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Intracranial epidermoid cysts About 44 cases

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Abstract
Introduction: Intracranial epidermoid Cysts, also known as pearl tumor or cholesteatoma, is a rare, slow-growing benign lesion that accounts for 0.2–1.8% of intracranial tumors. As early as 1922, Cushing defined this tumor as epidermoid cyst of central nervous system for the first time, and later this disease was reported successively.
Materiels and Methods: Our study concerns 44 cases of diagnosed intracranial epidermoid cysts, collected in department of neurosurgery of cherchel hospital and ait idir hospital for a period of 20 years (from 2000 to 2020).
Results: Epidermoid cysts are benign tumors that account for approximately 1% of intracranial tumors, typically lateral in location, epidermoids are true ectodermal inclusion cysts lined by an epithelium. The average age of patients was 34 years old, more common in females and with 24 females to 20 males. The average duration of symptoms before diagnosis is 2 years. The clinical signs were diverse ranging from epileptic seizures, cranial nerve damage, to neurological deficit associated in most of our patients with a headache. Neuro-imaging (CT scan/ MRI) are the key exams. MRI is important especially in the development of surgical planning. Collected radiological data was in favor of intracranial epidermoid cyst; with 30 cases located in the cerebellopontine angle, and 4 in the posterior fossa.
We used different approaches depending on the location of the cyst. The evolution was satisfactory for the majority of patients. However, complications were noted. We deplore the death of 04 patients presented with meningitis.
Conclusion: Intracranial epidermoid cysts should be treated by radical surgical resection; however, that is not always the case, and the remaining tumor is the source of recurrence.
Key words: *epidermoid cysts; Cerebellopontine angle; ectodermal.*

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I. Introduction:

Intracranial epidermoid Cysts, also known as pearl tumor or cholesteatoma, are a rare, slow-growing benign lesion that accounts for 0.2–1.8% of intracranial tumors (1). As early as 1922, Cushing (2) defined this tumor as epidermoid cyst of central nervous system for the first time, and later this disease was reported successively.

Most of congenital epidermoid cysts are due to a failure in separating the surface ectoderm from the underlying structures, with its sequestration along the embryologic sites of dermal fusion forming eyes, ears, and face. (3,4). Alternatively, ectodermal cells may be transported later with the development of the optic vesicles, with a physio-pathological mechanism explaining localizations in the anterior cranial fossa and around the orbit. The most common site is the cerebellopontine angle (CPA), where epidermoid cysts account for 4.6% to 6.3 % of space-occupying lesions (5). Epidermoid cysts in the CPA, which spread along the cisterns of the posterior fossa, may intimately involve multiple cranial nerves and vascular structures and may be adherent to them and to the brainstem.

The present report details the clinical, radiological and surgical aspects of 44 patients with intracranial epidermoid cysts who were diagnosed and operated on.

Clinical Material and Methods

This is a retrospective study of 44 patients who were diagnosed and treated for an intracranial epidermoid cyst over the period from 2000 to 2020 in the neurological department of cherchel hospital and ait idir hospital. The diagnosis was confirmed a high-resolution brain MRI in multiplannar sections, especially the FLAIR and DWI sequences.

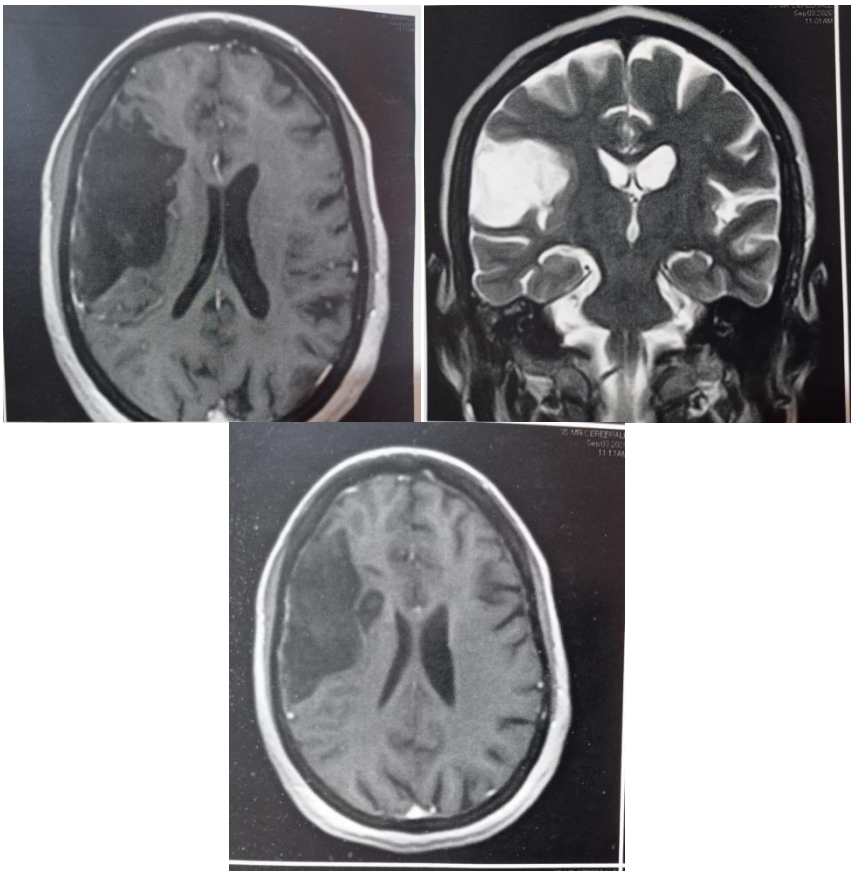

Figure 01: Brain MRI in sagittal T1, T2 weighed, FLAIR and diffusion sequences presenting a parieto-temporal epidermoid cyst.

The approaches used depended on the cysts locations. Postoperatively, all our patients were put under antibiotic and corticoid treatment. All precautions were taken before the operation while explaining the patient the nature and difficulties related to this surgery.

A radiological evaluation is systematically done in all our patients using an MRI.

II. Results:

Our series concerns 44 patients who were diagnosed and treated for an intracranial epidermoid cyst. The study included 24 females /20 males; the average age was 34 years old.

* The symptoms were diverse, depending on the location of the lesion.
* The most common symptom in CPA cysts was hypoacousia, followed with trigeminal neuralgia, while supra-tentorial cysts presented with signs of intracranial hypertension and seizures.

Initial Symptoms	Cases	**%**
Hypoacousia	23	52,27 %
Trigeminal neuralgia	19	34 ,18%
facial paralysis	08	18,18%
Mixed nerve damage	06	13,63%
Ataxia	15	34,09%
IH	15	34,09%
Decreased visual acuity	4	9,09%
Epileptic seizures	8	18,18%

Table 01: Initial symptoms presented in our patients.

Neurological examination	Cases	**%**
VIII	19	34,18%
VII	08	18,18%
V	15	34,09%
Mixed nerves	06	13,63%
Cerebellar dysfunction	13	29,54%
Pyramidal syndrome	12	27,27%
Member deficit	09	20,45%
Papillary oedema	. 4	9,09%

Table 02: Neurosurgical examination

Neuro-imaging (CT scan/ MRI) are the key exams. MRI is important especially in the development of surgical planning. Collected radiological data was in favor of intracranial epidermoid cyst; with 30 (68,18%) cases located in the cerebellopontine angle, and 4 (9,09%) in the posterior fossa, and 26 (59,09%) presented with a hydrocephalus.

The lesions presented with the typical image of an epidermoid cyst, CT scans showed a homogeneous hypodense lesion with irregular borders and without contrast enhancement, and the MRI findings consisted of T1 and T2 sequences, where the lesions presented as isointense to CSF and without enhancement, the lesions where the signal was too similar to the CSF, we used both the FLAIR and DWI to differentiate between our lesions and archnoid cysts.

Surgical management was the chosen method to treat all of our patients; the goal was to achieve a maximum excision with the preservation of the neurological functions. A total removal of the cyst was not possible in all of our cases. The epithelial capsule was peeled from the structures when it was possible to prevent recurrence. The second goal of the resection was to prevent the tumor contents from spilling into the subarachnoid space as they are intensely irritant and can cause aseptic meningitis, also the surgical field was irrigated to ensure the removal of any remnants and to avoid dispersing remnants to other areas of the brain.

All out patients who presented with a hydrocephalus (59,09%) benefited from a Ventriculo-peritoneal shunt.

We used a retro-sigmoid approach in all the patients with a CPA location, the procedure was performed under a neurophysiological control to preserve the functions of different nerves in the area; while the supra-tentoriel approach differs according to the cycts' locations; and the posterior fossa was approached by the sub-occipital approach.

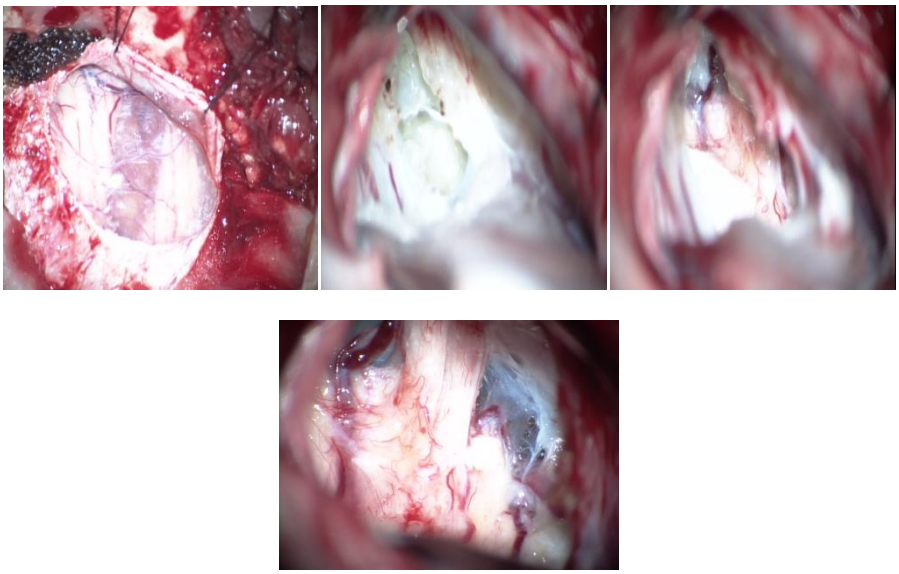

Figure 02: Operative view of the CPA epidermoid cyst.

The extent of resection is determined by the tumor adhesion to surrounding structures and by the pattern of extension. We achieved a total resection in 19 cases (43,18%) , 15 patients (34,09%) benefited from a sub-total resection, while 10 patients (22,72%) had only a partial resection.

Post operative complications were typical with meningitis in 5 (11,38%) patients, 02 of these cases were aseptic meningitis; CSF fistula in 4 (9,09%) patients, and we lost 04 (9,09%) patients.

We noted a decrease in symptoms for 32 (72,72%) patients while 8 (18,18%) patients kept the same symptoms.

III. Discussion:

Epidermoid or "pearly" tumors were described by Cruveilhier10 and designated the "most beautiful tumors of the body" by Dandy (6). Epidermoid cysts have a thin capsule of stratified, keratinized squamous epithelium (7). They grow as a result of desquamation of epithelial cells, which later break down into keratin and cholesterol; they may also expand and become enormous by neoplastic cellular growth. (7,8)

Intracranial epidermoid cysts are an uncommon finding, accounting from 0.3% to 1.8% of all intracranial masses; The cisterns of the CPA and parasellar region are the most common site for development of an epidermoid cyst (5,9,10,11,12,13,14,15). Vestibular schwannomas and meningiomas are the two most common tumors of the CPA, followed by epidermoid cysts (16,17). Intracerebral location is rare and can be confused with cystic primary or metastatic tumors (18,19). In our study, CPA location was in fact the most common site with a percentage of 68,18%.

In literature a slight male predilection was reported, while in our series we found a slight female predilection with a ratio of 24 females to 20 males and most patients present them in the first 4 decades of life (3,4) similar to our cases where the average age was 34 years old.

Epidermoid tumors grow slowly from the accumulation of breakdown products of desquamated epithelial cells, The symptomatic onset of epidermoid cysts is usually slow, lasting 2 or more years, same as our series where the mean of symptoms was 2 years. Although some patients with remitting signs and symptoms28 or with rapid onset (5,12) have been reported.

The type of symptoms depends on the location of the cysts; their growth mechanism from the accumulation of keratin produces a chronic inflammatory reaction that increases their adherence to vascular and neural structures.

Epidermoid cysts of the CPA cause the symptoms and signs of a slowly expanding mass in that region (20) including ataxia, nystagmus, facial pain, paresthesias, and weakness (9,13,20-24). The involvement of the facial nerve or unilateral hearing loss (5,12,15,25) has been reported as the most common sign. Our patients were typical in their symptoms where and presented with CPA nerves deficit.

The supra-tentorial cases were diverse and the symptoms depended on the location of the cysts. Rarely, an epidermoid tumor may leak keratin into the surrounding cisterns and cause aseptic or chemical meningitis, fortunately none of our patients presented with this complication. However, the meningeal irritation from the keratin can also contribute to the development of hydrocephalus; n our series the rate of hydrocephalus was relatively high with 59,09% of the cases.

Computerized tomography scans demonstrate an hypodense area with density values similar to, or slightly higher than, that of CSF (10,26,27,28,29). The margins are usually well defined but with irregular contours, which do not generally enhance and rarely present calcifications or perifocal edema (9,10,12,28). It is reportedly rare for the lesion to show capsular enhancement (8,15,20,30). MRI findings consisted of T1 and T2 sequences, which are isointense to CSF and without enhancement. Fast fluid-attenuated inversion recovery (fast-FLAIR) MRI or echo-planar diffusion-weighted imaging (DWI) is superior to conventional MRI in detecting epidermoid tumors (31). On the FLAIR imaging, there is often a heterogeneous/dirty signal higher than CSF. On the DWI, the tumor has an increased signal and appears bright when compared to the CSF. Our findings were similar to literature, and we used the same sequences (FLAIR and DWI) to better differentiate our cysts and other lesions; we had 30 patients with a CPA locations, and 59,09% of our cases presented with an adjacent hydrocephalus.

Some epidermoid tumors may have atypical radiological features and can be confused with other tumors (32,33). One of the main problems encountered in epidermoid diagnosis is distinguishing CSF from tumor. The most common differential diagnosis is the archnoid cysts, the dermoid cyst, and the metastatic tumors.

The radiological definition will determine the choice of surgical approach depending on the main extension of the cyst (24,34). Complete excision with preservation of the cranial nerve function should be the goal of the management of epidermoid tumors. Total removal may not be possible without inducing severe deficits; therefore, subtotal resection is acceptable in most cases. Neurophysiological monitoring aids in preventing cranial nerve and brainstem injury. The extent of resection is determined by the tumor adhesion to surrounding structures and by the pattern of extension. The growth mechanism from the accumulation of keratin can produce a chronic inflammatory reaction that increases their adherence to vascular and neural structures.

In our series we first treated the Hydrocephalus with a ventriculo-peritoneal shunt and afterwards managed our cysts, we used a retro-sigmoid approach in all our CPA locations similar to literature were the retro sigmoid approach is favored (35) while the Posterior fossa location we used the sub-occipital approach. Although the surgery was very demanding, we achieved a total resection in 19 cases (43,18%) , 15 patients (34,09%) benefited from a sub-total resection, while 10 patients (22,72%) had only a partial resection.

In literature more than 75% of the patients have an excellent functional prognosis with an independent and useful life after the operation (24,36). We had approximativly the same rate at 72,72% of our cases. The 20-year survival rate is 92.8% (36). The function of the trigeminal nerve and the facial nerve may recover after decompression, but the outcome of the symptoms related to the cochlear nerve is less certain.

In our study post operative complications were typical with meningitis in 5 (11,38%) patients, 02 of these cases were aseptic meningitis; CSF fistula in 4 (9,09%) patients; while in previous reports, complications varied from Cranial nerve deficit or worsening; Aseptic meningitis; CSF fistulas and leakage; and Infections.

Operative morbidity and mortality from the attempted removal of these cysts have declined remarkably in the last 20 years. Prior to the advent of the operative microscope, operative mortality ranged from 20% to 57%.15 This rate dropped to approximately 6% for operations performed since 1951.25 Compared to our study where we lost 04 (9,09%) patients due to post operative meningitis.

The main problem encountered with these cysts is recurrence, related to incomplete removal of the cyst (12,12,13,14,22,37). In such cases, CT scans and MR images should be used to closely observe the evolution of these lesions.

IV. Conclusion:

Intracranial epidermoid Cysts, also known as pearl tumor or cholesteatoma, are a rare, slow-growing benign lesion that accounts for 0.2–1.8% of intracranial tumors. They represent several difficulties in their management, starting with the diagnosis where several MRI sequences are best suited to the diagnosis, and the second difficulty resides in the surgical management where the total resection is not always possible and might be the source of possible recurrence. Our series like the previous studies highlights such difficulties, and confirms the necessity for more investigations in the management of these cysts.

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