

## Epidermoid cyst of the brain: A common cystic lesion in rare location

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### ABSTRACT

Epidermoid cysts are common benign tumors comprising around 1% and 2% of all intracranial tumors. Their usual locations include the parasellar region and cerebellopontine angle, and less commonly, the Sylvian fissure, suprasellar region, cerebral, and cerebellar hemispheres. Epidermoid cysts located in the brain stem are rare. These epidermoid cysts are similar to epidermoids arising in the skin which contain cheesy and flaky-white soft pulaceous material. Epidermoid cysts are very slow-growing tumors having a similar growth pattern of the epidermal cells of the skin and develop from the remnants of epidermal elements during the closure of the neural groove and disjunction of the surface ectoderm with neural ectoderm between the 3<sup>rd</sup> and 5<sup>th</sup> weeks of embryonic life. The ideal treatment of choice is the removal of cystic components with the complete resection of the capsule. We are presenting an interesting case of an epidermoid cyst in the frontal lobe in a 42-year-old male along with radiological investigations.

**Key words:** Embryonic life, Epidermoid cyst, Intracranial tumors

Epidermoid tumors of the brain are congenital, benign, and very slow-growing tumors [1]. These tumors are very rare, accounting for 1–2% of all intracranial space-occupying lesions [2]. Epidermoid tumors are usually located in the cerebellopontine angle, parasellar region, fourth ventricle, and suprasellar chiasma [1-3]. Epidermoids are 90% intradural and 10% extradural lesions. The most common location is the cerebellopontine angle (40–50%), suprasellar cistern (10–15%), fourth ventricle (17%), middle cranial fossa, and interhemispheric area (<5%) and is very rare in the spinal regions. These tumors are congenital and originate from the mesoectoderm [2]. During the 3<sup>rd</sup>–5<sup>th</sup> weeks of embryonic life, epidermoid tumors arise from the ectodermal cells trapped within the neural tube at the time of closure [2]. Since these are slow-growing tumors, the patient usually presents during the fourth decade and their growth is similar to the epidermal cells of the skin. The tumors appear to grow along the cisternal spaces and are rarely seen infiltrating the brain parenchyma [1].

We are presenting the case of an epidermoid cyst in a 42-year-old male with the presentation, radiological imaging, surgical management, and histopathological diagnosis.

### CASE REPORT

A 42-year-old male patient was brought to our hospital with the complaints of sudden stiffening of the right upper limb in flexed

position along with altered sensorium. This episode lasted for 15 min after which, the patient was normal. The patient had insidious onset back pain for the past 4 months. There was no history of vomiting, neck pain, neck stiffness, or headache. He was a known hypertensive and diabetic since the age of 36 years and is on regular antihypertensive and oral hypoglycemic medications.

On clinical examination, the patient was conscious and oriented with normal vital signs. Cardiovascular, respiratory, and abdominal system examinations were normal. A nervous system examination revealed the Glasgow Coma Scale of E4V5M6 (E-Eye opening, V-Verbal response, M-Motor response) with no neurological deficits. There were no signs of papilledema, history of focal seizures, speech disturbance, cranial nerve deficits, and signs of meningeal irritation. The motor and sensory system appears normal. Superficial and deep reflexes appear to be intact. A provisional diagnosis of low-grade glioma was given and the patient underwent further investigations.

Plain computed tomography (CT) scan brain was done which revealed a well-defined hypodense lesion in the right high frontal region measuring 2.5×2 cm, suggesting the possibility of granulomatous lesion versus brain contusion. Magnetic resonance imaging (MRI) brain with contrast was done which showed evidence of a well-defined non-enhancing T2 hyperintense/T1 hypointense lesion measuring 2.8×2.7×2.5 cm in the right high frontal lobe cortical and subcortical region. There was no evidence of acute infarction, hemorrhage, and abnormal parenchymal or

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meningeal contrast enhancement. MRI report impression was low-grade glioma which suggested histopathological correlation.

Surgery was planned on the basis of MRI report, for which the right frontoparietal craniotomy was done around the coronal suture. Dura was opened in a “C”-shaped manner with its base toward the median. Right frontal pale tissue was identified and corticotomy was done over the lesion. A rubbery grayish-white tumor was identified, and excision of the entire lesion was done through microdissection. There were no extensions of the lesion seen. Hemostasis was achieved, and the dura was approximated. The tissue was sent for histopathological examination. The postoperative period was uneventful. The patient recovered well without any neurological deficits or symptoms.

Histopathological sections of the multiple pale brown soft-tissue fragments showed multiple scattered basophilic amorphous calcified matrix and solid nests of mature squamous epithelial cells. The surrounding area also showed fibrocollagenous connective tissue. There were no brain parenchymal tissues noted in the given fragments (Fig. 1). Diagnosis of the epidermoid cyst was made. Immunohistochemistry was done, and the diagnosis was confirmed to be epidermoid cyst as cytokeratin stains were only positive (Figs. 2 and 3). The patient recovered well, and his symptoms eased off after surgery. He is under regular follow-up for the past 4 months without any acute symptoms.

## DISCUSSION

Epidermoid cysts or tumors are benign, slow-growing tumors with a higher incidence in the fourth decade of life with a male predominance. Intracranial epidermoid tumors comprise about 1.5% of all intracranial neoplasms. These lesions appear as cysts in imaging studies [2,3]. They are also known as cholesteatoma or pearly beautiful tumors. Epidermoids were described by French pathologist Cruveilhier as the “most beautiful tumors of all the tumors” as they appear pearl white. Since these tumors were first described by him, they were also known as Cruveilhier tumors [1-3].

The most common location of the epidermoid cyst is the retrosellar cerebellopontine angle [1-3]. Other locations with the least common occurrence include parasellar-sylvian fissure, suprasellar chiasmatic, and basilar posterior fossa [2,3]. These cysts develop from the ectodermal cell rests that displace and become trapped in the neural tube at the time of closure. They appear as exophytic lesions with irregular cauliflower-like outer

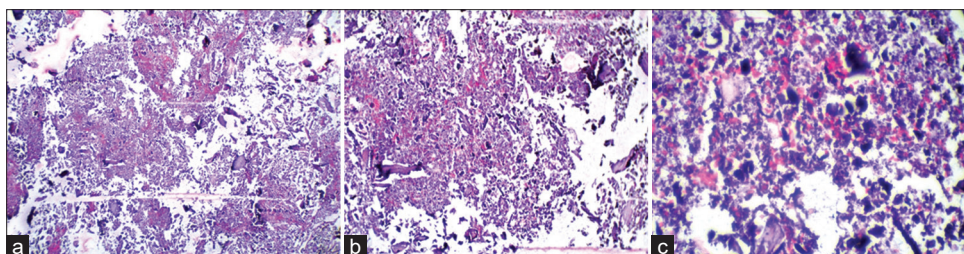
surfaces, it may also be seen encasing nerves and vessels [3,4].

Epidermoids appear pearly white due to the accumulation of keratin and cholesterol crystals; these are the breakdown products from the desquamating epithelial cells [1-3,5]. The outer capsule of the cyst has the epithelial layer and inner cystic fluid [1,3]. The cyst may contain inner tumor fluid, which can cause secondary infection at other sites [2,4].

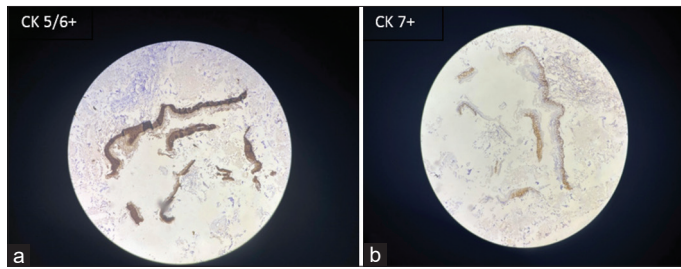
The clinical manifestation of epidermoid cysts is due to the compression of neighboring structures, resulting in signs of raised intracranial pressure, cerebellar symptoms, seizures, and cranial nerve deficits. Headache is the most common presenting symptom in many patients [1,2,4].

The important differential diagnosis for epidermoid cysts include dermoid cysts, arachnoidal cysts, neurocysticercosis, hamartomatous lipoms, neuroenteric cyst, and other cystic neoplasms, but these can be ruled out using radiological imaging such as CT or MRI [2]. CT shows hyperdense or hypodense or isodense lobulated mass depending on the content of the cyst. Hyperdensity in CT may be due to the combination of leukocytes, proteinaceous material, saponification of keratinized debris, and lipid debris; occasional calcification may also be noted [1-3]. MRI is the standard diagnostic tool in these cases. Epidermoid cysts appear hypointense on T1-weighted, hyperintense on T2-weighted, fluid-attenuated inversion recovery) with hyperintense restriction on diffusion-weighted imaging (DWI) without contrast enhancement. The absence of tumor edema differentiates epidermoid cysts from gliomas in case of intrinsic lesions. DWI is useful for extrinsic lesions to distinguish between arachnoid cyst and abscess [2,3].

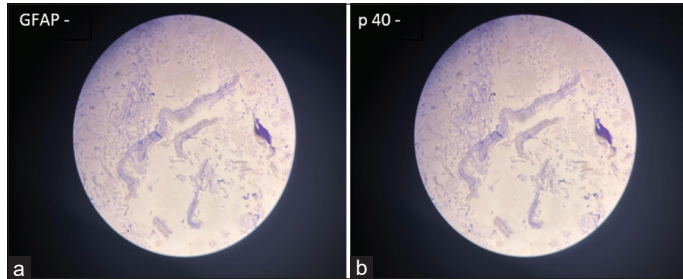
Surgical resection is the treatment of choice [3]. Microscopic sharp dissection of every bit of the capsule is essential to prevent recurrence. Partial removal leads to recurrence after a prolonged period due to the slow growth rate of the tumor [3]. Subsequent surgery is indicated only when the patient is symptomatic. Inadequate removal or spillage of the contents into the subarachnoid space may lead to chemical meningitis and adhesions [6-8]. Rupture of the cyst during surgery may also cause aseptic meningitis, hence to prevent this complication, surgeons excise the capsule in toto by sharp dissection and copious irrigation of the surgical field with hydrocortisone and postoperative intravenous dexamethasone prevents meningitis in such cases [9-11]. Malignant transformation into squamous cell carcinoma is a known but extremely rare occurrence reported in recurrent epidermoids [3,6,10]. Total



**Figure 1:** Photomicrograph showing (a) multiple scattered basophilic amorphous calcified matrix (hematoxylin and eosin stain,  $\times 10$ ); (b) solid nests of mature squamous epithelial cells (hematoxylin and eosin stain,  $\times 10$ ); (c) solid nests of mature squamous epithelial cells with surrounding fibrocollagenous connective tissue (hematoxylin and eosin stain,  $\times 40$ )



**Figure 2:** Photomicrograph of tumor cells showing (a) strong positivity for Cytokeratin 5/6 (CK 5/6 immunohistochemical stain,  $\times 10$ ); (b) Cytokeratin 7 positive mature squamous cells (CK 7 immunohistochemical stain,  $\times 10$ )



**Figure 3:** Photomicrograph of tumor cells showing (a) negative staining for Glial Fibrillary Acid Protein (GFAP immunohistochemical stain,  $\times 10$ ); (b) negative staining for p40 (p40 immunohistochemical stain,  $\times 10$ )

resection of the lesion is mandatory to avoid recurrence and complications postoperatively [5,7].

## CONCLUSION

Epidermoid cysts outside the typical location are extremely rare lesions with atypical imaging features. Diffusion-weighted imaging is definitive for the diagnosis. Currently, the debate is on the optimal treatment for epidermoid cysts. Gross total resection of the lesion is the definitive treatment that prevents recurrence or aseptic meningitis. Hence, in our case also, total resection of the lesion was done. Cases showing invasiveness into the vault of the skull, meninges, or cerebral sinuses need total excision which

may be surgically challenging and there are increased chances of postoperative complications such as cerebrospinal fluid leakage and chemical meningitis.

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