

Papillary Tumor of Pineal Region in a 5-Year-Old Male Child: A Rare Entity

Abstract

Papillary tumor of the pineal region (PTPR) is a rare grade II to III pineal lesion. These tumors mostly occur in adults, only rarely in children, with six cases in children under the age of 16 years (10.2%) up to now. We report the case of a 5-year-old male child presenting with worsening headaches, abnormally enlarged head since birth and visual disturbances. Imaging reveals a mass in the region of the pineal gland. The third and lateral ventricles were enlarged. The patient underwent a gross-total surgical resection of pineal mass through a suboccipital supracerebellar approach and tissue sent for histopathological examination and an available immunohistochemical workup has been done which confirmed the diagnosis of papillary tumor pineal region. This case highlights the histopathological features, imaging along clinical presentation similar to those in the original description of this rare entity PTPR. More studies are required to determine the prognosis and standard treatment protocol of this rare entity.

Keywords: Malignant tumor, papillary tumor of the pineal region, pineal gland

Introduction

Papillary tumors of the pineal region are located in the pineal gland which is located in the center of the brain. Papillary tumors of the pineal region (PTPRs) were first described by A. Jouvét *et al.* in 2003 and were introduced in the World Health Organization (WHO) classification of central nervous system (CNS) in 2007. Papillary tumors of the CNS and particularly of the pineal region are very rare, and therefore, diagnosing them is extremely difficult.^[1]

Case Report

A 5-year-old male child presented with worsening headaches, abnormally enlarged head since birth, and visual disturbances of several months duration. Noncontrast computed tomography (CT) revealed a 2.5 cm × 2.8-cm mass near the posterior aspect of the third ventricle, which resulted in obstructive hydrocephalus. The third and lateral ventricles were enlarged. Brain magnetic resonance imaging (MRI) was subsequently performed. A precontrast sagittal T1-weighted image showed a mass in the region of the pineal gland, which was isointense on T1 and T2-weighted images. The mass was enhancing avidly

on contrast [Figure 1]. The patient underwent a gross-total surgical resection of pineal mass through a suboccipital supra-cerebellar approach. The patient tolerated the procedure well and tissue was sent for histopathological examination and diagnosed papillary tumor pineal region which was further confirmed on immunohistochemical. Histopathologically, the tumor displayed distinctive features described for PTPR. This tumor was an epithelial-appearing lesion with focal papillary architecture, as well as other densely cellular areas that showed ependymal-like differentiation. Within the cellular areas, true rosettes and tubules were identified, consisting of cells with clear or vacuolated cytoplasm. Nuclei were round to oval with stippled chromatin in the papillary sites, vessels were layered with large, pale to eosinophilic columnar cells. Mitotic activity is reported to vary from 4 to 10 per 10 HPF, and necrotic foci were identified. Occasionally, PAS-positive islands were found. While microvascular proliferation was absent, vessels were hyalinized with a clear demarcation between the tumor and the residual normal pineal gland [Figures 2 and 3].

The evolution in the classification of pineal tumors has led to the emergence of PTPR.

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Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_81_19

Quick Response Code:



How to cite this article: Gupta K, Khursheed S, Nayil K, Khursheed S, Makhdoomi R. Papillary tumor of Pineal Region in a 5-year-old male child: A rare entity. Asian J Neurosurg 2021;16:824-6.

Submitted: 13-May-2019

Revised: 22-Jun-2019

Accepted: 19-May-2021

Published: 18-Dec-2021

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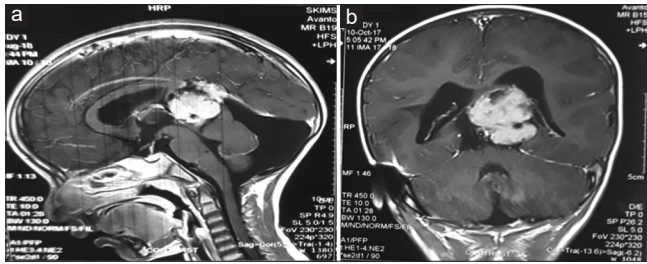


Figure 1: (a) Sagittal T1-weighted contrast magnetic resonance imaging showing a mass in the region of the pineal gland, with a thin rim of intrinsic signal hyperintensity. (b) Coronal T1-weighted contrast magnetic resonance imaging showing a mass in the region of the pineal gland, with a thin rim of intrinsic signal hyperintensity

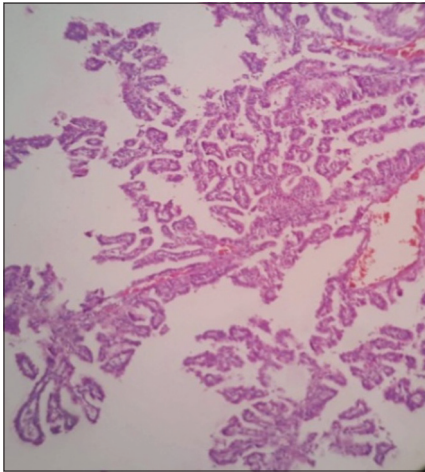


Figure 2: Hematoxylin and Eosin stain in Papillary tumor of the Pineal Region (x400) Extensive papillary formation lined by cuboidal to columnar cells with maintained basal polarity with mild to minimal atypia

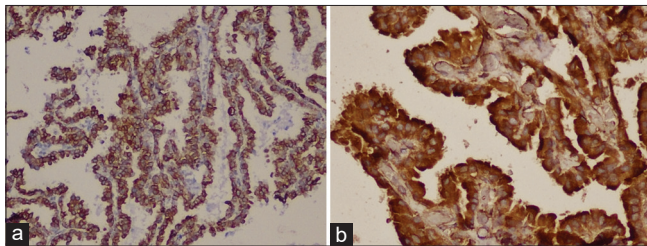


Figure 3: (a) Cytokeratin 18 Positivity in cells on immunohistochemistry. (b) Epithelial membrane antigen positivity in cells on immunohistochemistry

Discussion

Based on a series of six tumors with identical histological features, PTPR was first described as a distinct entity in 2003.^[2] To date, there are now 59 reported cases, including six cases. In children under the age of 16 years (10.2%),^[3,4] PTPR generally presents with nonspecific symptoms, including headache and visual disturbances due to obstructive hydrocephalus.^[5] Neuroimaging usually displays a large (2–4 cm), well-circumscribed contrast-enhancing tumor, which occasionally has cystic elements.^[6,7] Previous MRI literature has reported a heterogeneously enhancing mass in the pineal region.^[2,8] PTPR is prone to local recurrence, even after surgery chemotherapy,

or radiotherapy. PTPR is now viewed as a separate entity from choroid plexus papilloma and ependymoma, although there is significant overlap.^[5] PTPR displays prominent decoration with cytokeratins, which are most characteristically seen in papillary structures. CK18 is the cytokeratin most often associated with PTPR, although KL1, AE1/AE3, and CAM 5.2 also commonly decorate these tumors. Due to the rarity of PTPRs, no set positive or negative cutoffs have been established for these markers.^[5] In PTPR, staining is also seen with NSE and S-100. Focal decoration with chromogranin, synaptophysin, and EMA is also described. In contrast to the strong GFAP decorations in ependymomas, GFAP immunoreactivity in PTPR is usually focal or absent.^[8,9] Choroid plexus tumors frequently stain for membranous Kir 7.1 or cytoplasmic stanniocalcin-1; however, these two markers are absent in the majority of PTPR.^[5] Transthyretin is more often expressed in choroid plexus tumors as well. Because of the few reported cases of PTPR, there are no current standard treatment options beyond gross total resection. New areas of interest in the diagnosis and prognosis of PTPR continue to develop as pathologists further delineate this newly described entity. Measurements of melatonin levels, as well as the elucidation of the enzymes in melatonin synthesis of pineal tumors, have been recently explored as possible diagnostic and treatment indicators. As more pathologists have become familiarized with the cytological presentation of PTPR, the ability to diagnose the tumor on intraoperative smear preparations has increased. By combining observations such as smear population, architecture, background, calcification, single cells, cell morphology, location, and radiology, the addition of cytologic imprints can help improve the intraoperative diagnostic power over frozen section alone.^[10]

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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