

## Papillary Tumor of Pineal Region: A Case Report

Ahmed Khaled<sup>1</sup>, S.M Mahbulul Alam<sup>2</sup>

1. Sr. Consultant, Histopathology Lab, Evercare Hospital Dhaka
2. Sr. Consultant, Histopathology & Coordinator Lab Medicine, Evercare Hospital Dhaka

### Address for Correspondence:

Dr. Ahmed Khaled  
Sr. Consultant,  
Histopathology Lab, Evercare Hospital Dhaka.  
ahmed.khaled@evercarebd.com

### ABSTRACT

#### Abstract

Pineal region tumors constitute about 0.4%-1% of intracranial tumors in adults. These tumors may arise from pineal gland itself or structures around it which are all termed as pineal region tumor. Papillary tumor of the pineal region is a non-parenchymal tumor of the pineal region. First described in 2003, it is an extremely rare tumor. We have presented a case of 27-year-old female patient presented as a pineal tumor. After successful excision, the tumor proved to be Papillary tumor of Pineal region (PTPR) both histomorphologically and immunohistochemical study.

**Keywords:** Pineal region tumors, Papillary tumor, histomorphology, immunohistochemistry.

### INTRODUCTION

Papillary tumor of pineal region (PTPR) is defined as "A neuroepithelial tumor localized in the pineal region and characterized by a combination of papillary and solid areas with epithelial like cells and immunoreactivity for cytokeratin (especially CK18)<sup>1</sup>.

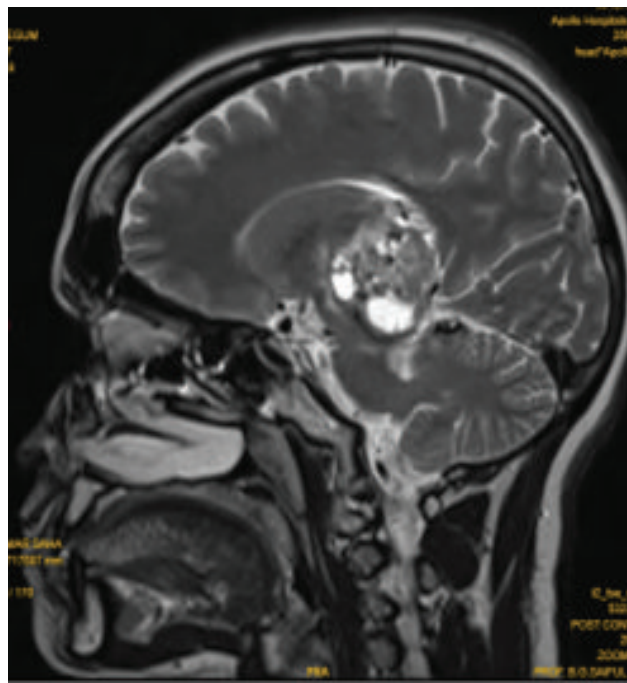
PTPR is a newly recognized distinct entity in the 2007 WHO nomenclature. This rare tumor is thought to possibly arise for the specialized ependyma of the subcommisural organ<sup>2</sup>. Tumors of the pineal region are rare lesions, accounting for only 1% of all intracranial tumor. PTPR have morphological features in common with a number of other papillary like tumors that occur in pineal region including pineal parenchymal neoplasm, choroid plexus papilloma, Papillary ependymoma, metastatic papillary carcinoma, papillary meningioma and germ cell neoplasm<sup>2</sup>. Therefore, distinction between PTPR and other papillary tumor is important from treatment and prognostic point of view.

This tumor is very rare therefore proper treatment and prognosis are not well documented and remain controversial. A study showed 5-year survival estimate of 73% by Kaplan- Meier analysis<sup>3</sup>. In this article, we have reported the case of a 27-year-old female with PTPR to share our experience with others.

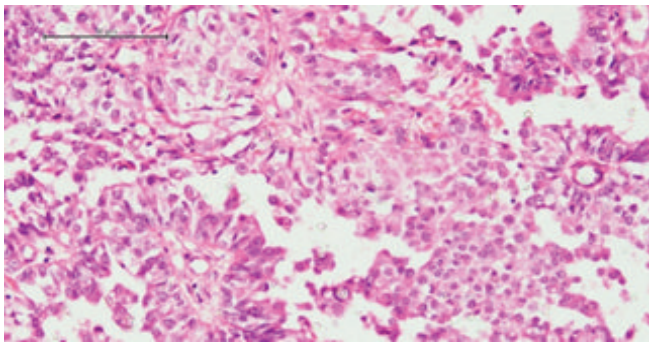
### CASE REPORT

A 27-year-old female patient got admitted through neurosurgery outpatient department of Evercare Hospital with the complaints of progressive weakness of right side of the body for 6 months, unable to sit and standup without support for same duration. On examination her GCS was E4V4M6, pupil was 2mm equal

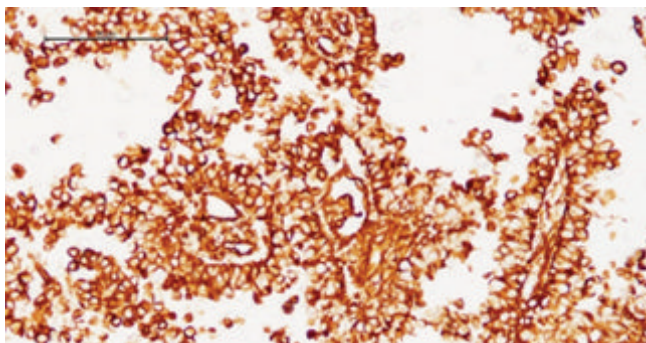
and reactive to light, decreased hearing on right side, pulse 102/min, BP was 110/70, respiratory rate 18/min, Temp 98° F and Spo2 wall 98%. Abdominal ultrasound detected no significant abnormality. Contrast MRI and CT scan of brain reveal a mass in the pineal region measuring 3.9x3.1x3 cm (Fig:1) and the mass was excised. Histopathological examination reveals proliferation of epithelioid appearing cells arranged mostly in papillary pattern with fibrovascular core and focally solid sheet like appearance. Mitosis is occasional and no necrosis is seen. (Fig:2).



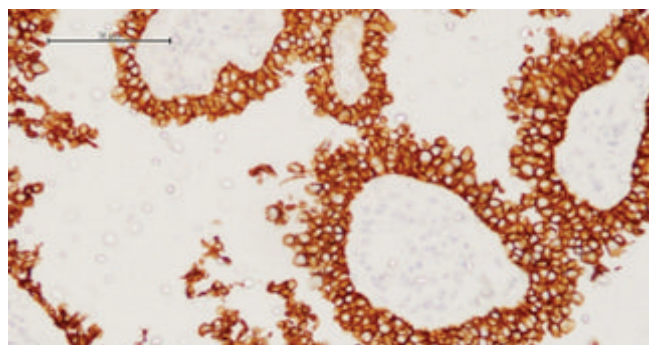
**Fig 1:** MRI Brain (sagittal section T2 WI) reveals large tumor at the pineal region.



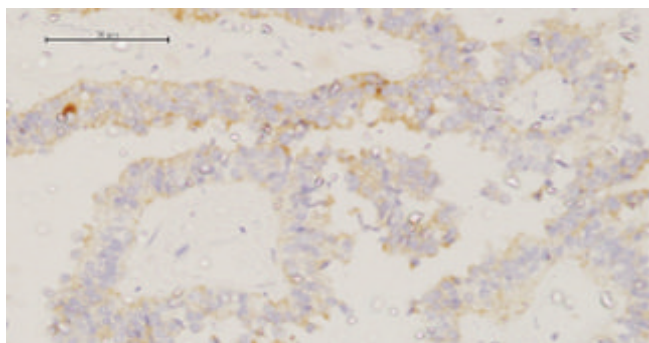
**Fig 2:** papillary structure with fibrovascular core lined by cuboidal to columnar cells in PTPR (HE stain ×400)



**Fig 3 (a):** strong immunoreactivity for cytokeratin



**Fig 3(b):** strong immunoreactivity for vimentin



**Fig3 (c):** strong immunoreactivity for CD 56

Immunohistochemically these tumor cells show strong expression for Pancytokeratin, Vimentin, CD56, NSE, S100, (Fig 3: a,b,c) and were negative for GFAP, SALL 4, CHROMOGRANIN. Proliferative index was low (2%). Both histomorphologically and immunohistochemically a diagnosis of papillary Tumor of the pineal region (PTPR) was made.

**DISCUSSION**

Papillary tumor of pineal region is a very rare and newly described entity first included in the WHO-2007 classification of brain tumor. Origin of this tumor is not known definitely; however, it is postulated to have originated from specialized ependymal cells of the subcommissural organ<sup>4</sup>.

These tumors are usually well circumscribed, and the size may vary from 2.5 to 4 cm. There is variable age range from very early childhood to old age, but mean age is 32 years and mostly common in young people. There is not much difference in frequency between male and female<sup>4</sup> In CT imaging these lesions are hypodense in nature and show enhancement with contrast. MRI shows hypointense in TI weighted (TIW) sequence with contrast enhancement. Symptoms of the patient are mostly related to obstructive hydrocephalus due to compression of cerebral aqueduct. Tumor progression usually seen in almost 75% of the patients<sup>4</sup>.

Immunohistochemistry plays a vital role in differentiating papillary tumor of pineal region from tumors of similar morphology, which includes pineal parenchymal tumor, papillary ependymoma, choroid plexus tumor, papillary meningioma, and metastatic papillary tumor. Prominent expression of neuronal marker differentiates from pineal parenchymal tumor, absent or low CK7/CK20 and low Ki-67 is helping differential from metastatic tumor of similar morphology. Papillary meningioma dose not express cytokeratin but PTPR does. However choroid plexus papilloma shows cytokeratin and transthyretin expression similar to PTPR. MAP-2 immunomarker is needed to differentiate between these as PTPR show MAP-2 expression<sup>4</sup>.

As the reported cases of PTPR are very few, there are no standard treatment protocol except gross total resection<sup>3</sup>. Radiotherapy has role in adjuvant setting with cases with high risk of local recurrence<sup>6</sup>. PTPR are characterized by frequent local recurrence but only occasional spinal dissemination and no metastasis<sup>4</sup>.

The real challenge of surgery and radiotherapy is as these treatment modality targets pineal region and neighboring critical anatomical structures; post treatment cognition defects must be considered. Besides most of the patients are young, so long term effect of treatment and quality of life must be carefully evaluated. Multidisciplinary treatment for pineal region tumors is best option<sup>5,6</sup>.

We have described a case of PTPR in a young patient in this case report. This case of PTPR has similar clinical characteristics, morphology and immunohistochemical profile described in other literatures<sup>2,3,4,5</sup>.

### **CONCLUSION**

PTPR is a rare CNS tumor which needs more detailed case study to understand the prognosis and optimization of treatment protocol.

### **REFERENCES**

1. Louis DN, Ohgaki H, Wiestler OD, Cavence WK, eds. World Health Organization classification of Tumors of the central Nervous system (Revised 4th ed). Lyon: International Agency for Research on Cancer (IARC): 2016.
2. Hua X, Yang P, Zhang M, Zhao Y and Wang B. Papillary Tumor of the pineal region: A case report and review of the literature. *Experimental and Therapeutic Medicine* 10:1375-1379, 2015.
3. Fèvre-Montange M, Hasselblatt M, Figarella-Branger D, et al. Prognosis and histopathologic features in papillary tumors of the pineal region: a retrospective multicenter study of 31 cases. *Journal of neuropathology and experimental neurology*. 2006;65(10):1004-1011.
4. Patil M, Karandikar M. Papillary tumour of pineal region: A rare entity. *Med J DY Patil Univ* 2013; 6:302-4.
5. Zaimoglu M, Yakar F, Dogan I, Caglar YS. Papillary tumor of the pineal region. *Int Surg J* 2017;4(12):4076-8.
6. Lancia A, Becherini C, Detti B, et al. Radiotherapy for papillary tumor of the pineal region: A systematic review of the literature. *Clinical Neurology and Neurosurgery*. 2020; 190:105646.