

## Case Report:

### EPIDERMOID CYST OF THE LATERAL VENTRICLES–A RARITY

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#### ABSTRACT:

Epidermoids are congenital lesions of epidermal origin, representing 0.2% to 1.8% of all primary intracranial tumours. They arise from epithelial remnants at the time of neural tube closure, between the third and fifth week of foetal development. Epidermoid cysts occurring within the lateral ventricles are rare. They are slow growing benign tumors usually presenting with non specific signs of deterioration of mental functions including recurrent headaches, seizures, paresis, chemical meningitis, symptoms of fat embolism, olfactory delusion, psychomotorv and visual disturbances. Epidermoids grow by accumulation of keratin and cholesterol, which are the breakdown products of desquamated epithelial cells. They grow linearly rather than exponentially and hence are slow-growing lesions. Therefore, they clinically manifest. We report a rare case of epidermoid cyst involving lateral ventricles in a 45 year old female presented with symptoms of headache, vomiting, seizures , diminition of vision and quadriparesis. Intra ventricular epidermoids are rare but must be included in diferential diagnoses of Ependymomas,Subependymomas & Choroid plexus Papillomas. We present this case because of its rarity of occurrence in the lateral ventricles.

**KEYWORDS:** Epidermoid cyst , Lateral ventricles , Intra- ventricular neoplasm

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#### INTRODUCTION :

Epidermoids are congenital lesions of epidermal origin, representing 0.2% to 1.8% of all primary intracranial tumours <sup>(1)</sup>. They arise from epithelial remnants at the time of neural tube closure, between the third and fifth week of foetal development <sup>(2)</sup>. Their preferred sites of formation are the cerebellopontine angle and supra-sellar region protruding in the subarachnoid space. Intra ventricular Epidermoid tumors are slow growing, benign, congenital neoplasm of the CNS. Epidermoids occurring within the lateral ventricles are very rare. They are slow growing and the clinical presentation is non- specific like deterioration of mental functions

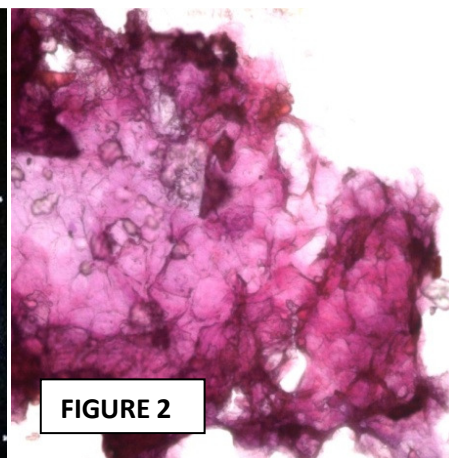
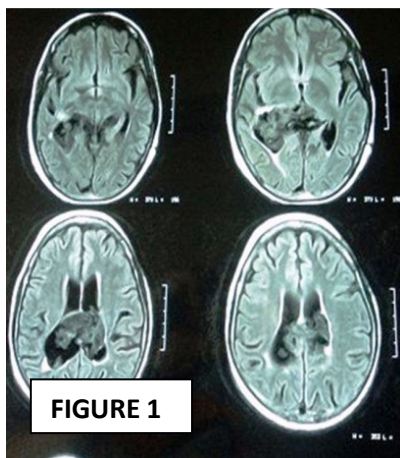
. Epidermoids of the lateral ventricles, suggesting an origin in the basal cisterns and extension through the choroidal fissure into the lateral ventricle with expansive growth. Rarely, these lesions may rupture, resulting in dissemination of the intracystic contents into the subarachnoid space and ventricles, and consequently, present with recurrent episodes of aseptic meningitis<sup>(3)</sup>. Spontaneous rupture of lateral ventricle epidermoid cyst can lead to diffuse dissemination of the fat component in the subarachnoid space, cerebrospinal fluid (CSF) cisterns, and ventricle system. However, the rupture of the cyst is considered a serious and frequently lethal complication.

**Case Report:**

A 45yr female presented to neuro-surgery out-patient department with complaints of severe headache since 15 days ,vomiting and seizures since 10 days and weakness of the all four limbs since 5 days. Patient also complained of diminution of vision since 2 days. Clinical examination showed mild pallor, disoriented speech, impairment of the visual acuity. On neurological examination, there was quadriparesis , more profound on the right side of the body, there was disorientation and psychomotor deceleration. Power in the right upper and lower limbs 3/5 and left upper and lower limb was 4/5 .Vision in the right eye was 6/18 and in the left eye was 6/18 with bilateral papilloedema .The score of mini – mental test was low (17/30) psychologist examined the patient and concluded that the cause of the symptoms is organic.

**RADIOLOGICAL FINDINGS**

**Magnetic Resonance Imaging (MRI) :** MRI of the patient showed a large irregular mass lesion measuring 5 x 4 x 2cm of altered signal intensity appearing hypo intense on T1, hyper intense on T2, FLAIR seen in atria and occipital horns of both lateral ventricles margins the lesion is seen on both sides of intra ventricular septum. ( **FIGURE 1** )



**IMPRESSION** –Features suggestive of Intra ventricular mass lesion involving atria & occipital horns of both lateral ventricles - ? Epidermoid cyst.

The patient underwent craniotomy and some amount of the tissue excised and sent for squash diagnosis (intra-operative cytology). Intra-operatively the appearance of the tumor was pearly white and avascular, occupying the whole of the atrium , occipital horns and body of both lateral ventricle.

**Squash Diagnosis ( Intra-operative Cytology )**

Cytosmears made from the tissue showed anucleate squames, acellular eosinophilic keratinous material .( **FIGURE 2** )

**IMPRESSION - Epidermoid cyst.**

Entire tumor excised after the squash diagnosis and the tissue sent for histopathological examination for confirmation.

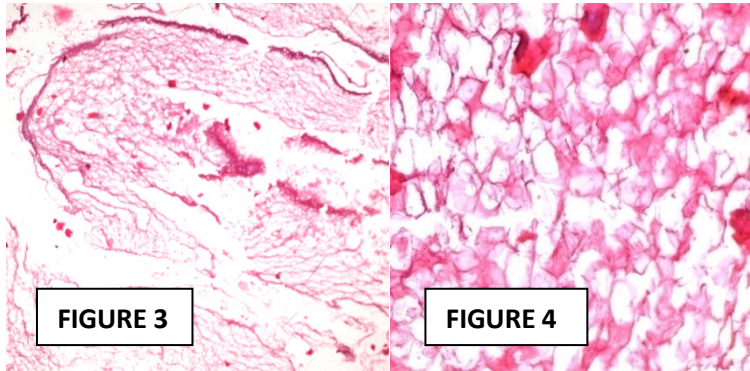
**Gross Findings:** Received multiple pearly white to grey white friable soft tissue bits altogether measuring 3 x 2.5 x1 cm.

**Histopathology Examination:** Multiple sections studied show plenty of anucleate squames, acellular eosinophilic material , lamellated keratin with foci of calcification. ( **FIGURE 3& 4** )

**Impression - Epidermoid cyst**

**FIGURE 1- MRI showing tumor in the both lateral ventricles with mild to moderate dilatation of the ventricles.**

**FIGURE 2 – Squash cytology – showing anucleate squames ,acellular eosinophilic keratinous material.**



**FIGURE 3 ( LOW POWER VIEW) – Showing flattened epithelial lining , lamellated keratin , anucleate squames.**

**FIGURE 4(HIGH POWER VIEW) – Showing polygonal anucleate squames with thin cell wall and clear cytoplasm and focal keratinous material.**

## DISCUSSION

Epidermoid tumors are uncommon, representing only 0.2-1.8% of all intracranial tumors. Epidermoids occurs within the lateral ventricles are very rare. They are also referred to as epidermoid lesions, intracranial cholesteatoma, inclusion tumors, pearly tumors or tumor peelee. Our extensive review of literature yielded only nine reported cases (**Table 1**). Pathogenesis of intra-ventricular epidermoids is uncertain. They arise from epithelial remnants at the time of neural tube closure, between the third and fifth week of foetal development. Probably, there is a direct relation to the development of choroidal vessels, which explains the lateral migration through the choroidal fissure and subsequent entrapment of neuroepithelial cells<sup>(4)</sup>. The tumors have a connection with median/paramedian structures, and the view that these lesions commence from quadrigeminal cisterns

also needs serious consideration. Epidermoids grow by accumulation of keratin and cholesterol, which are the breakdown products of desquamated epithelial cells. They grow linearly rather than exponentially, this mode of growth makes them slow-growing lesions. Therefore, they clinically manifest in the later decades of life. Most of the epidermoids do not present with hydrocephalus. This is because of the smooth and pliable nature of the cyst wall and its content, which allow progressive, slow molding of the surrounding neural structures. Moreover, it allows maintenance of CSF pathways. Because of the rarity of lateral intraventricular epidermoid, no fixed pattern of clinical presentation has been described and usual clinical presentations described in the literature are non-specific, like headache, cognitive deficit or psychiatric symptoms. In contrast to the present knowledge that the CSF obstruction is not

seen even if this lesion is in the vicinity of the foramen of Monro . Our patient had obstructive type of hydrocephalus due to compression of the 3rd ventricle. This resulted in papilloedema and diminished vision, which improved after total surgical removal of the lesion.

Considering the symptomatic and clinical improvement in our patient, surgical total removal is a better option to manage these types of lesions.

**Immuno-histochemistry (IHC)** -The lining epithelium of the epidermoid cyst is immunoreactive for cytokeratins and epithelial membrane antigen(EMA).In contrast to the cyst of neuroglial origin there is no immunoreactivity of the lining epithelium with Glial Fibrillary Acidic Protein(GFAP)<sup>(5)</sup>. In the recent study of 54 patients operated on for epidermoids of the brain only 9 cases were reported in the Lateral Ventricles <sup>(6)</sup>.

<b>Table : 1 Literature review of lateral intraventricular epidermoids</b>	
<b>Name of author</b>	<b>No. of cases of lateral ventricular epidermoid</b>
Franko <i>et al.</i> , (2008)	1
Bhatoe <i>et al.</i> , (2006) <sup>(5)</sup>	1
Akar <i>et al.</i> , (2003)	1
Menq <i>et al.</i> , (2006)	4
Koot <i>et al.</i> , (2003)	1
Rajendra <i>et al.</i> , (2012) <sup>(7)</sup>	1

**CONCLUSION**

Epidermoids are the rare lesions of the lateral ventricles . In addition to the rarity of these lesions in the lateral ventricle, intraventricular epidermoid can attain a large size in a short period and can manifest at a young age. It can also cause obstruction to the CSF pathway and can manifest clinically as loss of

vision and other features of hydrocephalus. Gross total removal is achievable; however, residual tumour capsules adherent to neurovascular structures should be left behind to minimize complications. Left over residual capsules leads to recurrence of the epidermoids.

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