Giant intracranial epidermoids: is total removal feasible?

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OBJECT Epidermoid tumors arise from misplaced squamous epithelium and enlarge through the accumulation of desquamated cell debris. Optimal treatment consists of total removal of the capsule; therefore, giant and multicompartmental tumors are particularly challenging. A conservative attitude in handling the tumor capsule is common given concerns about capsule adherence to neurovascular structures, and thus the possibility of recurrence is accepted with the intent of minimizing complications. This study focuses on the outcome of surgery in patients with giant epidermoid tumors for which total capsule removal was the aim.

METHODS The authors conducted a retrospective analysis of all patients with giant epidermoid tumors treated by the senior author (O.A.), who pursued total removal of the capsule through skull base approaches. Patients were divided into 2 groups: one including patients with de novo tumors and the other consisting of patients who presented with recurrent tumors.

RESULTS Thirty-four patients had undergone 46 operations, and the senior author performed 38 of these operations in the study period. The average tumor dimensions were 55 × 36 mm, and 25 tumors had multicompartmental extensions. Total removal of the tumor and capsule was achieved with the aid of the microscope in 73% of the 26 de novo cases but in only 17% of the 12 recurrent tumor cases. The average follow-up among all patients was 111 months (range 10–480 months), and the average postsurgical follow-up was 56.8 months (range 6–137 months). There were 4 recurrences in the de novo group, and every case had had a small piece of tumor capsule left behind. One patient died after delayed rupture of a pseudoaneurysm. In the de novo group, the average preoperative Karnofsky Performance Scale (KPS) score was 71.42%, which improved to 87.14% on long-term follow-up. In the group with recurrences, the KPS score also improved on long-term follow-up, from 64.54% to 84.54%. In the de novo group, 3 cases (11.5%) had permanent cranial nerve deficits, and 4 cases (15.4%) had a CSF leak. In the recurrence group, 3 cases (25%) had new, permanent cranial nerve deficits, and 1 (8.3%) had a CSF leak. Two patients in this group developed hydrocephalus and required a shunt.

CONCLUSIONS Total removal of the capsule of giant epidermoid tumors was achieved in 73% of patients with de novo tumors and was associated with improved function, low morbidity and mortality, and a lower risk of recurrence. Surgery in patients with recurrent tumors was associated with higher morbidity and persistence of the disease.

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KEY WORDS brain tumor; epidermoid cyst; epithelial capsule; skull base approaches; endoscopic techniques; microsurgical removal; oncology

It is important, however, that the wall of the cyst, composed of a stratified squamous epithelium which is the only living, growing part of the neoplasm, be removed completely in order to prevent recurrence. Simple evacuation of the cyst will not suffice.

Paul Bucy⁸

Epidermoids, known as the “pearly tumor,” are benign, slow-growing lesions that constitute 1.2% of brain tumors.

They are commonly believed to arise from misplaced stratified squamous epithelium during closure of the neural tube. They enlarge by accumulating desquamated epidermal cell debris and insinuate into the subarachnoid space, at times extending into multiple compartments. By the time a patient presents for medical attention, the epidermoid may have reached a giant size.^{6,9,14,31,43,57} Although many authors acknowledge that the ultimate goal

ABBREVIATIONS CN = cranial nerve; DWI = diffusion-weighted imaging; KPS = Karnofsky Performance Scale.

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* Drs. Aboud and Abolfotoh contributed equally to this work.

of treatment is total removal of the capsule, a conservative attitude has prevailed because the capsule adheres to neurovascular structures at the cranial base (arteries, perforators, cranial nerves [CNs], and brainstem). Thus, many surgeons are inclined to leave part of the capsule behind, thereby accepting the risk of recurrence.^{4,6,23,40,44,46,56} Others advocate only a decompressive procedure with subtotal removal.^{7,10,38} Owing to the slow linear growth of the epithelium and the rate at which debris accumulates,⁵ recurrences are claimed to be late and are frequently underreported.

Regardless of the argument that the capsule is a mere remnant of epithelium, some epidermoids grow faster than natural epithelium and enlarge or recur through cellular proliferation, and thus act as a true benign neoplasm⁵³ and even occasionally a malignant one!^{1,11,27,36,37,47,53} with leptomeningeal metastasis from the intracranial malignant transformation of an epidermoid tumor.^{25,33} Furthermore, malignant conversion has been reported as the recurrence of a previously benign epidermoid.^{21,53,54} Several investigators have commented that since the capsule is the “living portion” of the tumor,^{8,12,34} recurrence is inevitable with a residual capsule. Total removal of the capsule, which is now possible with microsurgical techniques, can prevent recurrence, and if only a minute portion of the capsule is overlooked during surgery, recurrence may at least be long delayed. This approach minimizes the complications associated with repeated surgery and subsequently improves patient outcome and quality of life. Furthermore, such an approach eliminates neoplastic or even malignant transformations.

The task of eliminating the epidermoid capsule is admittedly challenging, particularly in patients with giant tumors that extend into multiple compartments. This study is a retrospective analysis of a series of patients harboring giant epidermoid tumors that expanded into multiple compartments. They underwent surgery performed by the senior author (O.A.) with the goal of total removal of the capsule. This report aims to address the feasibility of total removal of the capsule of giant epidermoid tumors and its impact on complications and outcome. We postulated that total removal of the capsule not only prevents or delays recurrence but also minimizes complications and improves long-term outcome.

Methods

We conducted a retrospective study of all patients with intracranial epidermoid tumors surgically treated by the senior author (O.A.) in the period from 1987 through 2013. The aim of treatment was total removal of the tumor capsule. We reviewed each patient's clinical findings, images, operative procedures, postoperative outcome, and follow-up. As this is a retrospective study, it has the shortcomings of such a method, including selection bias, a span of more than 2 decades, and nonadjudicated outcome analysis. But the rarity of these tumors and the lengthy time period needed for study leads to such a method.

Patients were divided into 2 groups: those with de novo tumors without prior treatment and those having a tumor recurrence after surgery at another institution. In calculating

the total number of surgical procedures performed by the senior author in all patients, we counted second operations that were performed in patients in the de novo group, bringing the total to 38. Tumor size was measured in two dimensions on MRI studies and was calculated using Centricity software (Centricity, GE Healthcare) and OsiriX for the Macintosh (OsiriX).

Results

The series comprised 34 patients, 18 males and 16 females, with an average age of 35.77 years (range 9–67 years) at the first surgery performed by any surgeon. Twenty-six patients had de novo tumors without prior treatment, and 8 patients had a recurrence after surgery at another institution. The average age at surgery for recurrence (12 cases) was 44.9 years (range 20–63 years; Fig. 1). A total of 38 operations were performed for resection, 26 in patients with de novo tumors and 12 in those with recurrences (8 patients presented to us with recurrences, and 4 had recurrences after our initial surgery).

The average size of the tumor before surgery in the de novo group was 55.03 × 35.48 mm, while in the recurrence group it was 54.38 × 38.73 mm. The largest tumor measured 102 × 76 mm, while the smallest was 12 × 26 mm. Before any surgery, 72% of the tumors had at least 1 diameter ≥ 40 mm. These tumors had primary locations with extensions into various compartments in the same or adjacent cranial fossa. Details of the primary tumor locations and extensions for the 2 groups are summarized in Figs. 2 and 3.

Clinical Findings

The duration of symptoms in this series varied widely, from 5 days with hydrocephalus to a 30-year history of headache. The average duration of symptoms was 28.43 months. Symptoms also varied according to tumor location. Cranial nerve involvement was common in tumors in the cerebellopontine angle, while deteriorating vision was the most common presentation for patients with primary or secondary suprasellar epidermoids. Headache was common for tumors in any location and ranged from severe and

FIG. 1. The age distribution of patients at presentation. SD = standard deviation.

FIG. 2. De novo tumors involved 26 surgeries. Pie chart indicates distribution of primary tumor locations. Middle fossa refers to both the middle fossa and the temporal fossa. Bar graphs indicate the frequency of areas of extension of the cerebellopontine angle (*right graph*) and preoptine cistern (*left graph*) tumors. Temporal fossa refers to both the middle fossa and the temporal fossa. Figure is available in color online only.

FIG. 3. Recurrent tumors involved 12 surgeries. Pie chart indicates distribution of primary tumor locations. Bar graphs indicate the frequency of areas of extension of the cerebellopontine angle (*right graph*) and the suprasellar (*left graph*) tumors. Figure is available in color online only.

refractory to pain medication, to mild and lasting for years. The main symptoms at presentation are listed in Table 1.

Imaging

For all cases, we reviewed preoperative, postoperative, and follow-up MR images with and without gadolinium enhancement and diffusion-weighted images, as well as CT scans. Findings differed between the de novo and recurrent groups. Keratin contents were all positive in the diffusion sequences. Faint capsule enhancement was present in only 2 cases (7.7%) in the de novo group, while the capsule enhanced significantly in 5 cases (41.7%) in the recurrent group. Septation of the tumor was rare in patients with de novo tumors, occurring in only 4 cases (15.4%), while it occurred in 5 cases (41.7%) of recurrent tumor. At the second recurrence, all cases showed an enhanced capsule and multiloculation.

Surgical Technique

To achieve a wide exposure, to access all parts of the lesion, and to pursue total removal of the capsule, we used the skull base approaches listed in Table 2 and shown in Fig. 4. Because several CNs were compressed, involved, encased, or adherent to the brainstem (at times bilaterally), we used intraoperative neurophysiological monitoring, including somatosensory evoked potentials and brainstem auditory evoked responses, and extensive individual monitoring of all potentially involved CNs. Neuronavigation has been used routinely since 1993.

After evacuating the keratin contents with controlled suction, the surgeon performs meticulous microdissection with two hands to tease adherent capsule from the arachnoid layer, thereby freeing the neurovascular structures. Traction is avoided and sharp microdissection is used (Fig. 5). In all de novo cases, a plane of dissection can be developed between the capsule and a compressed overlying arachnoid; it is essential to identify and confirm this plane and preserve it as long as possible. When it is unclear which portion is tumor capsule and which is arachnoid, a frozen section is sent for analysis (Fig. 6). If this analysis

TABLE 2. Approaches used in 38 surgeries for epidermoid tumors

Surgical Approach	No. of Cases
Transmastoid retrosigmoid	12
Combined petrosal	8
Posterior petrosal	6
Cranio-orbital zygomatic	5
Transcondylar	2
Pterional*	2
Total petrosectomy	1
Zygomatic	1
Anterior petrosal	1

* Used twice in the same patient for 2 recurrences.

shows arachnoid only and no epithelial tissue, there is no need to pursue the membrane further.

In most of the patients with de novo tumors, the capsule was thin, avascular, or fibrotic and was easily dissected from neurovascular structures; however, it adhered to neurovascular structures in some cases (Fig. 7). Small perforators can traverse the contents of the cyst, perhaps because of the wrinkling or enfolding of the epithelial layer. In all surgeries for recurrent epidermoid tumors in this series, the capsule was thick and adhered to surrounding structures. We also noticed that the capsule became increasingly thick and fibrous with subsequent recurrences, a situation that can lead to the formation of loculated cysts of CSF or yellowish fluid that can compress surrounding structures.

The endoscope was used in 13 operations. It can reveal remnants of tumor hidden around the corner from the direct microscopic axis or into the foramina (internal auditory meatus, Meckel's cave, and jugular foramen). It assists in reaching a supratentorial extension when a purely posterior fossa approach is used (Fig. 5F).

The surgical site is walled off to minimize the spread of irritating substances and the risk of aseptic meningitis. Irrigation is used after resection is complete, and any debris is suctioned away. High doses of steroids are administered intraoperatively and slowly tapered over 2 weeks after surgery. Postoperative MRI is performed with diffusion sequences to verify gross removal of the contents and as a baseline for follow-up (Fig. 8).

Extent of Resection

Our evaluation of resection was based on the surgeon's notes in the operative reports regarding the extent of capsule removal. Microscopic total removal of the capsule was achieved in 19 cases (73%) in the de novo group but in only 2 (16.7%) of the 12 surgeries on recurrent tumors. Unfortunately, there is no postoperative imaging modality or tool that can be used to evaluate the degree of capsule resection. Diffusion weighted imaging confirms total removal of the contents, but postoperative DWI always showed bright signal in patients with a residual capsule from which all debris was thought to be totally removed (Table 3), probably because of microscopic flakes attached to the residual capsule.

TABLE 1. Signs and symptoms at presentation in 34 patients

Signs & Symptoms	% of Patients
Headache (variable degrees)	42
Weakness (variable distributions)	20
Hearing loss (variable degree)	18
Diplopia	16
Gait disturbance & ataxia	12
Trigeminal neuralgia	12
Facial numbness	10
Facial weakness (variable degree)	10
Vertigo & tinnitus	8
Swallowing difficulty	8
Seizures	6
Deterioration of vision	6
Hydrocephalus	4
Hemifacial spasm	2

FIG. 4. **A:** Preoperative axial T2-weighted MR image showing the high-intensity epidermoid. **B:** Preoperative sagittal T1-weighted MR image showing the lesion. **C:** Intraoperative photograph depicting the wide exposure obtained through a posterior petrosal approach to a giant epidermoid tumor that extends infra- and supratentorially.

Outcome

The average follow-up was 111 months (range 10–480 months), and the average postsurgical follow-up was 56.8 months (range 6–137 months). There was one perioperative death (de novo group) from delayed rupture of a pseudoaneurysm after a suture repair of the A1 segment. In the de novo group, the average Karnofsky Performance Scale (KPS) score before surgery was 71.42% (range 50%–90%), which improved to 76.66% immediately after surgery and further improved to 87.14% during the follow-up period. In the group with recurrences, the average KPS score before

surgery was 64.54% (range 60%–80%). After surgery, this score improved immediately to 71.81% and continued to improve to 84.54% during the follow-up period. Table 4 shows the KPS scores, which were available in 31 patients.

Among the 26 de novo cases, new, permanent CN deficits occurred in 3 cases (11.5%). Transient CN deficits occurred in 9 cases (34.6%) and CSF leakage in 4 cases (15.4%). Three patients with CSF leakage required reexploration and 1 patient had a seizure.

In the recurrence group, new, permanent CN deficits occurred in 3 cases (25%), and transient CN deficits oc-

FIG. 5. A giant epidermoid tumor in the posterior fossa treated through a transmastoid retrosigmoid approach. **A:** Axial T2-weighted MR image. **B:** Sagittal T1-weighted MR image showing the suprasellar extension. **C:** Intraoperative photograph showing sharp dissection of the involved neurovascular structures. *Arrow* indicates the anterior-inferior cerebellar artery (AICA); *arrowhead*, the AICA–posterior inferior cerebellar artery trunk. **D:** Intraoperative photograph showing separation of the capsule from the thin arachnoid layer engulfing the basilar artery (BA). **E:** Intraoperative photograph demonstrating the surgical field after a two-handed separation of the capsule from the arachnoid layer over CN VII. **F:** Intraoperative photograph showing hidden remnants that were pursued endoscopically for removal. C = tumor capsule; PCA = posterior cerebral artery; III = oculomotor nerve; VI = abducens nerve; VII = facial nerve.

FIG. 6. A and B: Photomicrographs of frozen sections obtained from two patients during intraoperative consultation. The cyst wall has a stratified squamous epithelial lining along with a granular layer (arrows), and the cyst has epidermal keratinous contents (asterisks). **C and D:** Photomicrographs of frozen sections obtained from the same patients during intraoperative consultation. Note the arachnoid membrane with meningotheelial cell groups (asterisks) along with psammomatous calcifications (arrows) commonly associated with these cells. No evidence of stratified squamous epithelium or keratin formation is visible. H & E, original magnification $\times 200$.

occurred in 4 cases (33.3%). CSF leakage requiring reexploration occurred in 1 case (8.3%) and hydrocephalus requiring shunt placement occurred in 2 cases (16.7%). In 3 cases of cerebellopontine angle tumors, in which a piece of tumor was left behind, multiple transient CN deficits and a recurrence developed. In the 8 patients who underwent initial surgery elsewhere, all had considerable postoperative residual tumor, as subtotal resection was the initial intent of surgery. Six of these patients (75%) had permanent CN deficits from their original surgery.

FIG. 7. A minute piece of tumor capsule was left behind, because of its adherence to the BA, and led to multiple recurrences. **Left:** Intraoperative photograph obtained at the original surgery. Arrowhead indicates the perforators. **Right:** Axial T1-weighted MR image showing a recurrence that extended to the quadrigeminal cistern. BS = brainstem.

Recurrence

Recurrence was diagnosed when progressive symptoms were associated with an increase in tumor size with a bright signal on DWI or extension into a new compartment. At recurrence, the tumor frequently changed its primary location, as it extended from the periphery of the original resection to other areas. Moreover, it showed more frequent capsule wall enhancement and multiloculations (Fig. 9). In the de novo group, 4 cases (15.4%) had a recurrence, while 5 cases (41.7%) in the recurrence group had a subsequent recurrence. In the de novo cases, we performed a Kaplan-Meier estimate to compare the rate of recurrence between 2 groups divided according to capsule removal: Group 1 had no residual capsule and Group 2 had a residual capsule (Fig. 10).

Follow-up was calculated for each operation. It was relatively short for patients with repeated recurrences, at an average of 56 months. The average calculated among all patients was 111 months. The average recurrence-free survival period after the first surgery was 121.5 months (range 33–150 months), while the recurrence-free period before the second recurrence decreased to 54.5 months (range 36–96 months).

Discussion

Epidermoids are benign lesions with rare reports of malignancy or malignant progression.^{7,13,44} There is no alter-

FIG. 8. An example of the emphasis on postoperative MRI studies for baseline follow-up, highlighting the importance of diffusion images. **A:** Preoperative axial enhanced T1-weighted MR image. **B:** Preoperative axial diffusion-weighted image with positive bright signal. **C:** Preoperative coronal T2-weighted MR image. **D:** Preoperative sagittal T1-weighted MR image. **E:** Postoperative axial T1-weighted MR image. **F:** Postoperative axial diffusion-weighted image showing no bright signal after total removal. **G:** Postoperative coronal T1-weighted MR image. **H:** Postoperative sagittal T1-weighted MR image.

native to surgery for epidermoid cysts,^{18,51,56,57} especially in younger patients.^{2,20,42} Since the squamous stratified epithelium that constitutes the capsule is the real disease, total removal of the capsule is acknowledged to be the ultimate treatment.^{8,18,51,57} Because the capsule adheres to the brainstem, CNs, and perforating arteries, however, a conservative attitude toward removal has prevailed,^{6,22,40,44,46,51,56} and many surgeons leave part of the capsule behind, accepting a higher risk of recurrence, especially for tumors extending into multiple sites.^{9,51} Deterred by the risk involved in dissecting the capsule, other authors have opted for a decompressive procedure by evacuating the contents^{7,38,44} or have not even attempted to handle the capsule.⁵⁵

With a capsule remnant, recurrence is inevitable with time, and the capsule cannot be totally removed in recurrent cases. The use of microsurgical techniques, skull base approaches, intraoperative neurophysiological monitoring, neuronavigation, and endoscope-assisted dissection facilitates complete removal of these lesions in many cases.^{44,51,57} Our study shows that total capsule excision can be done in a large number of cases and not only minimizes future recurrence but is also associated with fewer complications

after the first surgery. This approach also eliminates the added morbidity of repeated surgery that has such a drastic impact on patient quality of life, especially since patients are relatively young at their first presentation.^{2,4}

Surgical Outcome and Extent of Tumor Resection

The early high rates of morbidity and mortality in patients with epidermoid tumors has fallen dramatically in the microsurgical era.^{46,57} The most frequent surgical complications are aseptic meningitis and multiple CN deficits. Some authors commonly believe that surgical complications can be decreased by avoiding dissection of the capsule that adheres to neurovascular structures.^{23,46,56} Lunardi and colleagues³⁸ concluded that the absence of operative mortality among patients who underwent subtotal removal and the late recurrence justify subtotal removal.

We attempted total removal, diligently pursuing the capsule not only to diminish the chance of recurrence, but also to reduce complications. It is well known that the remnants of capsule and cyst material are the main causes of the aseptic meningitis and hydrocephalus described in some reports.^{7,24} Our findings are supported by an analysis of major, modern large series reported in the literature. Tables 5–7 list 22 major reports divided into 3 groups to compare the postoperative results of different strategies related to the degree of tumor resection: Group 1: conservative, total removal in less than 25% of cases; Group 2: moderate, total removal in 28%–60% of cases; and Group 3: extensive, total removal in more than 62% of cases. The mean rate of total removal was 12% in the first group, 46% in the second, and 78% in the third.

TABLE 3. Residual tumor after de novo and recurrent surgeries

Group	No. of Cases (%)	
	Intraop Residual Capsule	Postop Residual on DWI
De novo (26 surgeries)	7 (26.9%)	6 (23.1%)
Recurrent (12 surgeries)	10 (83.3%)	11 (91.7%)

TABLE 4. Karnofsky Performance Scale scores for 31 patients*

Group	Time	No. of Cases					
		100%	90%	80%	70%	60%	50%
De novo (20 cases)	Preop		2	8	3	6	1
	Immediate postop		6	9	3	1	1
	On FU	7	10	2			1
Recurrence (11 cases)	Preop			1	3	7	
	Immediate postop		1	4	5		1
	On FU	2	3	5		1	

FU = follow-up.

* Karnofsky Performance Scale score data were available in only 31 patients.

Mortality rates appear higher in Group 1 and certainly not less in Group 2, compared with Group 3, in which aggressive attempts were made to achieve a high percentage of total removal. Aseptic meningitis is more common in patients undergoing conservative approaches, probably because of the residual lesion's continuous direct contact with brain tissue and CSF after partial or subtotal removal.^{14,24,40,44,51} This complication was almost absent in our series and has been less frequent in other contemporary reports with high rates of total removal and, perhaps, the routine prolonged use of high doses of corticosteroids.^{17,45}

A lower incidence of CN deficits was noted in a postoperative series with a high rate of total removal,³² but this result should be credited to the microsurgical techniques

and intraoperative monitoring rather than to conservative handling of the capsule or the deliberate goal of subtotal removal.^{44,51,57} Furthermore, since our series comprised giant tumors that involved multiple CNs, at times bilaterally, and a relatively low rate of postoperative CN deficits, we believe that the skull base approach is an important factor in minimizing postoperative CN deficits.

Neuroimaging

Epidermoid tumors usually appear as a low-density mass on CT and have low signal intensity on T1-weighted MRI, high signal intensity on T2-weighted images, and hyperintensity on diffusion sequences with no enhancement.

FIG. 9. An example of repeated recurrences with capsule enhancement, migration of the mass, and positive bright diffusion signals. Preoperative axial (A) and sagittal (B) T1-weighted MR images showing the tumor. Postoperative axial T1-weighted image (C) demonstrating residual tumor in the intrapeduncular cistern. Axial T1-weighted image (D) obtained at the time of the first recurrence, demonstrating capsular enhancement at the time of the first recurrence. Axial diffusion-weighted image (E) showing positive signal when the patient was experiencing deteriorating vision. Coronal (F) and axial (G) enhanced T1-weighted images depicting tumor extension high in the suprasellar area and low around the brainstem at the second recurrence. Axial diffusion-weighted image (H) with positive bright signals at the second recurrence.

FIG. 10. Kaplan-Meier graph indicating the recurrence-free survival time for de novo cases divided into 2 groups, according to the extent of capsule removal (total removal or not): Group 1, no residual capsule; Group 2, residual capsule.

Calcification, hyperdensity, and marginal enhancement are rarely seen.^{3,14,26,28,29,41,50,51} Thus, these characteristics depict the contents of the cyst and aid in the accurate preoperative diagnosis. Postoperative imaging can only confirm removal of the cyst contents by the absence of hyperintensity on diffusion studies. Follow-up radiographic monitoring is crucial for detecting tumor recurrence through an increase in the size of residual lesions. Magnetic resonance imaging is the best modality for documenting tumor growth.

Occasionally, partial or moderate rim enhancement has been reported, particularly with a recurrence, and is frequently caused by reactive granulation.²¹ Nonetheless, this enhancement has been thought to be an indication of neoplastic proliferation or a manifestation of malignancy.¹ Our cases with obvious enhancement did not show signs of malignancy. Radiological confirmation of recurrence is not always easy. The cavity that remains after tumor has been excised fills with CSF, which appears on CT scans as a low-density area that resembles the original tumor and may be confused with recurrence.^{18,39,51} Multiple loculations on MRI and enhancement of the periphery further cloud the picture. Therefore, intervention for a recurrence should be based on documented clinical symptoms or a progressive, compressive accumulation seen on DWI, which can solely differentiate an epidermoid from a retained cyst or encephalomalacia, especially in patients with multiple recurrences.^{46,50,51}

Recurrence and Late Outcome

With a residual capsule, recurrence is inevitable given that the epidermoid cyst is a mere accumulation of cellular debris from a remnant of keratinized squamous epi-

thelium. The variable rate of recurrence reported in the literature is attributed to a short follow-up period.^{32,45,55} As Tables 5–7 show, recurrence rates were lower in Group 3 (a higher rate of total removal) for comparable follow-up periods in the other groups.

Given a hypothesis of linear growth and a constant rate of debris accumulation, it is believed that the time required for the recurrence of a tumor the same size as the original is equal to the patient's age at the initial surgery plus 9 months.⁵ Justifying the practice of subtotal removal and conservative handling of the capsule,^{7,44,46,57} this hypothesis assumes that as little as a single cell remains postoperatively and that the recurring mass grows at the same rate as the original tumor.⁴⁴

Multiple investigators suggest that the actual proliferation of this epithelium beyond the normal remnant should be considered a neoplasm,^{51,53} even with an incidence of malignant progression; hence, they should be treated the same as other benign tumors for which surgical removal is the goal. Echoing other authors,^{19,24} Altschuler and associates⁴ stated that repeated surgical attempts on these tumors are difficult and fraught with neurological complications for the patient. Repeated surgery is associated with accumulated deficits and morbidity and the expectation of another surgery.⁵¹ With repeated surgery, an inflammatory reaction with severe adhesion takes place, and the opportunity for total removal is lost. Moreover, the period between surgical interventions shortens with every recurrence. These findings emphasize that results are better and recurrence is delayed when resection of the tumor and capsule is as close to complete as possible.

One of the most important factors that facilitate total

TABLE 5. Literature summary of studies in which total removal occurred in less than 25% of cases, Group 1

Authors & Year	Calcification (% cases)	No. of Cases	Total Removal (% cases)	Mortality (% cases)	Cyst Recurrence (% cases)	FU (yrs)	Aseptic Meningitis (% cases)	Complications (% cases)	Cranial Neuropathy (% cases)	Giant Tumor				Use of Combined Techniques (% cases)	% Multi- Staged Cases
										Occupying Multiple Compartments (% cases)	Supra- & Infratentorial Extension (% cases)	Major Complaint	Complaint (% cases)		
Berger & Wilson, 1985	15.38	13	0	0	0	4.5	15.3	30.77	23.07	15.3	69.2	CMS, TN	7.69	7.69	7.69 PFA, FTA
Sabin et al., 1987	8.69	23	8.6	4.3	21.73	6	13.04	34.78	21.73	78.26	43.47	GA, HL, HA, Vo	30.43	PFA, TC	4.34 PFA, TC
de Souza et al., 1989	NA	27	18.51	3.7	14.81	5	11.11	37.03	18.5	14.81	14.8	FP, HL, Vo	14.81	PFA, MFA	NA
Vinchon et al., 1995	NA	9	0	22.2	0	3.05	22.2	55.5	33.33	4.44?	44.4	HL, CMS, HA	0	55.55	SRA, STA
Tancredi et al., 2003*	NA	9	22.2	11.11	33.33	14.5	33.3	33.3	11.1	66.66	NA	Vo, WD	0	0	0
Desai et al., 2006†	8.33	24	25	0	4.16	5.2	0	0	0	20.83	83.33	HA, A, VL	0	4.16	ISA, IPPOA
Mean	10.8	17.5	12.385	6.885	12.338	6.375	15.83	31.905	17.95	33.38	51.04		8.82		14.34

A = ataxia; CMS = cerebellopontine mass sign; FP = facial paralysis; FTA = frontotemporal approach; GA = gait ataxia; HA = headache; HL = hearing loss; IPPOA = interhemispheric posterior parietooccipital approach; ISA = infratentorial supracerebellar approach; MFA = middle fossa approach; NA = not available; PFA = posterior fossa approach; SRA = suboccipital retrosigmoid approach; STA = subtemporal approach; TC = temporal craniotomy; TN = trigeminal neuralgia; VL = visual loss; Vo = vertigo; WD = walking difficulty.

* Epidermoid cyst of the fourth ventricle.

† Pineal epidermoid cyst.

TABLE 6. Literature summary of studies in which total removal occurred in 28%–60% of cases, Group 2

Authors & Year	Calcification (% cases)	No. of Cases	Total Removal (% cases)		Mortality (% cases)	Cyst Recurrence (% cases)	FU (yrs)	Aseptic Meningitis (% cases)		Complications (% cases)	Cranial Neuropathy (% cases)	Giant Tumor Occupying Multiple Compartments (% cases)	Supra- & Infratentorial Extension (% cases)	Major Complaint	Use of Combined Techniques (% cases)	Multi-Staged Cases (% cases)
Yamakawa et al., 1989	15.15	33	36.4	12.12	12.12	24.13	8.83	12.12	35.7	15.15	15.15	15.15	12.12	T, S, GD, HA	NA	0
Lunardi et al., 1990	NA	17	35.2	17.6	17.6	11.7	9	17.6	17.6	NA	17.6	NA	NA	CMS, TN, HS	0	0
Altschuler et al., 1990	7.69	13	54	0	30.76	15.38	4.6	0	30.76	30.76	15.38	30.76	38.46	GD, HA	7.69 RMA, FTA	7.69
Mohanty et al., 1997	8	25	48	8	8	0	3.6	8	56	100	20	100	76	CMS, CN5, CN7, FP, HL	4 RMA, STA	NA
Talacchi et al., 1998	17.85	28	57	3.5	14.3	30	8.6	14.3	28.57	60.7	10.7	60.7	60.7	HA, T	10.71	NA
Kaspera et al., 1999	NA	17	50	11.7	7	16.7	3.3	7	NA	NA	13	NA	23.5	NA	NA	NA
Kobata et al., 2002	6.66	30	56.7	0	6.6	6	11.5	6.6	53.33	NA	10	NA	NA	CN5, HS	0	0
Goel et al., 2006	5.20	96	48	2.1	2.1	2.1	4.3	2.1	5.2	87.5	23.9	87.5	87.5	HA, GD	2.08	NA
Ahmed et al., 2009*	NA	7	28.6	0	28.6	28.6	4.9	28.6	71.4	57.14	42.8	57.14	0	HA	NA	0
Mean	10.09	29.56	45.99	6.11	14.96	6.51	6.51	10.7	37.32	58.54	18.73	58.54	49.71		6.12	1.54

CN5, CN7 = deficits in cranial nerves 5, 7; GD = gait disturbance; RMA = retromastoid approach; S = seizure; T = tinnitus.

* Epidermoid tumors in 3 children.

TABLE 7. Literature summary of studies in which total removal occurred in 62% or more cases, Group 3

Authors & Year	Calcification (% cases)	No. of Cases	Total Removal (% cases)		Cyst Recurrence (% cases)	FU (yrs)	Aseptic Meningitis (% cases)		Complications (% cases)	Cranial Neuropathy (% cases)	Giant Tumor Occupying Multiple Compartments (% cases)	Supra- & Infratentorial Extension (% cases)	Major Complaint (% cases)	Use of Combined Techniques (% cases)	% Multi- Staged Cases
Yaşargil et al., 1989	5.71	35	97	4.65	0	5.2	18.06	30.23	20.43	100	20	HA, Di	0	0	0
Fornari et al., 1990	9.09	14	85.6	7.1	7.1	13	7.1	14.2	14.2	NA	NA	Vi, S	0	0	0
Samii et al., 1996	NA	40	75	2.5	7.5	5.7	2.5	17.5	10	NA	62.5	HL, Di	NA	NA	NA
Darrouzet et al., 2004	NA	8	62.5	0	0	3.7	12.5	50	50	100	NA	HA, Deq	25	0	0
Kaylie et al., 2005	NA	10	80	10	10	1.75	0	40	40	NA	NA	HL, HA, Vo	50 TL+TC	0	0
Safavi-Abbasi et al., 2008	NA	12	75	0	0	2.25	0	8.33	8.33	83.33	8.33	PyS/CMS, HA	8.33, LSOA, STA	8.33	8.33
Chowdhury et al., 2013	4.35	23	73.9	8.7	0	3.25	0	21.74	13.04	NA	NA	HA, Vo	0	0	0
Mean	6.38	20.23	78.43	4.7	3.51	4.98	5.74	26	22.29	94.44	25.97		13.89	1.39	

Deq = disequilibrium; Di = dizziness; LSOA = lateral suboccipital approach; PyS = pyramidal sign/cerebellar; TL = translabyrinthine; Vi = visual disturbances including restriction of eye movements.

removal is adequate exposure of the lesion.^{32,49,51} In the patients who came to us with a recurrence, exposure during the previous surgery had been limited and did not allow access to all extensions of the tumor, preventing the surgeon from resecting the capsule in these regions. We extended the previous approaches with wide skull base exposures³ to allow better access to all portions of the tumor.

Giant Tumors and Extent of Resection

Both old and recent series in the literature include some cases of giant tumors that extend into multiple compartments.^{4,40,44} These tumors are formidable, and the goal of total removal of the capsule is quite challenging. These cases have been fraught with a high risk of failure and recurrence.^{40,44} In such cases, surgeons have resorted to subtotal resection or a staged operation through 2 different approaches: the use of an endoscope to visualize beyond the restrictive limits of the traditional approach or the use of a combined infra- and supratentorial approach.^{15,44,48} Altschuler and colleagues⁴ found that patients who required multiple procedures resulting in fixed deficits had an extensive lesion and incomplete initial resection, which they believed to be attributable to a limited initial exposure. They contemplated whether the use of a more extensive approach in a first operation would have resulted in better tumor resection, improved outcome, and reduced recurrence.

We strongly believe, and the results of our study confirm, that full exposure through a skull base approach not only increases the likelihood of total removal of this lesion, including the capsule, but also alleviates operative complications, CN deficits, and aseptic meningitis. Our findings support the role of a skull base approach in effectively and safely removing the capsule.

Conclusions

Total removal and complete resection of the capsule can be achieved in the majority of patients with giant epidermoids and is associated with low morbidity and mortality. This goal is facilitated by a thorough preoperative evaluation, an approach that allows wide exposure, sharp microsurgical removal, extensive intraoperative CN monitoring, an endoscope-assisted technique, and neuronavigation. The pursuit of total capsule removal minimizes not only recurrences but also the risk of postoperative aseptic meningitis, CN deficits, and subsequent complications from repeated surgery for recurrences.

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