H. K., Erbeni A., Bertan V.: *Cranial dermoid and epidermoid cysts*. Neurochirurgia, 1990; 33 (1): 16–19.
10. Sundaram C., Paul T. R., Raju B. V., Ramakrishna Murthy T., Sinha A. K., Prasad V. S., Purohit A. K.: *Cysts of the central nervous system: a clinicopathologic study of 145 cases*. Neurol. India, 2001; 49 (3): 237–242.