Surgical Resection of Cerebellopontine Epidermoid Cysts: Limitations and Outcome

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Abstract

Objective Epidermoid cysts are benign slowly growing tumors commonly involving the cerebellopontine angle (CPA). The aim of this study was to analyze the surgical limitations, surgical strategies, complications, and outcome of resection of these lesions.

Material and Methods The clinical data and outcome of 32 cases operated for CPA epidermoid between 2007 and 2015 were retrospectively analyzed. The mean follow-up period was 42.6 months, and all patients were followed up at least for a whole year.

Results There were 15 males and 17 females. The median age was 37.6 years. Headache and cranial nerves dysfunction were the most common presenting symptoms. Surgery was performed in all patients using the standard lateral suboccipital retrosigmoid approach. In three cases, microvascular decompression of an arterial loop was performed in addition to tumor excision. Total resection was accomplished in 19 out of 32 cases (59.4%), subtotal resection in 7 cases (21.9%), and only partial excision was achieved in 6 cases (18.7%). There was no recurrence or regrowth of residual tumor during the follow-up period. We had a single postoperative mortality due to postoperative pneumonia and septic shock. New cranial nerves deficits occurred in 15.6% of cases but were transient in most of them.

Conclusion The favorable outcome of total resection of CPA epidermoids should always be weighed against the critical risks that accompany it especially in the presence of tight adhesions to vital neurovascular structures. The retrosigmoid approach is suitable for the resection of these tumors even if they were large in size.

Keywords

- cerebellopontine
- epidermoid cyst
- retrosigmoid approach
- outcome

Introduction

Epidermoid cysts are benign slowly growing extra-axial tumors that represent 0.2 to 1.8% of all intracranial tumors. Although they belong to the spectrum of “tumors of disordered embryogenesis,” however they manifest late in life.^{1–3} Nearly half of these cysts occur in the cerebellopontine angle (CPA) as they represent 5 to 7% of tumors in this region following only schwannomas and meningiomas in frequency. CPA epidermoids are known to have tight adhesions with multiple cranial nerves and vascular struc-

tures, making their total surgical excision a challenging task.^{3–6}

In this study, we summarize our experience in the surgical management of CPA epidermoid cysts with special emphasis upon the choice of the approach and the surgical outcome.

Patients and Methods

This study is a retrospective analysis of 32 consecutive cases surgically treated for CPA epidermoid at our institute

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between 2007 and 2015. The initial diagnosis was based on computed tomography (CT) and magnetic resonance imaging (MRI) appearance and was histologically verified in all cases. The main goal of surgery was complete tumor excision without compromising any neurovascular structures. This was attempted by the sequential opening of all involved arachnoid cisterns to achieve total resection whenever feasible. If parts of the tumor tightly adhered to important neurovascular structures, they were deliberately left behind.

Intraoperative neurophysiological monitoring was lacking in all cases due to economic reasons. Special care was taken to avoid any seeding of the irritant contents into the subarachnoid space. High-dose steroids were used both intraoperatively and for 1 week postoperatively to reduce the risk of aseptic meningitis. The extent of resection was evaluated based on intraoperative assessment and postoperative images. All patients included in this study were followed up postoperatively both clinically and radiologically at least for a whole year. The postoperative hearing assessment was based mainly on the subjective reporting of the patients. Pre- and postoperative audiometries were available in only three cases. The follow-up period ranged from 12 to 92 months with an average of 42.6 months.

Results

Clinical Features

Among the 32 patients in this study, there were 15 men and 17 women. Age ranged from 20 to 67 years with a mean of 37.6 years. All cases were symptomatic, and the mean duration from onset of symptoms to diagnosis was 28.5 months (range: 8 months–10 years). Headache and cranial nerves dysfunction were the most common presenting symptoms. Other clinical presentations included ataxia, hemiparesis, epilepsy, and tinnitus. Two patients suffered from repeated attacks of low-grade fever and headache, which could be attributed to leakage of the cyst contents into the cerebrospinal fluid (CSF) leading to chemical meningitis. Two cases were previously operated for partial resection before they presented to our facility. Clinical presentations are summarized in ►Table 1.

Radiological Features

All tumors were hypodense on CT scans. Areas of calcification were seen in only four cases (12.5%). MRI revealed the usual appearance of hypointensity on T1-weighted images and hyperintensity on T2-weighted images in all cases. Peripheral areas of enhancement were seen in four cases (12.5%). Diffusion-weighted images (DWI) were available in 22 patients, and they showed restricted diffusion in all of them. Recently, we rely significantly on DWI, not only in the diagnosis but also in the postoperative surveillance for recurrent or residual epidermoid. The location of the lesion and its extensions into adjacent cranial compartments are shown in ►Table 2.

Surgery

Surgery was performed in all patients using the standard lateral suboccipital (retrosigmoid) approach in the lateral

Table 1 Clinical presentations

Presentation	Number of patients	Percentage
Headache	17	53.1
Cerebellar manifestations ^a	7	21.9
Trigeminal neuralgia	6	18.8
Facial numbness	5	15.6
Diplopia	4	12.5
Facial weakness ^b	3	9.4
Lower cranial nerves-related symptoms	3	9.4
Hearing impairment	3	9.4
Hemifacial spasm	2	6.3
Hemiparesis	2	6.3
Recurrent meningitis	2	6.3
Seizures	2	6.3
Visual deterioration	1	3.1

^aCerebellar manifestations include dysmetria, nystagmus, and ataxia.

^bAll cases with facial palsy were grade 3 or less on House–Brackmann scale.

position. Even in the presence of contralateral or supratentorial tumor extension, surgery was performed through the same approach. To get a better supratentorial access, the tentorium was incised in three cases in this series, and the suprameatal petrous bone had to be drilled in one case. A significant arterial compression was found at the root entry zone in three cases, and microvascular decompression of the respective cranial nerve was performed in addition to tumor removal (►Figs. 1–3).

Outcome

Total resection (tumor and capsule) was accomplished in 19 out of 32 cases (59.4%), subtotal resection (with only remnants of the capsule remaining) in 7 cases (21.9%), and only partial excision (more than 20% of tumor remaining) was

Table 2 Location and extension of the lesion

Extension of the lesion	Number of cases (%)
CPA alone	13 (40.6%)
CPA + supratentorial extension	9 (28.1%)
CPA + foramen magnum extension	4 (12.5%)
CPA + midline extension	3 (9.4%)
CPA + supratentorial and foramen magnum extension	2 (6.3%)
Bilateral CPA + supratentorial extension	1 (3.1%)
Total	32 (100%)

Abbreviation: CPA, cerebellopontine angle.

Fig. 1 (A) Preoperative axial postcontrast T1 and axial FLAIR-weighted MRI sequences of a 27-year-old male patient presenting with left trigeminal neuralgia. (B) Postoperative MRI (axial T1 with contrast and axial FLAIR sequences) performed 6 months after surgical excision of the epidermoid. FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging.

Fig. 2 (A) Preoperative MRI (axial T1- and coronal T2-weighted) of a 51-year-old female patient presenting with epilepsy, headache, and tinnitus. (B) Postoperative MRI (axial T1- and axial and coronal T2-weighted) of the same patient after subtotal resection through a right retrosigmoid approach. A small part was left in the middle fossa to avoid arterial injury. FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging.

Fig. 3 (A) Axial postcontrast T1- and axial T2-weighted MRI sequences of a 45-year-old female patient presenting with headache, left hemiparesis, and ataxia. (B) Postoperative CT after the patient developed chemical meningitis and cerebellar edema that required VP shunt application. (C) Postoperative axial postcontrast T1-weighted and (D) postoperative coronal T2-weighted MRI performed 3 months after surgery. CT, computed tomography; MRI, magnetic resonance imaging; VP, ventriculoperitoneal.

achieved in 6 cases (18.7%). Dense adhesions of the tumor capsule to vital neurovascular structures or tumor extension beyond the midline were the reasons for incomplete tumor removal.

We had a single postoperative mortality in this series. This patient had preoperative lower cranial nerves palsy and suffered at the end of the first postoperative week from aspiration pneumonia which progressed in spite of intensive antibiotic therapy to septic shock. There was no recurrence or regrowth of residual tumor during the follow-up period. New cranial nerves deficits occurred postoperatively in five patients (15.6%). They were transient in four cases while a single case suffered from a permanent trochlear palsy. A single case developed a small cerebellar infarction with no long-term effects on the patient. A list of postoperative complications is shown in ►Table 3.

Table 3 Postoperative complications

Complication	Number of cases (%)	
New CNs deficit	Temporary	4 (12.5)
	Permanent	1 ^a (3.1)
Ataxia (temporary)	3 (9.4)	
Meningitis	2 (6.3)	
Hydrocephalus	2 (6.3)	
Pneumonia	1 (3.1)	
Cerebellar infarction	1 (3.1)	
CSF leak	1 (3.1)	

Abbreviations: CN, cranial nerve; CSF, cerebrospinal fluid.

^aTrochlear nerve palsy.

Discussion

Intracranial epidermoid cysts, also known as pearly tumors or cholesteatomas, are developmental lesions that are thought to originate from aberrant ectodermal cells that become trapped during neurulation between the third and fifth week of intrauterine development.^{2,7} They are benign, slowly growing tumors commonly occurring in the CPA, and the risk of their malignant transformation is very slim.⁷⁻⁹ They have a thin epithelial-lined capsule and their growth results mostly from desquamation of epithelial cells inside the cyst, forming shiny pearly debris, in addition to cholesterol and keratin secretion into the cyst.^{2,5,7,10}

Approach

Although several authors recommended the use of more intricate approaches or staged surgery in large CPA epidermoids,¹⁰⁻¹² most series including the present one relied mainly on the lateral suboccipital (retrosigmoid) approach even with extensive tumors.^{1,4-6,13-15} In our experience, the retrosigmoid approach allows adequate exposure even without having to retract the cerebellum with a rigid retractor, as these lesions expand the subarachnoid spaces significantly and create a wide surgical corridor which is usually sufficient.^{6,16} It is a simple approach which allows early identification of the cranial nerves, and it could be easily extended both supratentorially or to the foramen magnum region. The supratentorial parts of the cyst could be accessed by following it through the tentorial incisura and gently delivering its contents downwards. If a better access to the middle fossa and Meckel's cave was needed, the tentorium could be incised, or the suprameatal petrous bone could be

drilled.^{17,18} Also, some authors advocated the use of endoscopic assistance for safe removal of tumor parts extending into adjacent cranial compartments, which could be easily performed through the retrosigmoid craniotomy.^{14,18,19}

Extent of Resection

The optimal treatment of CPA epidermoids is total surgical resection of both the cyst contents and its capsule; nonetheless, the neurosurgical judgment at the time of surgery is necessary to ensure maximum resection while minimizing postoperative neurological deficits.^{6,9,10} Few authors recommend the aggressive resection of every bit of the capsule to decrease recurrence rates.^{3,20} This could be risky in some cases where tight adhesions resulting from a long-standing inflammatory process could be found between parts of the capsule and important neurovascular structures. Taking into account the slow growth rate of these lesions, a near-total excision with only some small remnants left in a middle-aged or elderly patient may mean a clinical cure.^{5,10}

Therefore, similar to many authors, we would sometimes leave parts of the capsule intentionally if they were tightly adherent to cranial nerves, perforating vessels, or to the brainstem itself.^{5,6} The lack of intraoperative monitoring was an additional limiting factor to our ability to manage adhesions of the capsule to the brain stem and cranial nerves.¹² Adopting this strategy, we were able to achieve a total resection in 59.4% of our cases and subtotal resection (with only remnants of the capsule remaining) in 21.9%. The rate of total excision reported in the literature is widely variable ranging from 0 to 95%, but there is a tendency of higher morbidity with more aggressive surgery.^{1,5,6,21}

We did not have any recurrence or regrowth of residual tumor during the follow-up period; however, we have to acknowledge the relatively short mean follow-up period in this series taking into consideration the slow-growing nature of these lesions.

Cranial Nerves

Hyperactive dysfunction of the cranial nerves was encountered in eight patients in this series; six patients (18.8%) had trigeminal neuralgia (TN), and two patients (6.3%) had hemifacial spasm (HFS), and they have all improved after surgery. The incidence of TN associated with CPA epidermoid is particularly variable in the literature ranging from 0 to 90.6%, while HFS has a much lower incidence ranging from 0 to 13.3%.^{4,6,21-27} Hyperactive dysfunction could be the result of local irritation from cholesterol seeping through the cyst wall or direct compression of the nerve at the root entry zone. It could also result from vascular compression of the nerve either by a displaced artery or from nerve displacement toward the artery by the tumor.^{4,10,13,24,27} A compressing vascular loop was evident in three of our cases, and microvascular decompression was performed in addition to tumor removal in these cases, and the preoperative hyperactive dysfunction improved postoperatively in all three cases.

Newly added postoperative cranial nerves deficits or worsening of preexisting deficits represent a common and serious complication especially when they involve the lower

cranial nerves. Most of these postoperative deficits, however, are only temporary and tend to resolve or improve over time, as was the case in our series.^{2,3,6,13,28} In their analysis of 50 cases of posterior fossa epidermoid cysts including 37 cases of CPA epidermoids, Gopalakrishnan et al¹ found a higher incidence of new postoperative deficits in patients undergoing total resection. This can emphasize the risk of aggressive dissection of densely adherent parts of the capsule in some cases. The only mortality in the present series resulted from worsening of a preexisting lower cranial nerves palsy, which led to aspiration pneumonia and septic shock. Therefore, we think that it might be safer to perform an elective tracheostomy in patients with preoperative lower cranial nerves palsy.

Chemical Meningitis and Hydrocephalus

The incidence of aseptic meningitis in the literature ranged from 0 to 40% and was 6.3% in this series.^{4,5,10,28} This is caused by the spillage of cholesterol breakdown products into the CSF and is usually more common after incomplete resection.²⁸ This problem seems to be less commonly encountered in recent series, which could be attributed to the universal use of hydrocortisone solution for irrigation and the administration of postoperative steroids.^{4,10,28} Seeding of the irritant contents into the subarachnoid space could also be reduced by isolating the surgical field using cottonoids whenever feasible.

We did not perform any CSF diversion procedures before definitive surgery even in patients having clinical and radiological signs of hydrocephalus. Although some authors recommended preoperative shunting for patients presenting with hydrocephalus,^{3,29} we believe that it is a reversible condition in most cases after tumor resection and that preoperative CSF diversion should be reserved only for emergency cases and cases with persistent clinically significant postoperative hydrocephalus. We had only two cases (6.3%) of postoperative hydrocephalus; in one case it was transient and was treated by external ventricular drainage for 1 week and in the other case a ventriculoperitoneal shunt was applied.

Conclusion

Total removal of all the contents together with the secretory capsule is usually considered to be the standard goal when operating on CPA epidermoid cysts. This goal can be achieved in many cases; however, in some cases, it may be acceptable to leave parts of the tumor behind if they were tightly adhered to blood vessels, nerves, or the brain stem, to avoid the risk of serious neurological deficits. The suboccipital retrosigmoid approach is an adequate and safe approach to remove these tumors even if they were large in size. Coexisting arterial compression at the root entry zone should be recognized, and microvascular decompression should be performed in addition to tumor removal (—Figs. 1–3).

Conflict of Interest

None.

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