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Case Report

A rare and challenging case of pineal gland tumor – A case report

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ABSTRACT

Pineal gland tumors are of rare occurrence and may arise from pineal parenchymal cells, the neighboring glia or residual stem cells. Due to its rarity pineal gland tumors are often misdiagnosed. The World Health Organsation (WHO) classifies and grades pineal parenchymal tumors from grade I to grade IV. We present a case report of a rare pineal parenchymal tumor (PPT) in an adult female which was diagnosed mainly on histopathology and aided by immunohistochemistry. The case report includes review of histopathological features and grading of pineal region tumors of intermediate malignancy which is necessary for further management of such cases.

1. Introduction

The pineal glands can show varied lesions ranging from pineal cysts, aneurysms, cavernous vascular malformations, arteriovenous malformations and tumors. Pineal region tumors account for less than 1% of intracranial tumors and are very rare in occurrence.[1] Pineal region tumors are classified as germ cell tumors, pineal parenchymal tumors and those derived from adjacent anatomical structures. Germinoma is the most common pineal tumor in Asian population.[2]

Pineal parenchymal tumors (PPT) constitute 15 to 30% of pineal region tumors.[3] The tumors range from well differentiated neoplasm with presence of pineocytomatous rosettes to poorly differentiated malignant embryonal neoplasm occurring within first two decades of life. We present a case of intermediate differentiation with variable biological behaviour and may be classified according to WHO as grade II or III.

2. Case History

A 54-year-old female presented with history of skid and fall from bike. Magnetic resonance imaging (MRI) was done which showed a lesion in the pineal region but the patient was lost for follow up. Nine months later patient presented with bilateral watering of eyes and repeat MRI showed pineal region tumor for which surgery was advised and total excision was performed. The specimen was received in the department of histopathology of our hospital.

Grossly nodular, dull white soft tissue 3 x 2 cm with few tiny fragmented bits of 0.2 x 0.2 cm was received. Cut Section was pale white and was all embedded.

The histopathology showed a well circumscribed vaguely lobulated tumor having large round nucleus, mild to moderate atypia separated by vascular channels. Also, tumor cells in sheets with background of fibrillary matrix was noted. These cells had perinuclear clearing. Few bi and multinucleated cells was noted. Focal areas showed atypia with hyperchromatic elongated nucleus. 3 to 5 mitotic figures / 10 hpf was seen. Peripheral normal brain tissue was noted.

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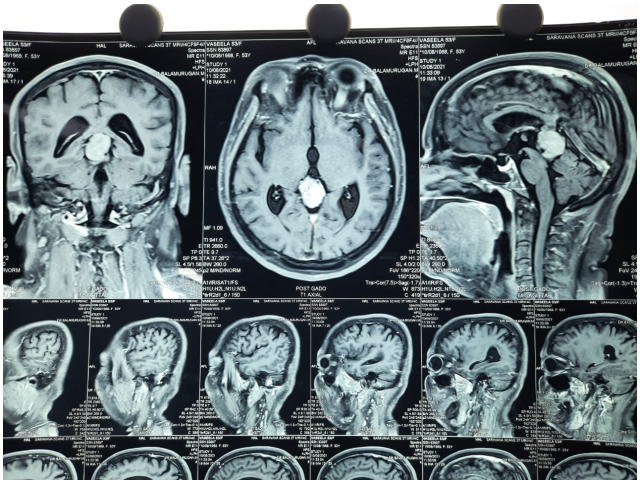

Fig. 1: MRI brain showing pineal region tumor.

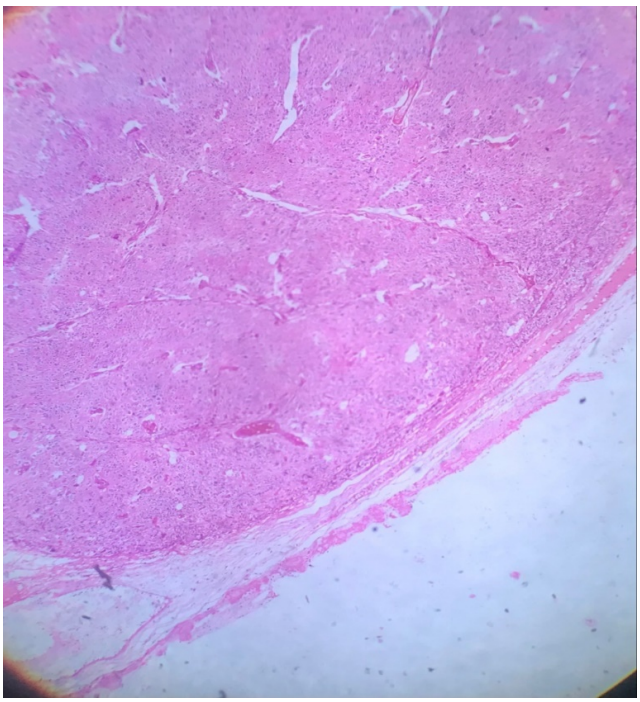

Fig. 2: Low power view of a well circumscribed lobulated tumor in pineal region. H&E stain 200x

Immunohistochemistry (IHC) was carried out with the following markers:

The IHC results was consistent with tumor of pineal origin. Correlating with H and E picture final diagnosis of pineal parenchymal tumor of intermediate differentiation, grade II was given. Adjuvant radio / chemotherapy was not given. The patient is on follow up and is recurrence free two years post surgery.

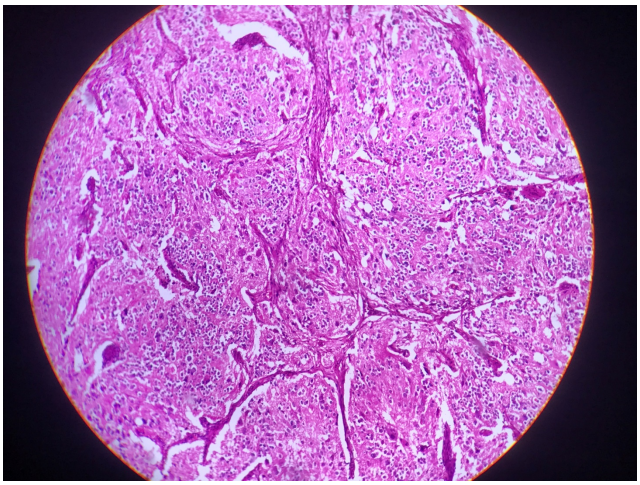

Fig. 3: Lobulated tumor having intermediate to large round nucleus separated by vascular channels. H&E stain 400x

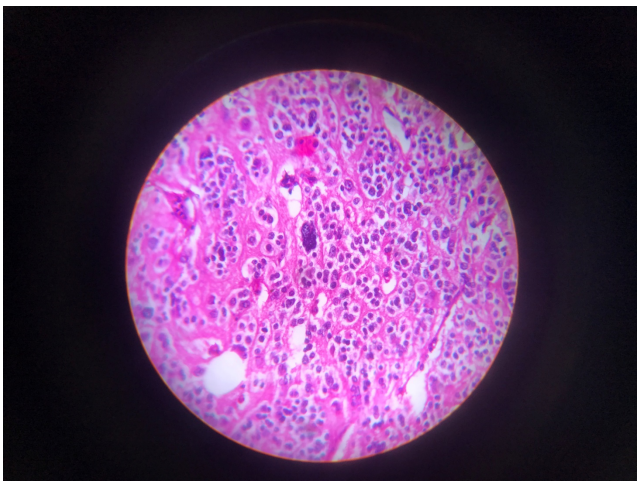

Fig. 4: Neurocytoma-like appearance in a pineal parenchymal tumour of intermediate differentiation. Tumour with moderate cellularity and round nuclei harbouring salt-and-pepper chromatin. H&E stain 400x

Table 1: IHC markersand result.

Marker	**Result**
EMA	Negative
GFAP	Negative
S100	Positive in few large cells and few small cells
Olig-2	Negative
Synaptophysin	Positive
CD56	Positive
Chromogranin A	Positive (dot like)
Ki67	3 to 5%

3. Discussion

The pineal gland got its name as "third eye" due to its pine cone shape and is situated in the epithalamus and its function is to modulate circadian rhythm through the production of melatonin.[4] Pineal gland parenchymal tumors include pineocytoma grade I, pineal parenchymal tumor of uncertain differentiation (PPTID) grade II or grade III, pineoblastoma grade IV, papillary tumor of the pineal region grade II or III and new addition in this group is Desmoplastic myxoid tumor of the pineal region, SMARCB1-mutant according to fifth edition of the WHO Classification of Tumors of the Central Nervous System (CNS), published in 2021.[5] All pineal tumors need surgery to obtain a tissue diagnosis and hence to reduce the symptoms caused due to impaired cerebrospinal flow from third ventricle into the cerebral aqueduct.[6] PPTIDs account for 45% of all pineal parenchymal tumors and the mean age at diagnosis of PPTID is around 41 years, although all ages can be affected.[7] It was first described by Schild et al in 1993.[8]

Patients usually present with signs of increased intracranial pressure like headache, vomiting secondary to obstructive hydrocephalus. Other symptoms are ataxia, Parinaud syndrome, or diplopia.[9]

On microscopy PPTID can exhibit two architectural patterns – Diffuse or lobulated with vessels delineating the lobules. The PPTIDs are moderately to highly cellular tumors with neoplastic cells displaying round nuclei, salt and pepper chromatin showing mild to moderate atypia. The cytoplasm of the cells is easily distinguished compared to pineoblastoma. The pleomorphic variant shows bizarre ganglioid cells with single or multiple atypical nuclei and abundant cytoplasm. The mitotic count is variable and range from 3.5 to 16.1%.[10]

According to study done by Jouvet et al, PPT with intermediate differentiation can be divided in two grades: Grade II showed lobulated, transitional, and diffuse PPT with strong immunolabeling for neurofilament and less than 6 mitoses and grade III was a lobulated and diffuse PPT with less than or more than 6 mitoses but without immunostaining for neurofilament.[11]

In the same study, four different morphological subtypes of PPTID were identified by Jouvet et al.

1. Lobular arrangement of cells with an endocrine vascularity.
2. Diffuse proliferation mimicking oligodendroglioma or neurocytoma
3. Lobulated and/or diffuse areas associated with other areas containing pineocytomatous rosettes corresponded to transitional forms.
4. Distinctly biphasic pattern including areas of typical pineocytoma and pineoblastoma.

Our case also showed vaguely lobulated and also focally sheets of tumor cells with round nucleus displaying salt and pepper chromatin. Intervening vascular channels dividing into lobules was seen. The mitotic index was low.

Immunohistochemistry of PPTID shows synaptophysin positivity, variable positivity for Neurofilament and chromogranin. S100 is expressed in ganglion cells of pleomorphic variant.

PPTIDs have better prognosis due to its localized presentation. They have a potential for local recurrence and craniospinal dissemination. Histologic grade, PPTID subtype, extent of resection, and neuraxis spread at diagnosis are important factors for outcome in PPT.[12] The main treatment option is microsurgery and is the standard modality, alternatives include stereotactic radiosurgery. For selected cases, adjuvant treatment to surgery also holds promise.

4. Conclusion

PPTID is of intermediate differentiation between pineocytoma and pineoblastoma. The biological behavior is variable and definite histological grading criteria is not defined in WHO. The extent of resection, histologic grade and craniospinal dissemination at presentation are important factors for outcome of pineal parenchymal tumors.

5. Conflict of Interest

None.

6. Source of Funding

None.

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