



Long term outcome in surgically treated posterior fossa epidermoids



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ABSTRACT

Objectives: To study posterior fossa epidermoids treated surgically at our institute and to compare their long term outcome with respect to extent of surgical decompression.

Materials and methods: Retrospective analysis of 50 cases of posterior fossa epidermoid surgically treated at our institute between 1997 and 2007.

Results: The mean duration from onset of symptoms to surgery was 2.5 years. Patients with cerebello-pontine angle (CPA) epidermoids presented predominantly with trigeminal neuralgia (35%) and hearing loss (29%) while patients with fourth ventricle epidermoids had features of raised intracranial pressure (ICP) and gait ataxia (69.2% each). The rate of recurrence was 9% in tumors considered totally removed and 93% in those subtotally removed. Of the 17 patients with recurrences, 3 (7.9%) underwent a second operation. The mean duration of follow up at first recurrence was 9.3 years.

Conclusion: Based on our experience, the rate of recurrence is significantly higher after subtotal removal as compared to total removal of epidermoids on long-term follow up. Symptomatic recurrence requiring re-exploration is evident only after a long duration (~10.9 year) following primary surgery. Hence, total removal without producing new neurological deficits should be the standard goal when operating on posterior fossa epidermoid cysts.

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1. Introduction

Epidermoid tumors account for around 1% of intracranial tumors and are known to occur at diverse locations in the neuraxis [1]. Posterior fossa epidermoids comprise a distinct entity since they insinuate across cisternal spaces, intimately involving multiple cranial nerves and vascular structures, thus making them a difficult tumor to completely resect. Attempts at total excision of the tumor capsule are often associated with significant morbidity in the postoperative period. On the other hand, partial removal risks recurrence and subsequent surgery [2]. The decision to radically remove these cysts must balance the risks of surgical complications vs the benefits gained.

We summarize our 11-year experience of surgically treated posterior fossa epidermoid tumors with the aim of reaching a consensus on the extent of tumor excision without compromising on postoperative clinical outcome and long term tumor recurrence.

2. Materials and methods

Retrospective analysis of 50 cases of posterior fossa epidermoid surgically treated at our institute between 1997 and 2007 was done. Data were collected regarding demographics, presenting signs and symptoms, management strategy, postoperative neurological complications and long term follow-up. Posterior fossa epidermoids were broadly classified based on their location into 2 groups: cerebellopontine angle (CPA) and fourth ventricle. CPA epidermoids were further sub-classified based on their extent (Table 1) [3]. Our primary aim during surgery was complete tumor removal without compromising neurovascular structures. The surgical approaches depended on the location of the main tumor which would often provide the gateway for reaching contiguous anatomic sites of tumor extension (Table 1).

Extent of tumor removal was determined based on intra-operative assessment and postoperative images. It was broadly divided into total (Group A) and subtotal (Group B) excision, the latter being re-classified as two subgroups: 'B1', residual lesion due to inaccessibility via the operative corridor chosen and 'B2', residual capsule adherent to critical neurovascular structures. Extent of removal was considered total if keratinous debris from the tumor and the entire tumor capsule were removed. The functional status at the time of discharge and at subsequent follow up was determined using the Modified Rankins Score (mRS). All

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Table 1
Tumor location, extension and their extent of removal with various surgical approach. CPA cerebellopontine angle, CA combined approach, FM foramen magnum, MCF middle cranial fossa, MFA middle fossa approach, MSOC midline suboccipital craniotomy, RMSOC retromastoid suboccipital craniotomy.

Tumor location	No of patient	Surgical approach	Total removal (A)	Subtotal removal (B1 + B2)	Residual lesion (B1)	Residual adherent capsule (B2)
CPA alone	12	RMSOC	9	3	0	3
CPA + transtentorial extension	10	RMSOC	6	4	2	2
CPA + MCF extension	7	2 MFA + 2 CA + 3 RMSOC	2 + 2 + 1	2	1	1
CPA + FM extension	3	RMSOC	1	2	0	2
CPA + transtentorial & FM extension	5	RMSOC	1	4	3	1
Fourth ventricle	13	MSOC	9	4	0	4
Total	50	–	31	19	6	13

patients underwent radiological follow-up studies immediately after surgery and at 1–2 years, depending on presence/absence of residual lesion.

All patients who had undergone surgical excision were sent a request letter to come for review at neurosurgery outpatient department. Long-term follow-up, both clinical and radiological, was analyzed and a Kaplan–Meier recurrence-free survival curve was plotted. The rate of recurrence and the need for re-exploration in total and sub totally excised tumors were compared.

3. Results

3.1. Patient demographics and clinical characteristics

Fifty patients presented with a diagnosis of posterior fossa epidermoids. All these patients were treated surgically and are evaluated in this review. There were 28 (56%) females and the age ranged between 19 and 66 years (mean 37.8 years). Clinical features at presentation depended on the tumor location as well as its extension. Patients with CPA epidermoids presented predominantly with trigeminal neuralgia, hearing loss and gait disturbance while those with lesions in the fourth ventricle had features of raised intracranial pressure (ICP) and gait ataxia (Table 2). The interval between onset of symptoms and diagnosis ranged between 1 month and 120 months (median 30 months).

Table 2
Presenting symptoms of CPA and fourth ventricle epidermoids CPA cerebellopontine angle.

Symptoms	CPA epidermoid n = 37 (%)	Fourth ventricle epidermoid n = 13 (%)
Gait disturbance	9 (24.3)	9 (69.2)
Trigeminal neuralgia	13 (35)	0
Hearing loss	11 (29.7)	1 (7.6)
Facial numbness	7 (18.9)	0
Cerebelar signs (e.g. dysmetria, nystagmus, gait ataxia)	7 (18.9)	8 (61.5)
Headache	6 (16.2)	9 (69.2)
Facial weakness	5 (13.5)	3 (23.1)
Diplopia	5 (13.5)	4 (30.1)
Lower cranial related symptoms	5 (13.5)	3 (23.1)
Hemifacial spasms	4 (10.8)	0
Hemiparesis	4 (10.8)	2 (15.4)
Seizure	2 (5.4)	0
Vision deterioration	1 (2.7)	0
Hemisensory anesthesia	1 (2.7)	0

4. Radiographic features

Computed tomography (CT) revealed all tumors to be hypodense with respect to brain tissue. A small area of peripheral calcification was demonstrated in 5 cases (10%), whereas subtle capsule-like enhancement was observed in 4 (8%). All patients were evaluated with magnetic resonance imaging (MRI) prior to surgery (Figs. 1 and 2). Epidermoids are iso- to hypointense on T1-weighted (T1W) and hyperintense on T2-weighted (T2W) images in comparison to cerebrospinal fluid (CSF) and brain tissue. Diffusion weighted images (DWI) were available in 40 patients, and it showed restricted diffusion in tumor. The location of the tumors based on imaging is given in Table 1.

5. Operative procedure

Standard operative approaches to CPA and fourth ventricle were used to achieve maximum tumor removal (Table 1). After durotomy, the tumor was visualized as a white, often cheesy mass filling the CPA or the 4th ventricle. After intracapsular tumor removal, extracapsular dissection was attempted to remove the capsule keeping in mind the adherent neurovascular structures. Often this necessitated incomplete removal of the capsule to preserve the integrity of the cranial nerves and blood vessels embedded in the lesion. In 11 (44%) cases of CPA tumor with extension to other anatomic sites, the entire tumor could be resected by retromastoid suboccipital craniotomy (RMSOC) alone. While, in the remaining 14 tumors, it was observed that a part of the capsule had to be left in 5 and a subtotal removal had been done in the rest due to inaccessible areas of tumor. 4 cases of CPA epidermoids with extension needed other avenues which included a middle cranial fossa approach in 2 (8%) cases and a combined subtemporal and retromastoid approach (combined approach) in the remaining 2 patients. This was deemed necessary as it was felt that the predominant component was placed anteriorly and the CPA component could be removed after splitting the tentorium and drilling the petrous apex. All fourth ventricle tumors were removed by a midline suboccipital craniotomy (MSOC). Most of the epidermoid cysts in this location could be completely excised except in 4 cases where the capsule was adherent to the fourth ventricle floor.

6. Complications

There was significant improvement in trigeminal and lower cranial nerve dysfunction especially after surgery as shown in Table 3. This improvement was not noticeable in patients who had preoperative facial weakness. New onset facial nerve palsy after removal of posterior fossa epidermoid was observed in 2 patients. 2 patients had lower cranial dysfunction in the immediate postoperative period necessitating tracheostomy in one patient and gradual weaning from ventilator support in the other.

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