ORIGINAL ARTICLE

Neurosurgical Management of Intracranial Epidermoid

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ABSTRACT

Objective: *Epidermoid cyst or cholesteatoma is a congenital slow growing lesion. The objective was to analyze clinical presentation surgery and outcome after surgery.*

Study Design: *Retrospective observational study.*

Materials and Methods: *This study represents a retrospective review of patients treated with epidermoid cyst, during a period of 18 years between 1995 to 2012. It was conducted at the department of Neurosurgery Sheikh Zayed Hospital, Rahim Yar Khan and department of Neurosurgery PGMI / LGH Lahore and department of Neurosurgery AMDC Farooq Hospital, Lahore.*

Results: *The age of our patients ranged from 18 to 40 years, with male predominance. The predominating symptoms were related to the 5^{th}, 7^{th} and 8^{th} cranial nerve and headache. Our study included 10 cases of epidermoid cysts; four of them were in CPA area, two suprasellar region and one was at pineal region. It was not always possible to determine if the signs and symptoms were due to local involvement by the epidermoids, increased intracranial pressure or both. The age of our patients ranged from 18 to 40 years, with male predominance. The retrosigmoid approach was used in 5 patients, ® Frontal craniotomy in 2 cases, Rt interhemispheric transtentorial approach in one patient and sub-temporal approach in one case. Parental portaline approach in 1 case. Total resection of epidermoid cyst was accomplished in 7 cases. To minimize recurrence, the residual epidermoid was carefully coagulated with the aid of microscope and bipolar diathermy without damaging surrounding neurovascular structures.*

Conclusion: *Surgical management of intracranial dermoid has encouraging and good results. Use of endoscope is good adjunct for complete removal of epidermoid from angles and corners of the lesion.*

Key Words: *Epidermoid, cerebellopontine angle, trigeminal neuralgia, Pineal region.*

Abbreviations: *CPA: Epidermoid, cerebellopontine angle, TN: trigeminal neuralgia, PR: Pineal region.*

INTRODUCTION

Intracranial epidermoids are rare, slow growing, benign that may arise from retained ectodermal inplants. Epidermoid cyst are the rare among all the intracranial neoplasm and incidence is 1% of all intracranial tumors.[1] The CPA is one of the most common sites affected. It can cause irritation of the cranial nerve, resulting in cranial nerve hyperactive dysfunction, such as trigeminal neuralgia, hemifacial spasm and may present symptoms of cranial nerves, cerebellar and brain stem dysfunction, parinaud syndrome and hydrocephalus.[2] Surgical treatment may show good resolution of presenting symptoms and signs.[3]

We retrospectively reviewed and analyzed the clinical characteristics of patients with trigeminal neuralgia due to CPA epidermoids. Intracranial epidermoid are slow growing benign tumors which contains keratin, cellular debris, cholesterol and lined with stratified squamous epithelium. Due to rarity of epidermoids presenting as TN: trigeminal neuralgia, the most of the cases reported are limited to case reports or short series.[4,9,11,12] Hearing impairment has been repo-

rted in epidermoid cyst in CPA, followed by TN, facial palsy, headache, vertigo, dizziness and diplopia. However, difficult listening over the telephone as sole manifestation of hearing impairment is rare.[11]

MATERIALS AND METHODS

This study represents a retrospective review of patients treated with epidermoid cyst, during a period of 18 years between 1995 to 2012. It was conducted at the department of Neurosurgery Sheikh Zayed Hospital, Rahim Yar Khan and department of Neurosurgery PGMI / LGH Lahore and department of Neurosurgery AMDC Farooq Hospital, Lahore.

Data were retrospectively collected from clinical, surgical, pathological and neuroradiological records.

RESULTS

Sex Incidence

There were 3 (30%) females and 7 (70%) male as shown in table 1.

Table 1: *Sex Incidence.*

Sex	Number	Percentage
Male	7	70
Female	3	30
Total	10	100

Age Incidence

Patients had age between 18 to 40 year (mean 30.5 years).

Table 2: *Age Incidence.*

Age	Number	Percentage
1 – 10	–	–
11 – 20	1	10
21 – 30	5	50
31 – 40	4	40
Total	10	100

Clinical Features

The mean preoperative duration of symptoms was 4.75 years. Five of 10 patients presented with trigeminal neuralgia, impairment of hearing, tinnitus facial numbers and 3 patients presented gait disturbance, blurring of vision and parinaud syndrome.

Compared to patients with classical TN due to vascular causes, epidermoid patients present at a younger age. Each patient had a prior history of preoperative medical treatment (using carbamazepine) and symptoms relief was transient or incomplete. None of the patient had undergone a prior surgical procedure for trigeminal neuralgia.

All patients were preoperatively evaluated for MRI. On MR studies mostly displayed hypointensity on T_1 weighted images, hyperintensity on T_2 weighted images. With the use of MR imaging, correct diagnosis of epidermoid was possible preoperatively in 9 cases.

Operation

A lateral sub-occipital approach through retrosigmoid was performed in 5 patients (one has also cyst at subtemporal region along with CP angle. In second stage sub-temporal approach was used later on) and one patient had right interhemispheric transtantorial approach for pineal region epidermoid cyst other approaches are in table 3. After opening the dura, the arachnoid was divided and tumor capsule was exposed. Its remarkable white and pearly appearance made immediate confirmation of epidermoid cyst. In all cases of CPA epidermoid, the trigeminal nerves were completely encased and distorted by the tumor without displacement: in one patient the nerve was compressed by both the tumor and anterior inferior cerebellar artery (AICA), on the same side. In this, patient, MVD of the trigeminal nerve was performed in addition to tumor removal. During operation irrigation and cotton pads were used to reduce spillage of the irritating cyst content into subarachnoid space. In all patient total resection of epidermoid content, and radical resection of the cyst capsule. Was done. All patients showed immediate relief of TN after surgery. Facial numbness and hearing disturbance did not show any evidence of improvement but disturbance in gait gradually improved. In one patient with midline post fossa craniectomy and removal of epidermoid compresses the 4^{th} ventrile, brain stem having 5^{th}, 6^{th}, 7^{th}, 8^{th}, 9^{th}, 10^{th}, 11^{th} nerves involvement, 5^{th}, 9^{th}, 10^{th} nerve recovered all other had partial recovery only.

Follow-up was 1–15 years 50% cases best follow-up after 10 years. During follow-up period of seven

Table 3: *Hospital demographic data, site, operation, outcome and HP.*

		Demographic Data						Follow-up	
Hospital	**Cases**	**Age in Years**	**Sex**	**Site**	**Operational**	**Immediate Outcome**	**Complication**	**3 Months**	**1 Year**
RYK	4	18	Male	CPA and subtemporal	CPA and 2nd ST approach	Good		Good	Good
		40	Female	Pineal regin	Interhemispheric transtentorial	Good		Good	Good
		30	Male	CPA	CPA approach	Excellent		Excellent	Excellent
		34	Male	CPA	CPA approach	Good	Recurrence at 9 year	Good	Fair
LGH Lahore	3	32	Male	Partial parafalcin	Interhemispheric parafalcin	Excellent	–	Excellent	Excellent
		25	Male	Pineal regin	Infratentorial supracerebellar	Good	–	Good	Excellent
		35	Male	CPA regin	CPA craniectomy	Good	–	Excellent	Excellent
Farooq Hospital Allama Iqbal Town Lahore	3	25	Male	® Frontal nerve frontal are sinus	® Frontal craniectomy	Excellent		Excellent	Excellent
		30	Male	Parasellar	Frontal craniectomy	Excellent		Good	Good
		38	Female	Midline CPA Brain stem with ventricle CPA	Midline craniectomy	Fair		Fair (Partial recovery of patients)	Fair
Total	10				CPA 5 cases Midline suboccipital craniectomy 2 cases Frontal cranioctomy 2 cases Parafalcin 2 cases ST cranioctomy 1 case			Excellent 4 cases	Excellent 5 cases
								Good 5 cases	Good 3 cases
								Fair 1 case	Fair 2 cases

*ST = Subtemporal approach. CPA = Cerebellopontine angle approach.

years, one patient has recurrence of epidermoid cyst and it was again re-explored and operated. No patients develop any sign of meningitis.

DISCUSSION

Complications

One patient had recurrence of CPA lesion at 4 year. He was operated again. One patient with epidermoid

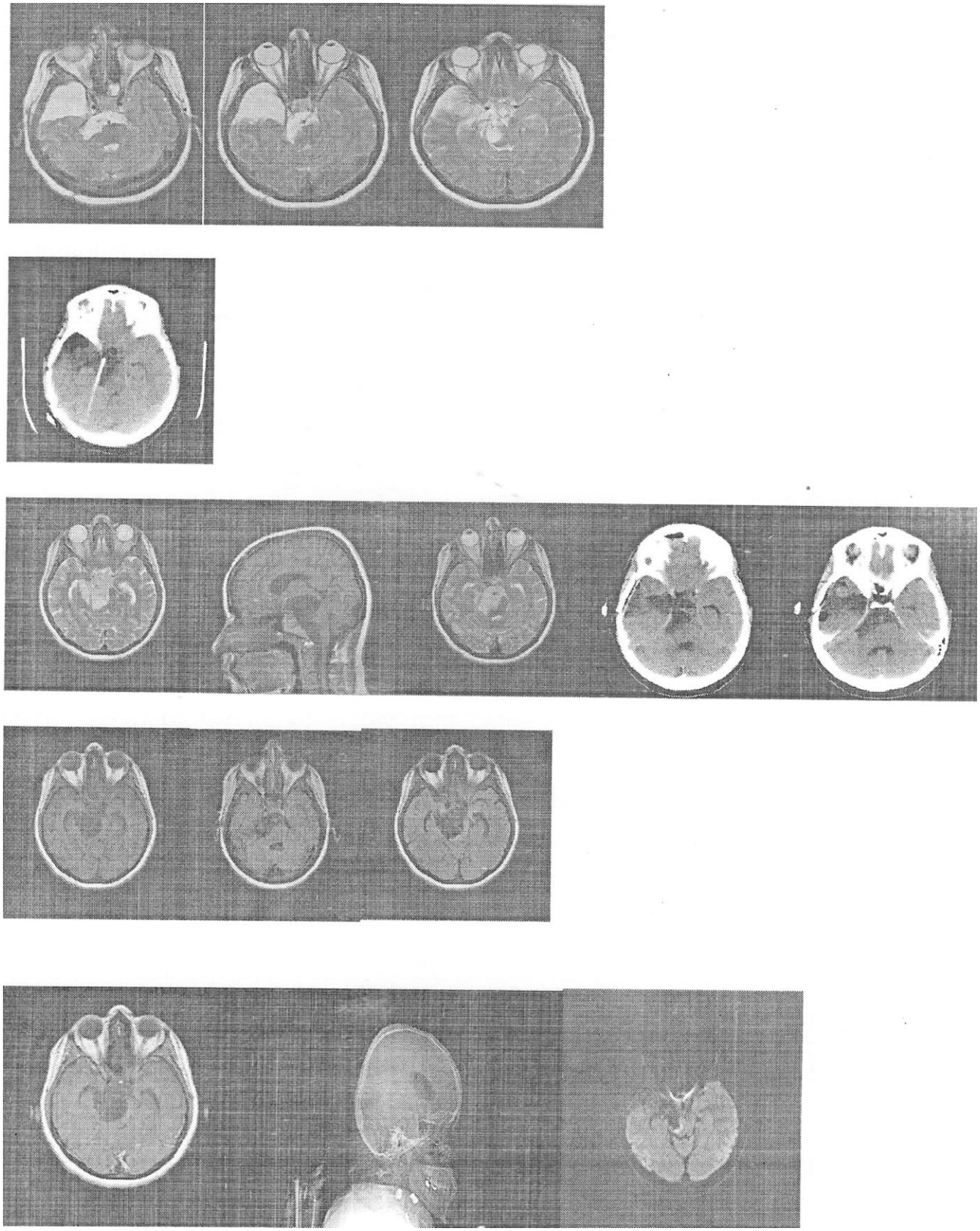

Fig. 1: *CT Scan Preoperative and Postoperative of Patient.*

improving the 4th ventrile and brain stem with pre-operative involvement of 5th, 6th, 7th, 8th, 9th, 10th nerves had recovery of 5th, 10th, 11th nerves while there was only partial recovery of remaining nerves 6th, 7th, 8th, 9th nerves. There was multisuperficial wound injection – one case which recovered with antibiotics.

Gracek argued that epidermoids originate from epithelial cells from vessels pouch while other state that their capsules consist of a thickening of aracnriod mater and develop within it. The cyst have also been postulated to be neuro-ectodermal in origin. None of our patients had history of trauma while epidermoids in the ear are most often infected, those of CPA are usually non-infected. Both behave in the similar manner by expanding and eroding bone.

Epidermoid cyst represent 0.2 to 1.4% of all primary intracranial tumor. CPA epidermoids constitute about 40% of all intracranial epidermoids.

Incidence of trigeminal neuralgia (TN) in patients with CPA epidermoids has been reported to vary from 0 to 90.6%. In our study 5 out of 6 patients with CPA epidermoid were associated with TN. Epidermoid cysts have extremely slow linear growth rate. Therefore, symptoms were long lasting, patients often presented relatively early when the predominant symptoms was trigeminal neuralgia. The symptoms and signs are caused by displacement of adjacent neurovascular structure. It is reported that hearing loss is most common symptoms in patients with epidermoids which was 37.6% followed by TN, (29.7%), dizziness or vertigo 9.4%, facial palsy (19.4%), headache (17.9%) and diplopia, 16.7%.[11,12]

Baker et al, and Jannetta reported that patients with cyst as the cause of trigeminal neuralgia (TN) have a clinical history that is not different from patients with classical manifestation of TN. But the occurrence of TN at a younger age and the long duration of symptoms are characteristic of TN patients with epidermoids, in contrast with TN due to vascular cause. In younger patients, with TN, therefore, the neuroimaging study must be performed carefully to avoid overlooking a small cyst in the CPA cistern.

The pathogenetic mechanism of pain in patients with CPA epidermoids remain uncertain. Direct compression of the nerve at the root entry zone, displacement of the trigeminal nerve, and compression against a blood vessel, at the root entry zone, or the combination of the two, have been postulated to be the cause. The keratin contents may directly cause irritation and hyperactivity of the nerve without an intervening vascular loop.[12]

However, the peculiar growth feature along with the wide extension of the tumor and its firm adhesion to critical neurovascular structures make total surgical removal not always possible nor advisable. We agree that the ideal goal in surgery for epidermoids is total removal, but not at the expense of neurological deficits. It is our strategy to attempt a total removal of the capsule. If the capsule is firmly adherent to critical neurovascular structures, we leave the adherent portion in place to minimize the risk of neurological sequelae.

Although a capsule remnant probably will result in recurrence, this will occur many years after surgery. Berger et al reported that it may take 30 to 40 years for recurrent symptoms to develop, a finding that supports the rationale for avoiding radical excision.[5]

Tumor removal itself may relieve symptoms, but is must be emphasized that epidermoid tumors commonly cause persistent distortion of trigeminal nerve even after satisfactory resection. Therefore, severing and removing the adherent tumor capsule and the arachnoid membrane to achieve neural axis straightening is mandatory for the cure and preventing of recurring pain. The surgeon also must pay attention to possible arterial compression at the REZ, and such an artery should be treated by MVD in addition to tumor excision. Kobata et classified the relationship between the CPA epidermoid and neurovascular structure into four types according to the compression pattern of the REZ of the cranial nerve. In type A, the nerve is completely encased by the tumor without displacement of the nerve axis. In type B, the nerve is compressed and distorted by the tumor. In type C, the nerve is displaced and compressed by the artery on the opposite side of the tumor, resulting in its being pinched by both the tumor and the artery.[12]

Now the latest trend is to use micro-scope and endoscope for the complete / maximum help to clear the remnents of epidermoids in the angles, corners at its peripheral part. We used endoscope in all 3 cases operated at Lahore General Hospital Lahore.

CONCLUSION

TN can be a typical symptom in epidermoid cyst in the cerebellopontine angle. This tumor can be removed easily via unilateral sub-occipital craniectomy and symptoms in our patients were relieved well and there was no recurrence symptom on the follow-up period till 9 years. Epidermoid patients were clinically indistinguishable from patients with trigeminal neuralgia from vascular cause, except symptom onset at a youn-

ger age. At operation, the root entry zone of trigeminal nerve should be examined for evidence of additional vascular compression. If vascular compression exists, MVD should be performed. Recurrence of tumor is rare in both total and sub-total removal cases, but long-term follow-up is required.

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