

## Papillary Ependymoma of the Spinal Cord: A Case Report with Summary of Prior Published Cases

### Abstract

Papillary ependymoma is a rare variant of ependymoma. It has been included in Grade II tumors of updated 2016 WHO classification of central nervous system tumors. Only a handful of cases of spinal papillary ependymomas have been reported so far. The differential diagnoses include choroid plexus papilloma, papillary meningioma, metastatic carcinoma, and papillary tumor of the pineal region. Here, we take the opportunity of reporting a rare case of spinal papillary ependymoma along with its squash cytological features and a summary of prior published cases.

**Keywords:** *Ependymoma, papillary ependymoma, spinal tumors, squash cytology*

### Introduction

Spinal canal lodges 10% of ependymomas with majority of them being classic ependymomas or myxopapillary ependymomas. Papillary ependymoma is a rare ependymoma that mainly affects the brain, and only three cases of spinal papillary ependymomas have been reported so far.<sup>[1-3]</sup> The updated 2016 WHO classification of central nervous system (CNS) tumors continues to place it in Grade II category. Its main differential diagnoses include choroid plexus tumors, metastatic papillary carcinoma, papillary meningioma, and papillary tumors of pineal region (PTPR).<sup>[4]</sup> Here, we take the opportunity of reporting a rare case of spinal papillary ependymoma including its immunohistochemistry (IHC) and squash cytology findings.

### Case Report

A 40-year-old male patient presented with back pain and focal sensory and motor deficits. Magnetic resonance imaging (MRI) spine revealed a well-circumscribed intramedullary space-occupying lesion in C6-D2 region with contrast enhancement [Figure 1a]. Complete excision of the tumor was done.

Intraoperative squash cytology was performed. The smears were cellular and composed of cohesive clusters of round to oval cells with bland nuclei and salt and

pepper type of chromatin. At places, the tumor formed large papillary structures with fibrovascular core having cells attached to blood vessel in multiple layers. No necrosis or mitosis was detected. Atypia was absent. In some places, thick bottle brush appearance was noted [Figure 1b-d].

Based on these findings and MRI, a diagnosis of ependymoma was suggested.

The histopathological sections showed well-formed papillae and finger-like projections lined by single or multiple layers of tumor cells. Several perivascular pseudorosettes and ependymal canals were noted. The tumor cells were monopolar and cuboidal or columnar in shape with a moderate amount of cytoplasm and bland nuclei having speckled chromatin. A smooth surface was formed by the apical surfaces of the tumor cells. The tumor lacked nuclear hobnailing. There was no evidence of necrosis, microvascular proliferation, endothelial proliferation, or mitosis. The tumor stained strongly for glial fibrillary acidic protein (GFAP) which was highlighted in the processes of perivascular pseudorosettes [Figures 2a-c]. It also showed positive staining for S100 and vimentin. Epithelial membrane antigen (EMA) showed apical membrane positivity [Figure 2d]. The tumor was negative for CK and synaptophysin. Ki-67 labeling index was 5%. Based on the histological findings and IHC pattern, a diagnosis of papillary ependymoma was made.

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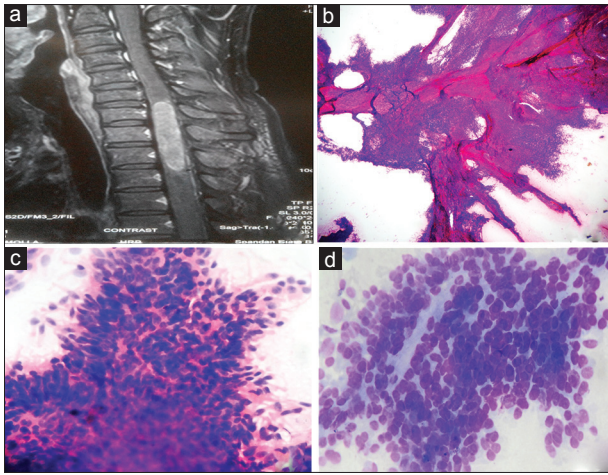
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**Figure 1:** (a) Magnetic resonance imaging shows well-circumscribed intramedullary space-occupying lesion at C6-D2 level. (b) Scanner view shows large papillary structures with fibrovascular core having cells attached to blood vessels in multiple layers (H and E). (c) Low power view shows cohesive clusters of round to oval cells with bland nuclei (H and E). (d) High power view of cells show chromatin of salt and pepper type (MGG)

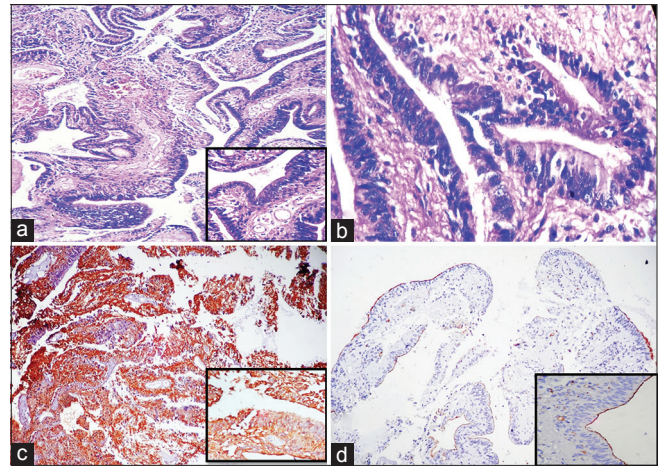
## Discussion

Ependymomas are slowly growing glial neoplasms that occur along the ventricular system or spinal canal. They can be found in any age group and affect males and females almost equally. In addition to the classic and anaplastic types, myxopapillary and tancytic variants of ependymoma have also been described in the spinal cord.<sup>[5,6]</sup> Papillary ependymoma is a rare variant of ependymoma which usually occurs in the brain. There are only a handful of cases of the same reported in the spinal cord. Being a papillary neoplasm, differential diagnoses of choroid plexus papilloma, papillary meningioma, metastatic carcinoma, and papillary tumor of the pineal region come into account.

The 2016 updated WHO classification of CNS tumors places papillary ependymoma in Grade II category. Clinical features and MRI findings of this tumor are nonspecific.

Papillary ependymomas have papillae and finger-like projections lined by single or multiple layers of cuboidal cells with smooth contiguous surface. In our case, histological findings were similar, and the hobnail appearance of choroid plexus papilloma and metastatic carcinoma was absent. Histologically, it is important to distinguish between papillary ependymoma and choroid plexus papilloma because both show papillary structures with fibrovascular core, but papillary ependymoma is Grade II, and choroid plexus papilloma is Grade I and the chance of recurrence is more in the former. Choroid plexus papillomas occur in ventricles, but rarely they may be found in the spinal cord. In choroid plexus papilloma, there is mostly a single layer of cells lining the fibrovascular core and multilayering is absent.

Although the histological features of papillary ependymoma and choroid plexus papilloma are different, sometimes it



**Figure 2:** (a) Low power view shows well-formed papillae and finger-like projections. Inset shows vessels lined by layers of tumor cells (H and E). (b) High power view shows papilla lined by single or multiple layers of tumor cells. True rosettes can also be seen (H and E). (c) Tumor cells show strong positivity for glial fibrillary acid protein. Inset shows glial fibrillary acid protein positivity at high power. (d) Tumor cells show apical membrane positivity for EMA. Inset shows the same at high power

becomes difficult to differentiate between the two. IHC plays an important role here. Papillary ependymoma shows strong positivity for GFAP, S100, EMA, and vimentin.<sup>[3]</sup> It is negative for CAM 5.2, CK7, CK20, and CK903.<sup>[7]</sup> Choroid plexus papillomas are negative for GFAP. They are reactive for vimentin and E-cadherin. They also express CAM 5.2, CK7, transthyretin, and Kir 7.1. Positivity for S100 is variable.<sup>[7]</sup> They are usually negative for neural cell adhesion molecule (NCAM), EMA, and CK20.

Our case showed perivascular pseudorosettes and ependymal canals that are also seen in classic ependymoma. Although the cells had bland nuclei with speckled chromatin like that of classic ependymomas, the Ki-67 LI was higher. It was 5%, while in case of classic ones, it is <1%.

Papillary meningioma is an important differential diagnosis which shows the presence of perivascular pseudopapillary architecture. The cells become discohesive and are arranged around blood vessels with a perivascular nuclear-free zone, but there is the absence of fibrillary processes of cells that are found in ependymomas. Furthermore, the ependymal canals are not present. Papillary ependymoma lacks necrosis and mitosis which may be sometimes seen focally in papillary meningioma.<sup>[8]</sup> Papillary meningiomas show strong and diffuse positivity for EMA. There is also an expression of somatostatin receptor 2A. They are negative for GFAP.

Papillary ependymoma shares many similarities with PTPR, a newly described entity. Both shows tumor cells covering blood vessels in layers. PTPR is negative for GFAP and shows positivity for EMA, E-cadherin, CK, and CAM 5.2.<sup>[2]</sup>

Myxopapillary ependymomas are fairly common in the spinal region. However, it shows characteristic myxoid changes in addition to papillary areas.

**Table 1: Summary of prior published cases of papillary ependymoma of spinal cord along with the present case**

Authors	Case number	Age (years)	Location	Histology	IHC
Dulai <i>et al</i>		8	Intramedullary thoracic spine	Papillary areas, pseudorosettes, and hobnailing	Positive: GFAP, NCAM, MAP2, vimentin, EMA, TTR, E-cadherin, AE1, AE3, CAM 5.2 Negative: Kir 7.1, Synaptophysin Ki67LI: 7%-8%
Mobley <i>et al</i>	Case 1 Case 2	7 17	Intramedullary thoracic spine Intramedullary Cervical spine	Both cases showed papillary and solid areas with monomorphic epithelioid cells arranged around vascular papillae	Positive: EMA, AE1, CAM 5.2, E-cadherin, GFAP, NCAM, TTR Negative: Synaptophysin Ki67LI: 10% Positive: EMA, AE1, CAM 5.2, E-cadherin, NCAM Negative: Synaptophysin, GFAP, TTR Ki67LI: 20%
Our case		40	Intramedullary, C6-D2	Papillary projections lined by single or multiple layers of cells, perivascular pseudorosettes, and ependymal canals	Positive: GFAP, EMA, S100, Vimentin Negative: Synaptophysin, CK Ki67LI: 5%

IHC – Immunohistochemistry; GFAP – Glial fibrillary acid protein; NCAM – Neural cell adhesion molecule; TTR – Transthyretin; EMA – Epithelial membrane antigen

Metastatic papillary carcinomas show atypia, mitosis, and necrosis.

Dulai *et al.* in a case report described one case of spinal papillary ependymoma and Mobley *et al.* in another case report described two cases of the same [Table 1]. The cases were either child or adolescent and the location of the tumor was cervical or thoracic spine. Histologically, all the cases showed papillae and pseudorosettes. Hobnailing was seen in one case. All the cases showed GFAP and EMA positivity and Ki-67 LI >5%.

Squash cytology of ependymomas and its rare variants including myxopapillary ependymoma and tanyctic ependymomas have been described earlier.<sup>[9,10]</sup> However, squash cytology of papillary ependymoma has not been reported in the past. The squash cytology smears from our case of papillary ependymoma were fairly cellular and composed of cohesive clusters of round to oval cells. No dispersed population was seen, and the pulled cotton appearance of other gliomas was absent. Nuclei were bland with salt and pepper chromatin. At places, the tumor formed large papillary projections having fibrovascular cores with cells attached to blood vessels in multiple layers. Grape-like papillary clusters of cells bulging from vessels or honeycomb pattern were not seen. These are features of choroid plexus papilloma. There was no evidence of necrosis or mitosis, and the cells lacked atypia.

Since there are only a few published cases of papillary ependymoma, hardly anything is known about the molecular pathogenesis of this disease.

## Conclusion

We take this opportunity to report a rare case of spinal papillary ependymoma along with its squash findings. A panel of immunostains is useful to confirm the diagnosis. Although papillary ependymoma is a WHO Grade II tumor like the classic ependymoma, its Ki-67 labeling index is higher than that of the classic one.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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