Cerebellopontine angle epidermoid cyst – case report

Torbiel naskórkowa w kącie mostowo-móżdżkowym – opis przypadku

Abstract:
We report a case of 31 year old man, with an epidermoid tumor of the right cerebellopontine angle (CPA). He has 1.5 year history of headaches. Non cranial nerves deficit was presented. Non hearing loss reported. Cerebellopontine angle epidermoid cyst is the third most frequent lesion after acustic neuromas, and meningiomas. Due to the size and the localization of the tumor in internal auditory canal, the middle cranial fossa approach was used to remove the tumor. The hearing and the facial nerve were preserved.

Keywords:
headache, cerebellopontine angle, epidermoid tumor

INTRODUCTION
Cerebellopontine angle tumors comprise about 10% of all intracranial tumors. Most common are vestibular schwannomas which account for ca. 90% of all tumors within the region. The second most common group of tumors are meningiomas diagnosed in 5-10% of cases, while the third group are epidermoid cysts accounting for 0.2 to 1.8% of tumors (1-4).

Epidermoid cysts develop as the result of developmental disturbances due to improper location of the ectoderm (1). The lesions develop between the third and the fifth week of gestation (2, 3). They are encountered at all ages. However, most diagnoses are made between the third and the fifth decade of life (3, 4). Epidermoid cysts are slow-growing, benign tumors, most often manifesting by trigeminal neuralgia. Unilateral tinnitus is equally common while unilateral hearing loss is observed less frequently. Other symptoms include deficits of cranial nerves V and VII manifested by hypoesthesia and muscle weakness (1,2,5). Rare symptoms include cerebellar disorders, diplopia and hydrocephalus (1-2). Cases of epidermoid cysts becoming malignant are very rare (6). Epidermoid cysts within the cerebellopontine angle are diagnosed on the basis of computed tomography or magnetic resonance imaging scans (5).

Case report
A 31-year-old male patient had been diagnosed at his site of residence for 1.5 year due to periodic, intense, dull headaches. The patient experienced occasional vertigo, mostly upon adopting
vertical position. As the tinnitus experienced as “ringing” was not intense or troublesome, the patient did not report it to his physician upon consecutive visits. He did not complain of any other hearing disturbances either. Six months earlier, the patient was diagnosed with chronic sinusitis treated by nasal steroids. The headache persisted despite initiation of treatment. Due to this fact, a magnetic resonance scan was performed. The study revealed segmental thickening of the right vestibulocochlear nerve, limited to the inner auditory canal. The size of the thickening was 9x7x7.5mm (Fig. 1). The lesion displayed a hyperintense signal in the T2-weighted image (Fig. 2.) and hypointense signal in T1-weighted image. Patient was referred for surgical treatment at the Clinic of Otolaryngology of the Medical University of Warsaw.

Preoperative examination revealed no cranial nerve deficits. No hearing disorders, deviations in pure tone audiometric examination or abnormalities in impedance audiometry were observed (Fig. 3, Fig. 4.). After preparation, patient was subjected to surgical removal of the tumor from the middle cranial fossa approach. The incision was performed in front of the earlobe to facilitate exposure of the squamous part of temporal bone, zygomatic arch and the superior segment of the outer auditory canal. Temporal craniotomy sized 2.5 x 3 cm was performed using a freise. The bone fragment was removed and the meninx of the middle cranial fossa was stripped so as to reveal the upper surface of petrous pyramid. Superior semicircular canal was located using a diamond fraise and the inner auditory canal was opened from above moving down towards the cerebellopontine angle (Fig. 5.). The meninx located beneath the superior petrosal sinus was opened initially followed by the meninx of the inner auditory canal.

The facial nerve stretched across the frontal surface of the tumor. Encysted tumor was filled with epidermal masses. The entire lesion was excised and sent for histopathological examination. Hemosstasis was verified within the cerebellopontine angle. Next, skull base defect plasty was performed using a fragment of the temporalis muscle and fascia.

Patient’s toleration of the surgery was good. Vertigo persisted on days 1 and 2 after the surgery, gradually decreasing on the following days. The patient denied experiencing tinnitus, impairment of hearing or disturbed recognition of speech. Due to the mild facial nerve paresis (House-Brackmann grade II), patient was referred for consultation with physiotherapist. Patient was discharged from the Clinic on the seventh day after the surgery in good general and local condition with recommendation of regular laryngological follow up.

DISCUSSION

Epidermoid cysts are formed as the result of abnormal organogenesis between the third and the fifth week of fetal development. They contain keratin, epidermal calls and cholesterol crystals (5,7). The wall of the cyst consists of normal epidermal cells (3). The most common location of epidermoid cysts include cerebellopontine angle, suprasellar region and third ventricle region (3-5,7).

The complaints reported by the patients are due to the cyst being located in the vicinity of cranial nerves. most common complaints include headaches, trigeminal neuralgia, tinnitus, and hearing dis-
Translabyrinthine approach is used for removal of large cerebellopontine angle cysts in patients with deep preoperative hearing loss. This approach allows for good control of internal structures, particularly the facial nerve. Proper surgical technique facilitates anatomical conservation of facial nerve in as much as 95% of cases (9). The disadvantage of the approach consists of the necessity of destroying the inner ear and a relatively large risk of cerebrospinal fluid leakage (10). Samii et al. (14) reported CSF leakage in 21% of all cases in their surgical material.

The middle cranial fossa permits conservation of hearing and is characterized by low number of neurological complications including facial nerve paresis (11). It is useful in surgeries of small tumors and complete conservation of hearing depends on the size of the lesion (10,12).

The objective of the surgery is to completely remove the tumor including the capsule while maintaining the function of adjacent vascular and neural structures. As shown by the analysis of extensive study material performed by Scheich et al. (12), CSF leakage was observed in 12% of surgeries performed from the middle cranial fossa approach. At the same time, CSF leakage was the most common symptom observed in the study population and usually resolved following conservative treatment.

Although epidermoid cysts are benign tumors, Link et al. (6) reported a small percentage of malignant transformations into squamous cell carcinoma. The pathogenesis of this transformation is unknown.

Fig. 3. Pure tone audiometry result before the procedure.

Fig. 4. Speech audiometry result before the procedure.
SUMMARY

Epidermoid cysts are tumors rarely found within the central nervous system. Trigeminal neuralgia, unilateral tinnitus, unilateral sensory hearing impairment and refractory headaches are indications for detailed diagnostics. The treatment consists of surgical removal of the cyst. The surgical approach depends on the size and the location of tumor, patient’s age as well as the experience of the operating surgeon. Small epidermoid cysts located within the outer auditory canal may be operated from middle cranial fossa approach with conservation of the function of adjoining structures.

Fig. 5. Removal of an epidermoid cyst using the middle cranial fossa approach.

References


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