Original Article

Surgical strategy for intracranial dermoid and epidermoid tumors: An experience with 33 Patients

Jose Carlos Lynch, Antônio Aversa, Celestino Pereira, Jânio Nogueira, Mariangela Gonçalves, Hêlio Lopes

Department of Neurosurgery, Hospital Federal dos Servidores do Estado and National Cancer Institute, Rio de Janeiro, Brazil

E-mail: *José Carlos Lynch - jclynch@globo.com; Antonio Aversa - aversadosouto@gmail.com; Celestino Pereira - celestino.pereira@br.inter.net;
Jânio Nogueira - JanioNogueira@inca.gov.br; Mariangela Gonçalves - goncalvesmbl@gmail.com; Hêlio Ferreira Lopes - Hlopes@inca.gov.br

*Corresponding Author

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Abstract

Background: The aim of this paper is to report on our surgical strategy and technique and to identify the best management for intracranial dermoids and epidermoids tumors (IDETs).

Methods: We retrospectively reviewed 33 consecutive patients (14 males and 19 females; mean age at surgery, 37.9 years) with pathologically confirmed IDETs who underwent surgical resection, with mean follow-up of 7.2 years.

Results: Gross total tumor removal was achieved in 24 cases (72.7%) with zero surgical mortality and a recurrence rate of 9%.

Conclusions: The surgical strategies used in this group of patients enabled total removal of most tumors without surgical mortality and with low morbidity and recurrence rates, proving to be safe and effective.

Key Words: Dermoids, epidermoids, intracranial tumors, microsurgery, surgical treatment

INTRODUCTION

Intracranial dermoids and epidermoids tumors (IDETs) are congenital, slow-growing tumors that develop between the third and fifth weeks of gestation from ectodermal remnants during neural tube formation in embryogenesis. Epidermoid or “pearly” tumors were described by Cruveilhier and designated the “most beautiful tumors of the body” by Dandy. Epidermoids are lined by a delicate capsule of stratified squamous epithelium while dermoids include, in addition to skin, numerous hair follicles as well as sebaceous and sweat glands. Epidermoids tumors grow linearly rather than exponentially and hence are slow-growing lesions by accumulation of keratin and cholesterol, which are the breakdown products of desquamated epithelial cells. These tumors are rare, comprising around 1-2% of all intracranial tumors. They are most commonly located in the cerebellopontine angle (CPA) and in the parasellar area. In the CPA, IDETs constitute 7.9% of all tumors. Despite the development of microsurgery and cranial base techniques, the surgical management of IDETs continues to be a formidable technical challenge to neurosurgeons because these tumors grow in close contact with neural and vascular structures that cannot be sacrificed or retracted. A controversial debate has continued for several years, which has not been completely settled to date: Should the neurosurgeon promote gross total tumor...
removal (GTR), which can result in unwarranted cranial nerve (CN) deficit and arterial injury, or should GTR be avoided and a safer, subtotal tumor removal (STR) be performed instead?

The objectives of the current paper are to report on our operative strategy and surgical technique and to identify the best operative management for IDETs.

MATERIALS AND METHODS

Data collection
This study included 33 consecutive patients with IDETs who underwent surgery between January 1986 and January 2013 at the Department of Neurosurgery of the Hospital Federal dos Servidores do Estado (HFSE), the National Cancer Institute (INCA), and a private clinic in Rio de Janeiro, Brazil. The medical charts, pre- and postoperative imaging, and pathological reports of the patients were retrospectively reviewed to ascertain the diagnosis of IDET, thereby creating a database from which information pertinent to the present study was collected. The intraoperative videos and/or photos of 21 patients were analyzed for nuances of the microsurgical technique. Informed consent was waived due to the retrospective character of the study. Computed tomography (CT) and magnetic resonance imaging (MRI) scans were reviewed with the radiology department [Figure 1a, b]. Control postoperative imaging studies were performed within the first 48-72 h after surgery to document the extent of resection and postoperative changes [Figures 2d and 3b]. The first clinic visit occurred approximately 15 days after hospital discharge, with subsequent visits at 2 and 6 months. Thereafter, the patients were scheduled for revision at 1-year intervals. They were contacted for imaging studies and clinic visits or interviewed by telephone. In this series, two recently introduced techniques were used: Neurophysiological monitoring and neuronavigation. The Glasgow Outcome Scale (GOS) defined the outcome.

Neurosurgical approaches and microsurgery techniques
The neurosurgical approach to IDETs depends on tumor location and growth pattern. We used a tailored surgical approach to manage each case: (i) CPA tumors were removed via a suboccipital retrosigmoid approach (39.3%); (ii) Sellar and parasellar lesions were accessed by a pterional craniotomy with wide splitting of the Sylvian cistern arachnoid membrane (18.1%), (iii) Vermian and fourth ventricular lesions were operated upon through a midline suboccipital craniotomy (15.1%); (iv) frontobasal and frontoparietal lesions (12.1%) were removed via a frontobasal craniotomy and a frontoparietal craniotomy, respectively. Considering that IDETs situated in the CPA are the most challenging to remove, we will describe our surgical techniques in managing 13 IDETs located in the CPA.

Surgical technique: We used the same standard microsurgery technique in all operations, following these general steps. Patient positioning: Under endotracheal general anesthesia, most patients were placed in a semi-sitting position, with the head flexed forward and secured in the Mayfield three-point fixation device. Some patients, however, were positioned in dorsal decubitus with contralateral head rotation and ipsilateral shoulder elevation (surgeon’s preference). An arterial line and a central venous catheter were placed in all patients. The involved region was shaved, prepped, and draped in a

Figure 1: T1-weighted MRI scans with contrast enhancement in cases of IDETs found in this series. a (sagittal) and b (axial).
sterile fashion. Routine antibiotics, dexamethasone, and manitol were used. The procedure was initiated with the use of a 2.5× surgical loupe and co-axial lighting for soft tissue incision and craniectomy. Skin incision and muscle dissection: An 8-cm vertical linear incision was made 3 cm medially to the mastoid and centered approximately 2 cm above the mastoid tip. The incision was carried down through the galea and periosteum over the suboccipital bone. Emissary veins that opened during subperiosteal dissection were controlled using bipolar coagulation and waxed immediately, with repeat waxing at the end of the surgery. The spinous process of the second cervical vertebra was the palpatory bony landmark guide to the position of the foramen magnum and enabled a safe subperiosteal dissection of the suboccipital region to be carried along the posterior C1 arch. The paravertebral muscles were detached from the occipital bone squama and progressively sectioned with a scalpel. A self-retaining retractor was gradually inserted into the wound, thus exposing the suboccipital triangle and maintaining the paravertebral muscles in the appropriate position. At that point, the posterior arch of C1 was identified. Craniectomy: Suboccipital craniectomy was performed unilaterally using a high-speed drill to thin the suboccipital bone. Then, a regular Leksell rongeur was used extending from the posterior edge of the occipital bone condyle to the inferior and lateral edge of the transverse and sigmoid sinus, respectively. In some of the more recent patients, craniotomy was performed using a power circular saw. The access thus achieved provided sufficient midline and lateral suboccipital exposure of the tumor. The operating microscope was introduced in the surgical field and the operation proceeded with magnification ranging from 10 to 20×. Opening the dura: The dura was opened by a horseshoe-shaped incision with its base toward the midline. The dural edges were tented up. The cisterna magna was incised and the cerebrospinal fluid (CSF), allowed to drain spontaneously. The combination of bone removal, dural opening, and CSF drainage provided excellent visualization of the entire CPA with minimal cerebellar retraction. Exposing the lesion: The tumor was then visualized under the arachnoid as a bright white lesion. Exposure was improved after gentle elevation of the cerebellum, which was maintained in position with fixed retractors. The arachnoid attachments along the cerebellum in the cerebellopontine cistern were opened and the arachnoid membrane enveloping the tumor was incised in the posterior aspect of the exposed area using microscissors. Then, the arachnoid membrane was gently dissected away from the tumor surface to the sides using microsurgery instruments. The arachnoid membrane was left intact to protect the brainstem from surgical dissection. At that point, the high magnification provided by the operating microscope proved particularly beneficial. Although IDETs are soft and usually poorly vascularized lesions, thorough hemostasis was obtained throughout the procedure using bipolar forceps to keep the field bloodless. Debulking the tumor: After low-current bipolar coagulation of a few fine vessels travelling on the tumor surface, under saline irrigation, the lesion was incised, penetrated, and progressively debulked from within, mobilized, and removed in piecemeal fashion. Careful attention was paid to identifying and preserving the arachnoid plane at the tumor–brainstem interface, which facilitated complete tumor resection and minimized small vessel and brainstem injury. Dissecting the tumor: As the surgery...
proceeded within the space provided by the progressive tumor debulking, the IDET was dissected and mobilized away from the facial, acoustic and trigeminal nerves, the brainstem, and blood vessels by gentle, meticulous microsurgical techniques. We used, at that point, fine micro scissors and several types of angled dissectors in addition to multiple microscope angulations at varying magnifications. As tumor debulking proceeded, the brainstem progressively relaxed and provided additional working space for dissection. When dealing with large lesions that compress and deform the brainstem, these tumors should be dissected from, and not against, the brainstem and CNs. The lesion was mobilized and, once rid of adhesions, removed from the surgical field with cupped forceps; small fragments were sucked away with the aid of irrigation. Surgeons should be aware that CNs and vessels are usually engulfed by the tumor; these delicate structures should be carefully dissected and spared. We maintained suction as low as possible during the microsurgical dissection to avoid injuries to those fragile structures. GTR is always attempted, but whenever dissection of the tumor from the basilar and vertebral arteries, their perforating branches, the brainstem, or CNs could entail risk of damage, we left a thin rim of tumor tissue attached to those structures.

During the operation, copious irrigation was provided and cotton pads were placed around the exposed area to reduce spillage of irritating cyst content into the subarachnoid space. Inspection of the tumor bed with the operating microscope is the then performed for verification of the extent of tumor resection. Some authors have recently proposed endoscope-assisted microsurgical resection for IDETs.\(^{[6,27]}\) However, that technique was not used in the current series.

Before closure, the patient’s blood pressure was brought to a normotensive level for at least 10-15 min and monitored for risk of oozing. The dura was closed primarily or with either a free pericranial graft or an artificial dural substitute. All open mastoid cells were sealed with bone wax to prevent CSF leaks. The superficial planes were closed in three layers using interrupted nylon sutures. Postoperatively, all patients were cared for in an intensive care unit before returning to the ward.

**Illustrative cases**

Patient 1: A 3-year-old girl presented with headaches and vomiting of 3-month duration. Her neurological examination revealed gait ataxia. An MRI scan detected a hypointense Vermian mass with expansion toward the fourth ventricle [Figure 2a]. A midline suboccipital craniotomy was performed and a pearly tumor with numerous hair follicles was identified [Figure 2b] and totally removed using standard microsurgical techniques [Figure 2c and d]. Postoperatively, transient dysphagia developed, which cleared completely after 2 months.

Patient 2: A 48-year-old male truck driver presented with a 2-year history of headaches and progressive left hemiparesis. MRI detected a large hypointense tumor located in the right Sylvian fissure with brainstem compression and a retrochiasmatic tumor expansion [Figure 3a]. After a pterional approach, nearly total tumor removal was obtained with decompression of the brainstem and removal of the retrochiasmatic portion of the tumor [Figure 3b-d]. The patient had an uneventful recovery.

Patient 3: A 68-year-old female developed progressive gait ataxia, diplopia, left-ear deafness, and left facial paresis of 2-year duration. An MRI scan showed a tumor located in the left-side CPA with brainstem distortion and contralateral shift [Figure 4a]. The patient was placed in a semi-sitting position and a retrosigmoid approach was achieved. The tumor occupied the left CPA entirely and engulfed most of the neurovascular structures [Figure 4b]. After STR, the seventh and eighth CNs were preserved and the brainstem, decompressed [Figure 4c]. Postoperative recovery was slow, but the patient eventually achieved a GOS 3. She died 3 months later due to pulmonary complications.

**RESULTS**

**Patient demographics and clinical characteristics**

Since this is a retrospective study, it has inherent biases and drawbacks that only a multicenter, prospective study could overcome. Thirty-three patients with IEDTs were identified, 29 epidermoids and 4 dermoids tumors. At the time of diagnosis, the patients’ average age was...
37.9 years, ranging between 1 and 78 years. In our study, 19 patients were female and 14 male. The neurological symptoms and signs varied according to tumor location and extension. Intracranial hypertension was observed in 9 (27.2%) patients. Gait disturbances were found in 8 (24.2%) patients, CN dysfunction was diagnosed in 9 (27.2%) individuals, and visual deficits in 6 (18.1%) patients. In our series, only three cases presented with trigeminal neuralgia.

A posterior fossa dermoid tumor associated to a pilonidal sinus, complicated by relapsing purulent meningitis and/or posterior fossa abscess was identified in three pediatric patients. But some authors consider this association as different entity.[3,10,15,24,26,34,35]

The demographics, neurological presentation, and results of the 33 patients analyzed in this study are summarized in Table 1.

### Neuroimaging

All patients underwent a CT and/or MRI scan. In general, the tumors appeared as low-density lesions on plain CT. MRI revealed that, in the majority of cases, the lesion was isointense to T1-weighted and hyperintense on T2-weighted images, similarly to the MRI appearance of arachnoid cysts. When available, the differential diagnosis was solved on inspection of the diffusion-weighted images, which showed epidermoids to be distinctly bright compared with arachnoid cysts and other tumors.[1,26] One tumor appeared as a high-intensity signal on T1 and as a mixed-intensity signal on T2. Ten (30%) lesions showed irregular enhancement after contrast injection. Two (6%) of them revealed calcifications [Figure 1a and b].

### Mortality, morbidity, extension of resection dilemma, and recurrence

There was no operative mortality until 30 days after surgery, but in the late postoperative period, three elderly patients with CPA lesions died from pulmonary complications. The follow-up period ranged from 1 to 21.5 years (mean, 7.2 years). Five patients were lost to follow-up. The dissection of the adherent capsule from the third, sixth, seventh, and eighth CNs led to palsies of the oculomotor, abducens and facial nerves. All five patients who developed facial paralyze, recovered. Two individuals with paralysis of the third nerve presented with partial recovery, other with sixth nerve did not improve. Permanent hearing loss occurred in three patients. Two pediatric patients with fourth ventricle tumors developed transient swallowing difficulty. Temporary postoperative complications, including wound infection, CSF fistulas that needed reoperation, deep venous thrombosis, and pulmonary complications, occurred in seven patients.

We achieved GTR in 23 (72.7%) patients and STR in 10 (27.3%), all confirmed by surgeon’s impression and/or postoperative imaging. Twenty-six (78.7%) patients achieved GOS 4 or 5.

### DISCUSSION

#### Patient demographics and clinical characteristics

We are in agreement with the authors who claim that epidermoid tumors occur more commonly in the fourth and fifth decades of life, whereas dermoid tumors are more frequent in children.[3,12,21-23,29] In the present sample, at the time of surgery, the patients’ mean age was 37.9 years, ranging between 1 and 78 years. In the literature, IDETs showed no definite gender predilection.[3,7,12,14,17,19,22,29] [Table 1].

Many studies[2,7,9,14-16,19,21,24,25,29,32,33] had come to share that the clinical symptoms varied among patients depending on tumor location, ranging from slight CN deficits to severe ataxia, CN dysfunction, and brainstem compression, as we observed in this series [Table 1].

#### Mortality, morbidity, extension of resection dilemma, and recurrence

Since the advent of microsurgical techniques, several series have shown that IDETs can be surgically resected with a good outcome.[5,10,15,17,24,26,34,35] Samii et al.[36] revealed that operative morbidity and mortality from IDET removal have declined remarkably in the last 20 years. Prior to the advent of the operating microscope, operative mortality ranged from 20% to 57%. Contemporary series have

| Table 1: Characteristics of 33 patients treated for IDETs |
|----------------|----------------|
| **Age at treatment:** 1-78 years (mean, 37.9 years) |
| **Female sex:** 19 (57.6%) |
| **Male sex:** 14 (42.4%) |
| **Location of the tumor** |
| CPA: 13 (39.3%) |
| Parasellar: 6 (18.1%) |
| Cerebellar: 5 (15.1%) |
| Temporal: 5 (15.1%) |
| Frontal: 4 (12.1%) |
| **Neurological presentation** |
| ICH: 9 (27.2%) |
| CN deficits: 9 (27.2%) |
| Gait disturbance: 8 (24.2%) |
| Seizure: 6 (18.1%) |
| Visual deficit: 6 (18.1%) |
| Motor deficit: 5 (15.1%) |
| **Pathology** |
| Epidermoid: 29 (87.5%) |
| Dermoid: 4 (12.4%) |
| GTR: 24 (72.7%) |
| Follow-up: 1-23 years (mean, 7.2 years) |
| Surgical mortality: Zero |
| Recurrence: 3 (9.0%) |
| GOS 4 or 5: 26 (78.7%) |

**Note:** IDETs: Intracranial dermoids and epidermoids tumors, CPA: Cerebellopontine angle, ICH: Intracranial hypertension, CN: Cranial nerve, GTR: Gross total tumor removal, GOS: Glasgow outcome scale
In our series, four CPA tumors and others advocate that the ideal management of IDETs is GTR. However, GTR could be dangerous for some patients and difficult to achieve because these tumors can be critically located, and there may be adherence or even encasement of vital structures by the tumor. Because IDETs “flow” into any available subarachnoid space, slowly increase their volume, and conform to the shape of the cavities they enter, CNs and arteries can be engulfed or displaced by the tumor. On diagnosis, many IDETs are already large or giant, and frequently have extended into multiple anatomic compartments. For these reasons, many authors recommend that GTR should be avoided to decrease mortality and morbidity. In our series, four CPA tumors spread along the cisterns of the posterior fossa and became partially trapped between the pons and the anteriorly displaced vertebralbasilar arteries, enveloping the perforating branches, and densely adhering to the brainstem. We performed a careful dissection at higher magnification to prevent injury of the fragile perforating vessels that were engulfed by the tumor. However, even a delicate dissection could entail risk of damaging these perforating vessels, which could provoke profound and permanent neurological disabilities. We cautiously left a rim of tumor attached to those structures (STR).

The best surgery for IDETs has been debated for a long time. The controversy is underlined by one major question: How far can we go with the intent of achieving GTR? On the one hand, if the surgeon persists with any attempt to remove every last residual lesion to achieve the cure of the patient, it could result in an unwarranted CN or arterial injury, thus increasing mortality and morbidity. On the other hand, experience clearly emphasizes that when tumors are removed incompletely, they tend to regrow after varying periods of time. As Sekhar and Wright clearly point out, the next surgeon operating on the patient will be confronted with severe adhesions of blood vessels and CNs to the brain, and with the inability to remove the lesion totally or nearly totally. Because IDETs are indolent, the risk of potential complications after IDET s resection, although they may occasionally occur spontaneously. In some series, this entity achieves more than 22%. To prevent this complication, it seems that excision of the capsule by sharp dissection and “copious irrigation” in the surgical field plus the use of IV dexamethasone intra- and postoperative reduced the incidence of chemical meningitis. Moreover, some authors proposed irrigation with hydrocortisone during the surgery, to also provide protection against chemical meningitis. We routinely used the aforementioned protocol and added cotton pads placed around the exposure to reduce spillage of irritating cyst content in the subarachnoid space. In our experience, aseptic meningitis was not a problem. We noticed this in only two patients. Another rare complication can be a malignant transformation of previous epidermoid tumor to squamous cell carcinoma. This situation should be considered when follow-up MRI show contrast enhancement at the surgical site, and/or whose condition deteriorates. In the literature, the interval from first operation to malignant transformation ranged from 6 months to 33 years. In these cases, treatment options include reoperation with adjuvant chemotherapy or radiotherapy.

Table 2: Contemporary surgical series on dermoid and epidermoid tumors (IDETs)

| Authors/years | N° cases | MORT. (%) | GTR. (%) | REC. (%) | F/U (years) |
|---------------|----------|-----------|----------|----------|-------------|
| Berguer et al.[3] /1985 | 13 | 0 | 0 | 7.6 | 4.6 |
| Sabin et al.[24] /1987 | 20 | 5 | 5 | 10 | 6 |
| De Souza et al.[27] /1989 | 30 | 3.7 | 18 | 14.8 | 9 |
| Rubin et al.[31] /1989 | 7 | 0 | 57 | 0 | 4.6 |
| Yamakawa et al.[23] /1989 | 15 | 6.6 | 47 | 20 | 8 |
| Yasargil et al.[34] /1989 | 43 | 0 | 95.4 | 0 | 5.2 |
| Lunardi et al.[16] /1990 | 17 | 12 | 35 | 17.6 | 9 |
| Gormley et al.[19] /1994 | 32 | 5 | 42 | 26 | N/A |
| Vinchon et al.[32] /1995 | 19 | 22.2 | 0 | N/A | 3 |
| Samii et al.[36] /1996 | 40 | 2.5 | 75 | 7.5 | 5.7 |
| Mohanty et al.[15] /1996 | 25 | 8 | 48 | 0 | 3.5 |
| Talachchi et al.[29] /1998 | 28 | 3.5 | 57 | 30 | 8.6 |
| Kobata et al.[15] /2002 | 30 | 0 | 56.7 | 6.6 | 11.4 |
| Chowdhury et al.[4] /2013 | 23 | 4.3 | 73.9 | N/A | 3 |
| Kata et al.[13] /2013 | 27 | 5 | 10 | 20 | N/A |

MORT: Mortality; GTR: Gross total removal; REC: Recurrence; F/U: Follow-up; N/A: Not available

We did not observe a single case of malignant transformation in this group of patients.

In this current series, there was no surgical mortality.

Published papers on IDETs have reported GTR rates ranging from 0% to 95.4%. In the current series, we achieved GTR in 72.7% of the patients. Our surgical objective was always to prioritize the patient’s quality of life; therefore, STR could represent a very acceptable goal in tumors encasing the basilar artery, the vertebral artery, or perforating vessels, or adhering to CNs. Previous studies with long-term follow-up have reported an overall estimated recurrence rate ranging from 0% to 26%. In the present series, the recurrence rate was 9% (three cases).
CONCLUSIONS

The neurosurgical management of IDETs remains controversial, but our results and literature data allow us to conclude that the best surgical strategy is GTR, using meticulous microsurgery techniques to obtain the cure of the patient. We achieved GTR in the majority of our patients without mortality and with low morbidity. However, GTR can be difficult to achieve because IDETs can be critically located, with adherence to, and involvement of, vital structures. Our surgical objective was always to prioritize the patient’s quality of life. Subtotal removal could represent a very acceptable goal in tumors encasing the basilar artery and perforating vessels or adhering to CNs. If the functional risk of GTR outweighs its potential benefits, an STR strategy should be adopted.

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