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Case Report

Pineal Region Tumor with Ventricular Extension with Hydrocephalus Removed by Supracerebellar Infratentorial Approach in a 19-Year-Old Female: A Case Report

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Abstract

Pineal region tumors are uncommon, accounting for d"1% of intracranial tumors in adults and 3–8% of pediatric brain tumors. Getting an epidermoid in pinealregion is also very rare. In 1928 Cushing was the first to report the pineal localization of the epidermoid cyst.We report a case of pineal epidermoid with ventricular extension with obstructive hydrocephalus, which was diagnosed depending on magnetic resonance imaging (MRI) findings. Knowing its benign nature, at first we planned for an Endoscopic Third Ventriculostomy and later on a gross total removal was done.

Keywords:*Pineal region tumor, Pineal epidermoid, Endoscopic Third Ventriculostomy*

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Introduction:

Intracranial epidermoid cysts comprise 0.2–1% of all intracranial tumors[1]. They are most commonly located in the cerebellopontine and parasellar cisterns, but have been reported throughout the neuraxis [2].The pineal region is exceptionally subject to such kind of tumor.Pineal gland is made up different cell types, accounting for diverse pathology of tumors in this region. These tumors are classified into four main types, i.e. germ cell tumors, pineal parenchymal cell tumors, glial cell tumors, and other miscellaneous tumors. Pineal epidermoid tumor belongs to miscellaneous tumors of this region.

Case Report

A 19 year old female was got admitted at BSMMU on 15/06/2023. She was in good health until 1 year ago, when she started complaining of early morning headache which was dull aching, aggravated by coughing, bending and sneezing that were relieved

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by common analgesics. At that time she had no complaints concerning vomiting nor visual disturbance. About one month before admission she was experiencing visual disturbance. On physical examination she was in good general health. Neurological examination gave entirely normal results except her fundus which revealed chronic papilloedema. Plain x-ray films of the skull demonstrated signs of chronic

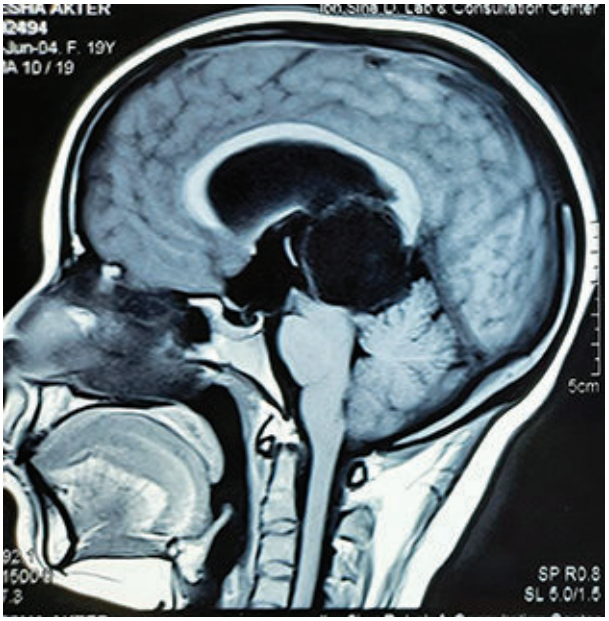

Fig.-1: *T1WI showing a hypointense*

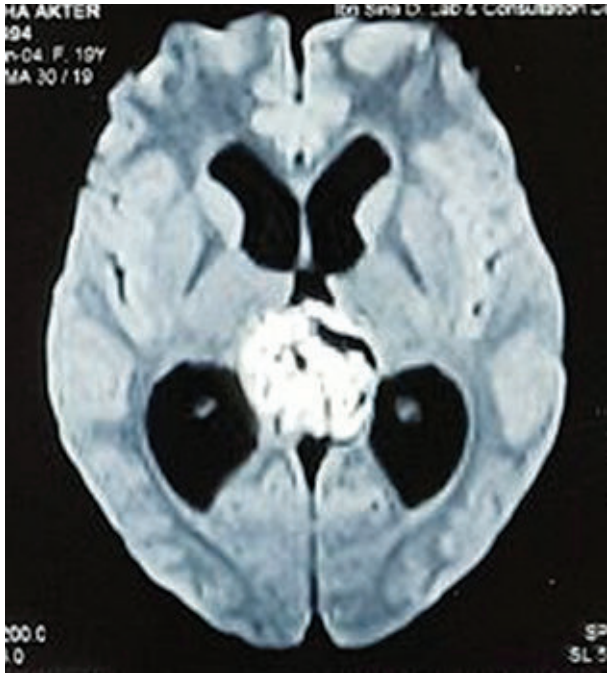

Fig.-2: *DWI showing high signal lesion at pineal region*

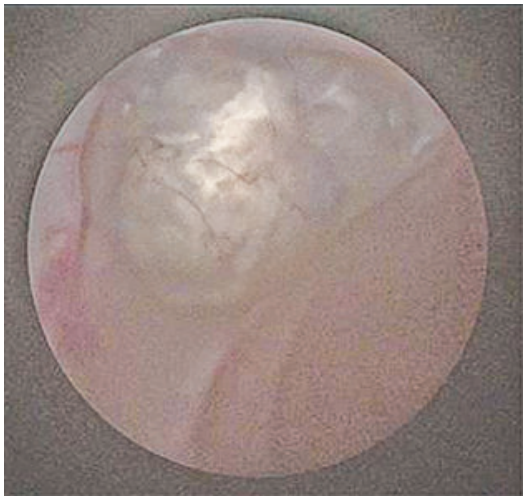

Fig.-3: *During ETV. Direct visualization of ventricular portion of tumor*

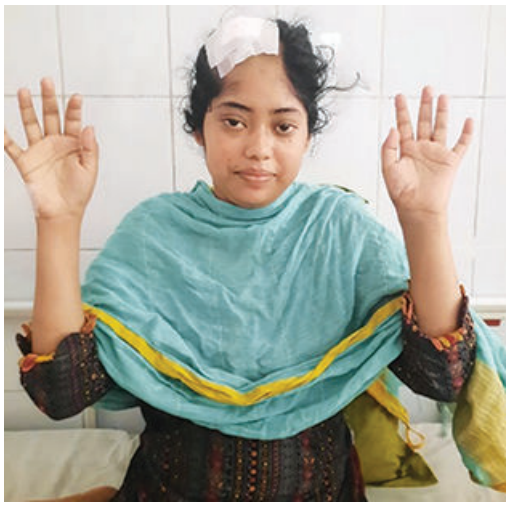

Fig.-4: *After doing ETV*

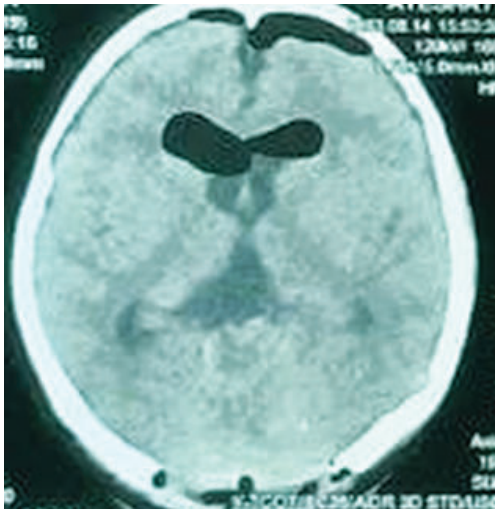

Fig.-5: *Postoperative CT Scan*

intracranial hypertension with copper beaten appearance and eroded posterior clinoids. There was no pathological calcification.A magnetic resonance imaging (MRI) scan demonstrated that the nonenhancing cystic lesion was of signal intensity comparable to that of CSF in all sequences at pineal region slightly more on right side compressing quadrigeminal plate and cerebral aqueduct causing obstructive hydrocephalus, Diffusion Weighted Image(DWI) showed marked diffusion restriction and high signal and venogram demonstrated displacement of the internal cerebral veins and vein of Galen upwards. Our clinicoradiological diagnosis was Epidermoid tumor with ventricular extension with acute hydrocephalus. As an emergency management we did her Endoscopic Third Ventriculostomy on 17/06/23. While doing this we saw the ventricular extension of lesion giving pearly white appearancewhich securedour diagnosis. After the procedure Patient's headache and visual disturbance were gone. Then we planned for her definitive surgery on 11/08/23.The patient was positioned in semisitting position. We approached through supracerebellar infratentorial route. Using microsurgical techniques, we explored the pineal region. Just in front of thickened arachnoid in the quadrigeminal cistern, there was a pearly tumor. Cyst wall was glistening; intratumor decompression was done. Cystic contents were avascular, pearly white, soft, and waxy debris. Contents were removed piecemeal, followed by performing extracapsular decompression. A gross total removal was achieved without any injury to neural or vascular structures. We had to leave in situ a tiny fragment of the capsule that was adherent to the right basal vein of Rosenthal. Histopathological report revealed Epidermoid Cyst.

Discussion:

Epidermal cysts develop when remnants of the ectodermal layer become trapped between two merging ectodermal surfaces. Other mechanisms, such as trauma and differentiation from multipotential cell rests, have also been implicated in the formation of epidermoid tumors. Epidermoids exhibit a linear growth pattern similar to normal skin, and due to their slow growth rate, these tumors can reach a relatively large size before causing symptoms in the patient. These tumors are typically found within the basal subarachnoid cisterns and often extend both above and below the tentorium[3]. They are notable for their ability to expand anatomical spaces in the region and encase major arteries and perforators within the cisterns[4]. On T1- and T2-weighted images, epidermoid tumors exhibit a signal intensity similar to cerebrospinal fluid, resembling arachnoid cysts[5]. Diffusion-weighted imaging (DWI) is the most effective test for distinguishing between epidermoid and arachnoid cysts. Epidermoids appear hyperintense on DWI, while arachnoid cysts appear hypointense. Contrast MRI provides insights into the tumor's relationship with major vessels. In our case, the tumor had displaced major vessels upward. The choice of the surgical approach depends on the tumor's extent and its proximity to major vessels. In our case, given the significant ventricular portion of the tumor and its upward displacement of major vessels, we selected the supracerebellar infratentorial route. Performing an endoscopic third ventriculostomy (ETV) prior to definitive surgery relieved the patient's symptoms and also facilitated our diagnosis through direct visualization. While performing ETV, a biopsy sample may be obtained, but we avoid it since the tumor appeared consistent with an epidermoid, and our plan was to achieve gross total removal via the supracerebellar infratentorial approach.

Conclusion:

Pineal epidermoid cyst is a very rare case. Gross total removal is the ideal treatment. Unfortunately, it's not always possible because of the characteristics of the tumor and the pineal region.If it is concomitant with obstructive hydrocephalus an ETV may be helpful to reduce symptoms as well as making a diagnosis .

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