Case report

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Papillary Tumor of Pineal Region: Clinical Manifestations, Histopathological Insights, and Immunohistochemical Profile: A Comprehensive Case Report

Shreekant Bharti[1], Avinash Singh[2], G Guralarasan[3], Punam Prasad Bhadani[4], Madhu Kumari[5]

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Abstract

Introduction: Papillary tumor of the pineal region (PTPR) is a unique rare neuroepithelial tumor arising from unique ependymocytes of the subcommissural organ, different from the pineal gland Affecting less than 1% of adult intracranial tumors, PTPR presented unique diagnostic challenges due to rarely and because of its nature papillary pattern does.

Case presentation: We report a 25-year-old man initially treated for acute hydrocephalus and later diagnosed with PTPR. Radiologic examination revealed a typical pineal area mass, and histopathology examination confirmed PTPR with prominent papillary structures. Immunohistochemistry played an important role in supporting the disease.

Conclusion: This article outlines a complex approach to the diagnosis of PTPR, including radiology, histopathology, and immunohistochemistry. Furthermore, it highlights the uniqueness of PTPR in pineal-parenchymal tumor classification, as outlined in the WHO classification for 2021. This report contributes to the paucity of literature on this rare tumor, and highlights comprehensive diagnostic approach when evaluating pineal region masses.

Keywords: Brain neoplasms; Papillary tumors of the pineal region; Pineal parenchymal tumor.

Author Affiliation: [1]Additional Professor, [2,5]Assistant Professor, [3]Senior Resident, [4]Professor and Head of Department, [5]Assistant Professor, Department of Pathology, All India Institute of Medical Sciences, Patna 801507, Bihar, India.

Corresponding Author: Avinash Singh, Assistant Professor, Department of Pathology, All India Institute of Medical Sciences, Patna 801507, Bihar, India.

E-mail: alpana.jain@gmail.com

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INTRODUCTION

Papillary tumour of pineal region (PTPR) is a rare neuroepithelial tumour formed by specialised ependymocytes of the subcommissural organ found in the lining of the posterior commissure rather than the pineal gland itself.[1] PTPR is highly rare, accounting for less than 1% of all adult intracranial tumors.[2] Tumors developing in the pineal area may have a papillary pattern, posing a diagnostic difficulty on imaging and histological findings.We present a case of a 25-year-old man who was initially operated on for acute hydrocephalus and then referred to our hospital for further investigation.

CASE PRESENTATION

Appropriate protocols were followed and patient informed consent was obtained. We present the case of a 25-year-old man who complained of severe nausea, headache and vomiting for one year. He underwent surgery for hydrocephalus. Follow-up NCCT Head revealed a well-defined hypodense lesion with an average attenuation of 17 HU, with areas of hyperdensity with an average attenuation of 48 HU involving the pineal region. The lesion is seen abutting the left thalamus, and inferiorly, the lesion is seen involving the central region of the midbrain, causing obliteration of the aqueduct. The lesion measures 4.0 x 3.5 x 3.0 cm (CCxAPxTR) likely dysgerminoma. The presence of an extracranial drainage tube is seen traversing through the defect in the right parietal bone, with its tip lying in the frontal horn of the left lateral ventricle. After 6 months of MRI brain plain and contrast with MR spectroscopy, a large, measuring 4.4X3.0X4.6 cm (APxTRxCC) well-circumscribed solid cystic mass was seen epicentred in the pineal gland and third ventricle region. The mass is occupying the third ventricle and reaching up to the foramen of Monro superiorly. The solid soft tissue component appears to be T1/T2 isointense and T2 FLAIR hyperintense. On post-contrast, avid enhancement of the solid component is seen. Impression of a likely pineocytoma (Fig. 1). Histopathology revealed a cellular tumour arranged in papillae, sheets, and pseudorosettes. These tumour cells are cuboidal to columnar, have mild pleomorphism, round to ovoid nuclei, fine chromatin, and moderately eosinophilic to occasional vacuolated cytoplasm (Fig. 2A & 2B). An immunohistochemical panel was applied. The tumour showed moderate to strong diffuse positivity for NSE (Fig. 2C) and CK8/18 (Fig. 2D). Ki67 is 3–5% (Fig. 2E). The tumour is negative for GFAP (Fig. 2F), EMA (Fig. 2G), chromogranin (Fig. 2H), and E-cadherin (Fig. I). Final diagnosis given was papillary tumour of pineal region.

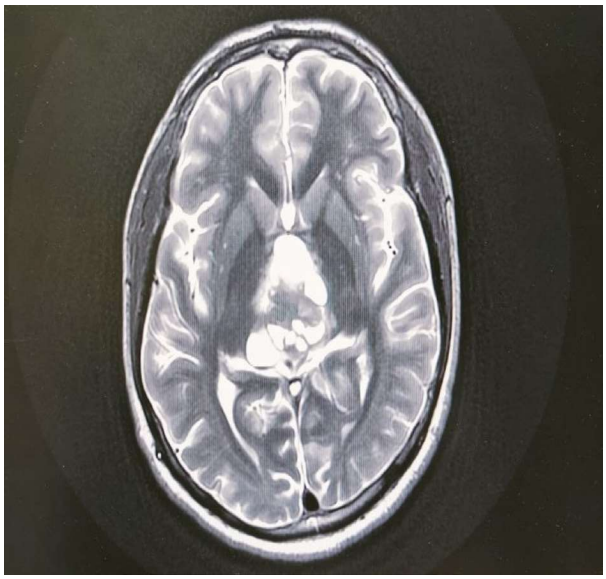
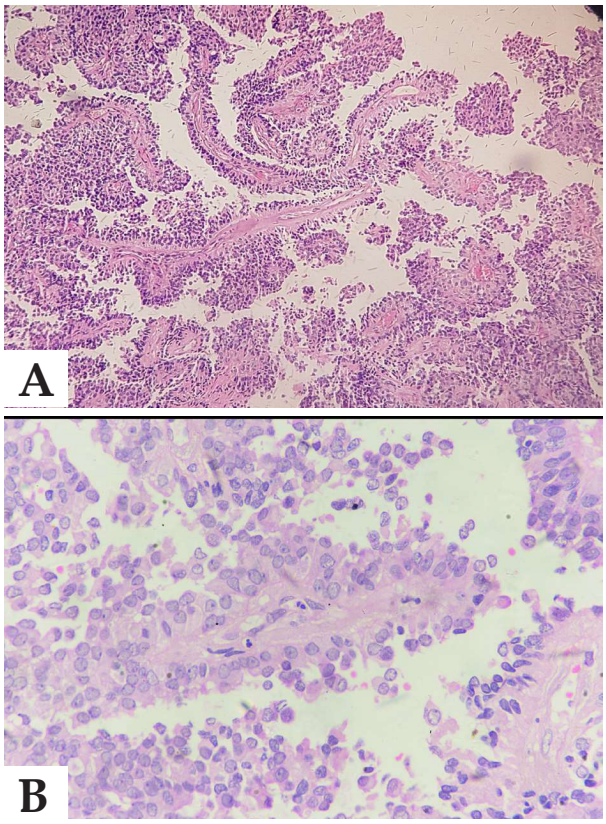


Fig. 1: Photomicrograph showing MRI brain contrast with MR spectroscopy, a large, measuring 4.4X3.0X4.6 cm (APXTRXCC) well-circumscribed mass was seen epicentred in the pineal gland and third ventricle. On post-contrast, avid enhancement of the solid component is seen.



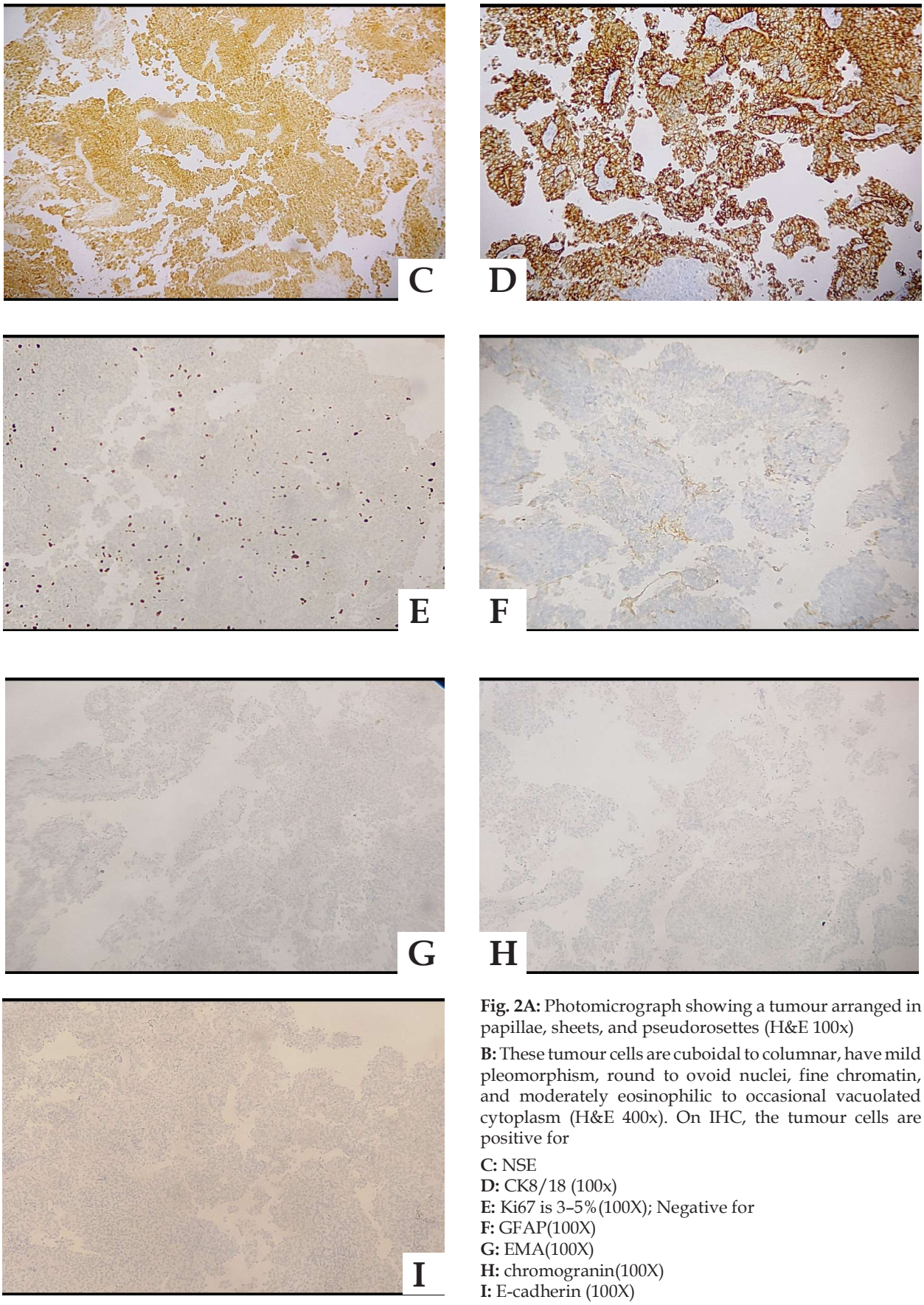


Fig. 2A: Photomicrograph showing a tumour arranged in papillae, sheets, and pseudorosettes (H&E 100x)

B: These tumour cells are cuboidal to columnar, have mild pleomorphism, round to ovoid nuclei, fine chromatin, and moderately eosinophilic to occasional vacuolated cytoplasm (H&E 400x). On IHC, the tumour cells are positive for

C: NSE
D: CK8/18 (100x)
E: Ki67 is 3–5%(100X); Negative for
F: GFAP(100X)
G: EMA(100X)
H: chromogranin(100X)
I: E-cadherin (100X)

DISCUSSION

The WHO 2021 classification of tumors of the central nervous system (fifth edition) differentiates five histological types of pineal parenchymal tumors including pineocytoma, pineal parenchymal tumors of intermediate differentiation, PTPR, pineoblastoma, and desmoplastic myxoid tumor of the pineal region, SMARCB1-mutant. Arabic (rather than Roman) numerals are utilized for grading within the types.[2] The term "PTPR" is based on the histopathological description of a tumor characterized by a papillary pattern, rosettes, and pseudorosettes. Microscopic evaluation often demonstrates a lesion with papillary areas lined by epithelioid tumors with eosinophilic cytoplasm, and numerous cells exhibiting clear or vacuolated cytoplasm. Evaluating mitotic and proliferation index were important in grading of PTPR as Grade 2 or 3.[2,3] This strange case justifies the complex process to accurately diagnose this rare tumor involving imaging, histopathology, and immunohistochemistry. As reported in the literature, PTPRs are usually masses larger than 3cm and well-defined and may have cystic features. The image shows enhanced contrast and decreased attenuation on CT scan. MRI shows hyperintensity on T2-weighted sequences.[3] This description corresponds to the findings of our patient. Because of its papillary and epithelioid appearance, PTPR is similar to other papillary-like malignancies, such as ependymoma, pineal parenchymal tumours, choroid plexus neoplasms, and adenocarcinoma metastases. To differentiate it, immunohistochemistry analysis is essential.[4,5] Immunohistochemical staining profiles were investigated in a series of 15 PTPR. In addition to cytokeratin, synaptophysin and glial fibrillary acidic protein expression, PTPR were examined for the presence of dot or ring-like epithelial membrane antigen (EMA The most distinctive immunohistochemical feature of PTPRs is their reactivity to keratin (especially CK18). PTPR also express vimentin, NSE, CD56 and transthyretin.[4,6] Immunohistochemical staining profiles were investigated in a series of 15 PTPR. In addition to cytokeratin, synaptophysin and glial fibrillary acidic protein expression, PTPR were examined for the presence of dot- or ring-like epithelial membrane antigen (EMA Unlike PTPR, ependymoma are often express GFAP and lack CK18 expression.[1] E-cadherin is commonly expressed in choroidal plexus tumors that are negative in PTPR.[7,8]

CONCLUSIONS

The present case exemplifies the spectrum of papillary tumors that may be considered as distinct in cases presenting with a pineal mass with proper radiological setting. The clinical presentation of a patient with PTPR and the various immunohistochemical expressions used to confirm the diagnosis are presented in our case report.

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