Case Report

Pineal epidermoid

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Abstract

**Background:** Tumors of pineal region are uncommon, accounting for ≤1% of intracranial tumors in adults and 3–8% of pediatric brain tumors. Epidermoid cysts account for 0.2–1% of all intracranial tumors. The majority occur in and around the cerebellopontine angle and suprasellar area. Getting an epidermoid in pineal region is very rare.

**Case Description:** We report a case of pineal epidermoid, which was diagnosed correctly as epidermoid depending on computed tomography (CT) and magnetic resonance imaging (MRI) findings. Knowing its benign nature, we accordingly planned for its near-total removal.

**Conclusion:** Most cases of pineal tumors present as obstructive hydrocephalus. They either require pre- or postoperative ventriculoperitoneal (VP) shunt. If properly planned, many benign pineal tumors may be successfully excised and, most importantly, postoperative VP shunt could be avoided.

**Key Words:** Epidermoid tumors, pineal tumors, three-quarter prone position

INTRODUCTION

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 41-year-old male presented to us with complaints of holocranial headache for last 1 year, progressive diminution of vision for last 6 months, ataxia for last 3 months, and memory disturbance for last 1 month. On neurological examination, we found bilateral papilledema with impaired vision in both eyes. Computerized tomography (CT) scan revealed well-defined hypodense and nonenhancing mass in the pineal region, and patchy calcification in the peripheral part of the tumor, with hydrocephalus [Figure 1a].

Magnetic resonance imaging (MRI) revealed complex heterogeneous, ill-marginated mass in pineal region, with extension encroaching midbrain and infratentorium. The mass was compressing the aqueduct and causing obstructive hydrocephalus [Figure 1b and c]. On T1-weighted imaging, the mass was hypointense and nonenhancing. On T2-weighted imaging, the mass was hyperintense. On fluid attenuated inversion recovery (FLAIR) and diffusion weighted imaging (DWI), the mass showed some restriction within the lesion with marked hyperintense signal [Figure 2a-c]. His serum α-fetoprotein, β-human chorionic gonadotropin (β-HCG), and placental alkaline phosphatase were...
within normal limits. Our pre-op diagnosis was pineal epidermoid. The patient was positioned in three-quarter prone position. We approached through occipital-transtentorial route. Using microsurgical techniques, we explored the pineal region. Just in front of thickened arachnoid in the quadrigeminal cistern, there was a pearly tumor [Figure 3a]. 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 near-total removal was achieved without any injury to neural or vascular structures [Figure 3b]. We had to leave in situ a tiny fragment of the capsule that was adherent to the vein of Galen. Postoperative period was uneventful. Patient improved neurologically. Histopathology confirmed it as a case of epidermoid [Figure 3c]. On post-op CT scan, there was minimal residual tumor and hydrocephalus decreased [Figure 3d].

**DISCUSSION**

Epidermal cysts arise when ectodermal remnants are trapped by two fusing ectodermal surfaces. Trauma and differentiation from multipotential cell rests are other mechanisms implicated in the origin of epidermoid tumor. Growth of epidermoid occurs at a linear rate as that of normal skin; because of slow growth rate, these tumors attain relatively large size before the patient becomes symptomatic. These tumors are commonly located within the basal subarachnoid cisterns. Extension of the tumor both above and below the tentorium is frequent.[1] The tumor is known for its potential to open the anatomical spaces in the region and the encasement of the major arteries and the perforators coursing within the cisterns.[2]

On T1- and T2-weighted images, these tumors have signal intensity as that of CSF, mimicking arachnoid cyst.[3] DWI is the best test to differentiate epidermoid from arachnoid cyst. Epidermoids show hyperintense
Figure 3: (a) Glistening tumor in quadrigeminal cistern. (b) Tumor displaced major vessels downward. (c) Histopathology showing keratin in layers. (d) Post-op CT scan showing minimal residual tumor and decreased hydrocephalus

signal in DWI, whereas arachnoid cyst shows hypointense signal. Contrast MRI also gives an idea about tumor relation to major vessels. In our case, the tumor had displaced the major vessels downward.

Surgeries for pineal region tumors are done to get sufficient tissue sample for histological diagnosis, relieving hydrocephalus and mass effect.\(^4\) Adjuvant therapy depends upon histological diagnosis. Only for germ cell tumors, depending upon elevated tumor markers, radio-or chemotherapy may be started.

Choice of operative approach depends upon the extent of tumor and its relation to major vessels. As in our case the tumor had a major supratentorial part with displacement of major vessels downward, we chose occipital–transtentorial route.

**CONCLUSION**

Though germ cell tumors constitute a significant portion of pineal region tumors, still 35–50% of pineal tumors are benign, which are radio-resistant. Therefore, tissue diagnosis of pineal region tumor is mandatory for optimal management of these tumors. Most cases of pineal tumors present as obstructive hydrocephalus. They either require pre- or postoperative ventriculoperitoneal (VP) shunt. If properly planned, many benign pineal tumors may be successfully excised and, most importantly, postoperative VP shunt could be avoided.

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