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## Papillary ependymoma

Papillary ependymoma is a rare variant of ependymoma and often gives rise to confusion with choroid plexus papilloma because of topographic, light microscopic and ultrastructural similarities.

## **Case reports**

With the increasing frequency of neuroimaging, incidental intramedullary cavities are diagnosed more frequently. Ng et al. presented a case of asymptomatic incidental intramedullary cervical cavity diagnosed as an idiopathic syringomyelia as initial MRI showed an isolated cystic image without contrasted component. The patient had no subsequent MRI follow-up but eventually showed symptoms 8 years later. By this stage of the disease, the MRI appearance had changed, showing a solid and enhanced component. The patient underwent surgical resection and histopathology concluded a papillary ependymoma (grade 2). This case illustrates how asymptomatic intramedullary cavities may hide an underlying tumoral process and why these cavities should not be considered as idiopathic syringomyelia by default, except after prolonged MRI follow-up <sup>1)</sup>.

In two cases of papillary ependymomas, Brain MRI revealed a large mass in the left lateral ventricle in one case and a 3cm sized mass in the pineal area and the 3rd ventricle in the other. Microscopically, the tumor was characterized by papillary and tubular structures. Immunohistochemically, the tumor cells in both cases expressed cytokeratins(CK22 and CAM 5.2) but did not express glial fibrillary acidic protein(GFAP), vimentin, epithelial membrane antigen, and S100 protein. This is a very unusual immunohistochemical feature for papillary ependymoma. Ultrastructurally, the tumor showed a mosaic pattern of tumor cells with frequent intercellular microrosettes having a few stubby microvilli, a few cilia and zonulae adherentes. The cytoplasmic processes were markedly reduced compared to conventional ependymoma. The cytoplasm did not contain intermediate filaments. Interestingly, the mitochondria showed abnormal features with a pleomorphic shape and abnormal cristae in both cases. These ultrastructural features enabled differentiation between papillary ependymoma and choroid plexus papilloma in addition to the light microscopic findings <sup>2)</sup>.

1)

Ng S, Aghakhani N, Bauchet L. Clinical image of a spinal ependymoma discovered 8 years after initial misdiagnosis as an idiopathic syringomyelia. World Neurosurg. 2020 Oct 3:S1878-8750(20)32181-1. doi: 10.1016/j.wneu.2020.09.162. Epub ahead of print. PMID: 33022432.

Park SH, Park HR, Chi JG. Papillary ependymoma: its differential diagnosis from choroid plexus papilloma. J Korean Med Sci. 1996 Oct;11(5):415-21. PubMed PMID: 8934397; PubMed Central PMCID: PMC3054185.

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