Original Article (BRAIN)

Incidence and Surgical Outcome of the Intracranial Epidermoid Cyst at Punjab Institute of Neurosciences Lahore, Pakistan

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ABSTRACT

Objectives: The incidence and microsurgical outcomes of intracranial epidermoid cysts in the Department of Neurosurgery III, Punjab Institute of Neurosciences (PINS), Lahore, Pakistan, are described in this case series.

Materials and Methods: This study was a data analysis of a case series of 15 patients (mean age, 40 years) of both gender with intracranial epidermoid cysts who had microsurgical surgical excision over five years.

Results: This study comprised 11 (73.3%) male and 4 (26.7%) female patients, 11 (73.3%) cases were infratentorial and 4 (26.7%) cases were in supratentorial region. The epidermoid was located in the CP angle in 11 (73.3%) patients, 3 (20%) in the midline supra sellar region, and 1 (6.66%) in the frontotemporal region. The presenting complaints were mainly headache in 11 (73.33%), cranial nerve palsy and cerebellar signs in 8 (53.3%) patients, Trigeminal neuralgia in 3 (20%) patients, Fits and hydrocephalus in 2 (13.3%) patients. There were 14 (93.3%) patients with GTR (gross total resection), 1 (6.6%) patients STR (subtotal resection). According to Karnofsky’s performance scoring (KPS), 3 (20%) patients improved, 11 (73.3%) patients had the same KPS, and 1 (6.6%) patient had a lower KPS.

Conclusion: The epidermoid cysts in the brain are usually found in the infratentorial region rather than the supratentorial region. Infratentorial lesions typically cause cranial nerve deficits, whereas the supratentorial area symptom is a headache.

Keywords: Cranial Nerves, Epidermoid Cyst, Cerebellopontine Angle.

INTRODUCTION

The intracranial epidermoid cyst is a congenital lesion developed by the migration of ectodermal cells after neural tube closure at the 3rd to 5th weeks of pregnancy. Epidermoid cysts are common in the 3rd and 4th decades of life. Men are more likely than women to get cysts. Intracranial epidermoid cysts have well-defined borders. It’s an uncommon tumor that accounts for 0.2–1.8% of all brain tumors. Mostly in the
The cerebellopontine angle (CPA, 40%), supra sellar cisterns (18%), cerebral, cerebellar hemispheres, lateral, Sylvian fissure, and fourth ventricles. The cerebellopontine angle (CPA) is the most frequent intracranial place for an epidermoid cyst (EC), accounting for 5–7% of all malignancies in this region after schwannomas and meningiomas. CPA epidermoid cysts are in the subarachnoid space or cisterns at the base of the skull. Most go unnoticed for years, which is why it is common to find substantial tumor infiltration at the time of diagnosis. Epidermoids of the CPA are difficult to completely resect due to their dispersed development pattern. There have been reports of surgical morbidity differences based on the extent of the lesion. Despite the need for total tumor removal to prevent a recurrence, total removal may not be achieved without causing serious deficits. These tumors contain soft keratinous desquamated epidermal cell debris that can be aspirated, the tumor capsule is sometimes tightly attached to important structures like the cranial nerves, brainstem, and blood vessels, and resection of this capsule can result in serious complications. The lesions proliferate while adhering to these vital structures, especially in CPA angle, and removing the capsule raises the risk of complications. Even if the contents tumor are completely removed during surgery, the likelihood of recurrence is significant if the tumor capsule is not excised. The tumor removal rate at the time of reoperation will be very low than the initial operation, and the most serious issue is that radiotherapy is ineffective for residual or recurrent epidermoid cysts, unlike meningiomas and schwannomas. As a result, the optimal treatment is extensive tumor resection, including the capsule, which allows for long-term tumor control while avoiding chronic neurological damage. We believe that removing CP angle epidermoid cysts under direct observation is critical for both drastic elimination and preservation of cranial nerve functioning. As a result, on the tumor size and extent, we choose the best surgical strategy for each patient in our department. The surgical results of 11 patients with CPA epidermoid cysts and four individuals with supratentorial epidermoid cysts in our institution are presented here. This study would provide an insight into the incidence of Intracranial epidermoid cysts and surgical outcomes at our institution.

MATERIALS AND METHODS

Study Design

This is the retrospective data analysis of case series of 15 patients (mean age, 40 years) with intracranial epidermoid cysts who underwent microsurgical removal over 5 years at the Punjab Institute of Neurosciences Unit 3 between January 2017 to January 2022.

Inclusion Criteria

All patients of both genders (mean age, 40 years) with intracranial epidermoid cyst who underwent microsurgical removal over 5 years at the Punjab Institute of Neurosciences Unit 3 between January 2017 to January 2022.

Exclusion Criteria

Pediatric population.

Surgical/Clinical Management & Data Collection

Between January 2017 and January 2022, 15 patients with cerebral epidermoid cysts (11 men and 4 females) with an average age of 40 years had microsurgical excision at the Punjab Institute of Neurosciences Unit 3. The signs and symptoms of presentation, the extent of surgical excision, post-op complications, and recurrence were all investigated. Pre-op imaging included CT scan and MRI with axial, coronal, and sagittal cuts in T1- and T2-weighted images, and the majority of
FLAIR and DWI sequences in all patients. The technique used was chosen by the tumor's location. The extent of resection was assessed through postoperative imaging. Total excision was defined as the complete removal of the lesion, sub-total excision as the removal of sections of the capsule adhering to essential neurovascular structures, and partial excision as the removal of more than 20% of the tumor. During the outpatient session, clinical follow-up was assessed. Every six months, then annually, a brain MRI was utilized to assess the occurrence of clinical and imaging recurrence.

RESULTS

Age and Gender Distribution
The research comprised a total of 15 patients with mean age, of 40 years and a range of 20 to 55 years. There was a male majority, with males accounting for 11 (73.3%) of patients and females accounting for 4 (26.7%).

Location of Tumors
With 11 (73.3%) cases being infratentorial and 4 (26.7%) instances being supratentorial. The epidermoid cysts were located in the CP Angle in 11 (73.3%) patients, 3 (20%) in the midline suprasellar, and 1 (6.66%) in the frontotemporal region (Table 1).

| Table 1: Location. |
|---------------------|
| Infratentorial      | 11     (73.3%) |
| Cerebellopontine Angle | 11     (73.3%) |
| Supratentorial      | 4      (26.7%) |
| Midline suprasellar | 3      (20%) |
| Fronto-Temporal region | 1      (6.66%) |

Presenting Complaints
The presenting complaints were mainly headache in 11 (73.33%), cranial nerve palsy and cerebellar signs in 8 (53.3%) patients, and Trigeminal neuralgia in 3 (20%) patients, Fits and hydrocephalus in 2 (13.3%) patients (Table 2).

| Table 2: Presenting Complaints. |
|----------------------------------|
| Complaints                      | No. of Patients | Percentages |
| Headache                        | 11              | (73.3%)     |
| Cranial nerve palsy             | 8               | (53.3%)     |
| Cerebellar signs                | 8               | (53.3%)     |
| Trigeminal neuralgia            | 3               | (20%)       |
| Fits                            | 2               | (13.3%)     |
| Hydrocephalus                   | 2               | (13.3%)     |

Surgical Resection
There were 14 (93.3%) patients with GTR (gross total resection), 1 (6.6%) patients STR (subtotal resection) (Table 3).

| Table 3: Tumor Resection. |
|---------------------------|
| Tumor Resection           | No of Patients | Percentages |
| Gross Total               | 14             | (93.3%)     |
| Sub-Total                 | 1              | (6.6%)      |

Surgical Outcome (Karnofsky Performance Scoring)
Based on pre-and post-operative Karnofsky performance scoring, 3 (20%) patients improved, 11 (73.3%) patients had similar KPS, and 1 (6.6%) patient had a lower KPS (Table 4). After surgery, one case encountered complications.

| Table 4: Karnofsky's performance scoring. |
|-------------------------------------------|
| Post Op KPS Status                        | No. of Patients | Percentage |
| Improved                                  | 3              | (20%)      |
| Similar                                   | 11             | (73.3%)    |
| Decreased                                 | 1              | (6.6%)     |
Fig 1(A): Pre-op MRI.
Figure 1(B): Post-op CT Scan.

Figure 1: Infratentorial (CPA) Epidermoid Cyst (Images used with permission).

Figure 2(A): Pre-op MRI.
DISCUSSION

An epidermoid cyst is a benign brain tumor that occurs between the 3rd to 5th weeks of embryonic life as a result of aberrant entrapment of ectodermal cells, which eventually grow into the epidermis, in the nerve tissue after neural tube closure. The otic and optic vesicles are created at the same time as the CPA and the parasellar area, and the ectodermal cells inside these structures result in the most common intracranial location of epidermoids.\(^{15,16}\) Cysts in the fourth ventricle are thought to be the result of ectodermal cell misplacement before neural tube closure, while cysts epidurally or inside the diploe occur after neural tube closure.\(^ {16}\) Epidermoid cysts account for 1% of all brain cancers, according to estimates.\(^ {12}\) Almost half of them arise in the CP angle, the third most common brain tumor after schwannomas and meningiomas, accounting for 5–7% of tumors in this area.\(^ {16}\) In this study, 11 (73.3%) of the instances were infratentorial, whereas 4 (26.7%) were in the supratentorial region. In 11 (73.3%) of the patients, the epidermoid was seen in the CPA (cerebellopontine angle), 3 (20%) in the midline suprasellar region, and 1 (6.66%) in the frontotemporal region. Hearing loss, trigeminal neuralgia, facial palsy, diplopia, migraines, seizures, and altered mental status are common symptoms of these slow-growing cysts.\(^ {18}\)

Headache was the most common presenting complaint in 11 (73.33%) patients, followed by cranial nerve palsy and cerebellar abnormalities in 8 (53.3%) people, trigeminal neuralgia in 3 (20%) patients, and fits and hydrocephalus in 2 (13.3%) patients (Table 2). The goal of surgical treatment was to remove the tumor and its capsule completely. Small portions of the capsule were left in places where it was difficult to separate the capsule from nerves, arteries, or the brainstem to
prevent the possibility of major neurological abnormalities from unintended damage to these structures. There were 14 (93.3%) patients with GTR (gross total resection) and 1 (6.6%) patient with STR in our study (subtotal resection) (Table 3). While individuals with epidermoid tumors have a decent long-term survival rate, their slow development pattern necessitates early detection and identification. According to pre-operative and post-operative Karnofsky performance scoring (KPS), 3 (20%) patients improved, 11 (73.3%) patients had similar KPS, and 1 (6.6%) patient had a lower KPS (Table 4). Unlike more common liquid cysts, which can be removed with a less invasive operation, epidermoid tumor removal necessitates extensive exposure to allow for capsular resection when possible. Capsular resection reduces regrowth and malignant transformation. One (6.6%) patient in our study underwent STR (subtotal resection) for a suprasellar midline epidermoid cyst due to the closeness of critical structures and vascular encasement. This becomes a big concern in persons whose original surgery was performed when they were younger because it tends to return after many years or decades.

CONCLUSION
The epidermoid cysts in the brain are benign congenital brain tumors that are mostly found in the infratentorial than supratentorial regions. The cerebellopontine angle is the most common intracranial site for epidermoids. Infratentorial lesions typically cause cranial nerve deficits, whereas the supratentorial area symptom is a headache. It’s reasonable to suspect an epidermoid cyst if the patient has basal cranial nerve palsies. The surgical goal is to remove the tumor and its capsule completely. As separating the capsule from nerves, vessels, or the brainstem is difficult, small portions of the capsule have to be left to prevent neurological complications from unintended damage to these structures, which tends to recur after many years.

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Additional Information

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Human Subjects: Consent was obtained by all patients/participants in this study.
Conflicts of Interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following:
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Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHOR CONTRIBUTIONS

| Sr. No. | Author’s Full Name          | Intellectual Contribution to Paper in Terms of                                      |
|---------|----------------------------|------------------------------------------------------------------------------------|
| 1       | Tariq Imran Khokhar         | Study design, methodology, and paper writing.                                      |
| 2       | Muhammad Naveed Majeed      | Data calculation and data analysis.                                                 |
| 3       | Khawar Anwar                | Interpretation of results.                                                         |
| 4       | Ali Faizan Bukhari          | Literature review.                                                                |
| 5       | Asif Bashir                 | Literature review and quality insurer.                                             |