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Epidermoid cysts of the cerebellopontine angle: Clinical features and treatment outcomes

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ABSTRACT

Objective: To report clinical characteristics, treatment outcomes and risk of recurrence in patients with surgically treated cerebellopontine angle epidermoids.
Methods: In 1994–2013, we operated 17 patients, including 7 with tumor limited to the cerebellopontine angle, 7 with cerebellopontine angle tumor penetrating supratentorially, and 3 with cerebellopontine angle tumor extending along skull base to contralateral cerebellopontine angle. All patients were followed-up for the mean duration of 126 months.
Results: On admission cranial nerve symptoms predominated. Total tumor removal was achieved in 5 patients, and incomplete removal (with small tumor remnants left on vessels, nerves, or brainstem) in 12 patients. Postoperatively, preoperative deficits worsened in 2 and new postoperative deficits occurred in 10 patients. The extent of tumor expansion had no effect on postoperative morbidity and risk of recurrence. During long-term follow-up, improvement or resolution of preoperative deficits was seen in 11 of 17 patients, and new postoperative deficits in 8 of 10 patients. Symptomatic recurrences after an average of more than 9 years were noted in 5 patients, 3 of whom were reoperated. Recurrences occurred in some younger patients and always in area of primary tumor. No effect of extent of tumor removal on risk of recurrence was found.
Conclusions: The extent of tumor removal had no effect on the risk of recurrence, and thus it may be acceptable to leave tumor capsule fragments adhering closely to nerves, vessels, or brainstem. During long-term follow-up, resolution or improvement of present preoperatively and new postoperative neurological deficits may be expected in most patients.

1. Introduction

Epidermoid cysts are benign, usually congenital brain tumors, that are formed between 3 and 5 weeks of fetal life due to abnormal trapping of ectodermal cells, later developing into the epidermis, within the nervous tissue during neural tube closure. During the same period of embryogenesis, the otic and optic vesicles are also being formed, and inclusion of ectodermal cells within these structures results in the most

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common intracranial location of epidermoids, i.e. in the cerebellopontine angle and in the parasellar region [1,2]. It is suspected, that cysts located within the fourth ventricle result from ectodermal cell misplacement before neural tube closure, and cysts located epidurally or within the diploe are formed after neural tube closure [1]. The incidence of epidermoid cysts has been estimated at about 1% of all brain tumors [1,3]. Nearly half of them occur in the cerebellopontine angle and they are the third most common brain tumors in this location after schwannomas and meningiomas, comprising 5–7% of tumors in this region [1–4].

Epidermoid cysts are lined with stratified squamous epithelium overlying a connective tissue lamina that closely adheres to the pia mater and is anatomically indistinguishable from the latter. These tumors, being located in the subarachnoid space, when they grow and spread, fill subarachnoid cisterns, encompassing the nerves and vessels, much less frequently displacing them. Only after the all available spaces like cisterns, fissures, ventricles are filled, a space-occupying lesion effect ensues. Tumor growth results mostly from desquamation of epithelial cells inside the cyst, forming pearly, shiny debris, and also cholesterol and keratin secretion into the cyst [1,2,4,5]. These tumors grow very slowly, and hence the duration of symptoms often spans many years [3,4,6,7].

In computed tomography (CT), epidermoid cysts are difficult to distinguish from normal spaces with cerebrospinal fluid, and their presence may be suspected based on a finding of an abnormal space-occupying non-enhancing mass with signal density slightly higher compared to that of the cerebrospinal fluid [1–5]. Identification of calcifications within the tumor, which are present in 10–25% of cases, may be helpful in diagnosis [1–3,5]. Hyperdense epidermoid cysts are seen in 3% of cases, related to previous intracystic hemorrhage, high protein content, or saponification of keratinized debris to calcium soaps [1]. However, the imaging method of choice is magnetic resonance imaging (MRI) with diffusion-weighted imaging (DWI) sequence [2,4,8,9]. In most cases, epidermoid cysts are hypointense in T1-weighted images, with a signal intensity intermediate between the cerebrospinal fluid and the brain, and are markedly hyperintense in T2-weighted images, iso- or slightly hyperintense relative to the cerebrospinal fluid signal [3–6,8–10]. In FLAIR sequence, their signal does not attenuate completely, unlike the cerebrospinal fluid signal. DWI is most important for the diagnosis and differentiation from other lesions, mostly arachnoid cysts, as the content of the epidermoid cysts show prominent diffusion restriction (i.e. they are markedly hyperintense in DWI) due to layered microstructure of the debris [2,4,6,8]. The CISS sequence is also useful, clearly showing nerves and vessels within the tumor [1]. Previously heavy-T2-weighted images were used for this purpose [2,9]. Minimal contrast enhancement within the cyst capsule is seen in one fourth of epidermoids [1,9].

2. Material and methods

Between 1994 and 2013, 17 patients were operated due to an epidermoid cyst of the cerebellopontine angle, including 11 women and 6 men. In all cases, preoperative brain MRI was performed, in the recent years also using the DWI sequence. Tumor size was assessed based on their largest dimension in MRI, which ranged from 30 to 83 mm (mean 54.3 mm, median 50 mm). Based on MRI and intraoperative findings, cerebellopontine angle epidermoids were divided into three groups: tumors limited to the cerebellopontine angle ($n = 7$) (Fig. 1), tumors located in the cerebellopontine angle and extending supratentorially ($n = 7$) (Fig. 2), tumors located in the cerebellopontine angle and extending along the skull base to the contralateral cerebellopontine angle ($n = 3$) (Fig. 3). Clinical condition of the patients before the surgery and at discharge was evaluated based on the presence of neurological deficits, whereas long-term outcomes were evaluated based on both: the presence of neurological deficits and the modified Rankin scale [11]. As some patients, mostly those operated many years ago, did not undergo early postoperative MRI, evaluation of the completeness of tumor removal was based mostly on the assessment by the operating surgeon. Long-term follow-up data were obtained in all patients. Duration of follow-up ranged from 16 to 184 months, with mean 126 months. A long-term follow-up MRI was performed in all patients. Recurrence was defined as meeting both conditions: tumor recurrence or progression of residual tumor in neuroimaging studies and at the same time exacerbation of preexisting or occurrence of the new neurological deficits.

The aim of the study was evaluation of treatment outcomes in patients with cerebellopontine angle epidermoids, taking into account preoperative patient's condition and evolution of new postoperative neurological deficits, and evaluation of the risk of recurrence.

Statistical analysis was performed using the Statistica, ver. 12.0 (StatSoft) including basic descriptive statistics, the Student t-test and contingency tables with the Pearson Chi-square test and the exact Fisher test. $p \leq 0.05$ was considered statistically significant.

3. Results

3.1. *Patient symptoms*

Patient age on admission ranged from 17 to 60 years, with mean 39 years. Duration of symptoms ranged from 50 days to 11 years, with mean 3 years, and was longer than one year in half of patients. All operated tumor were symptomatic, and vertigo was the most common initial symptom (Table 1).

On admission, cranial nerve symptoms predominated, including hearing impairment ($n = 10$), vertigo ($n = 8$), facial nerve paresis ($n = 7$), lower cranial nerve paresis ($n = 7$). Table 3 shows all symptoms and signs present on admission along with their changes at discharge from hospital and during long-term follow-up.

3.2. *Surgical treatment*

The goal of surgical treatment was complete removal of the tumor with its capsule. However, if it was difficult to separate the capsule in places where it adhered closely to nerves, vessels or the brainstem, small fragments of the capsule were left to avoid the risk of serious neurological deficits due to