# Papillary glioneuronal tumor presenting with low-pressure hydrocephalus from intraventricular spread: Case Report

Authors: Nicholas A Myers, BS[a] , Daniel F Marker, MD, PhD[b] , Kushal J Shah, MD[c,d] , Troy D Payner, MD[c,d] , Charles G Kulwin, MD[c,d]

## Introduction:

Papillary glioneuronal tumors (PGNTs) are rare, benign neoplasms that are typically solid or solid/cystic, well-circumscribed, parenchymal, solitary, and calcify. While intraventricular location, tumoral hemorrhage, and/or high-pressure obstructive hydrocephalus have been described, presentation with multifocal disease and normal pressure hydrocephalus (NPH)-like symptoms has not. We describe the novel presentation of a glioneuronal tumor distinctly presenting with NPH-like symptoms from intraventricular spread and the associated management.

## Case report:

Mid-50s male patient presented with unsteady gait for over a year, mild nonspecific visual changes and mild short-term memory impairment initially concerning clinically for NPH. Past medical history was significant for hyperlipidemia and solitary kidney. Neurological exam revealed a shuffling gait; the patient denied any other localizing symptoms. Brain MRI revealed a complex hemorrhagic and calcified lesion in the right atrium of the lateral ventricle, in the setting of mild, triventricular enlargement without transependymal flow, with a separate enhancing lesion at the outflow of the cerebral aqueduct. Surrounding the atrial mass were multiple large intraparenchymal cysts extending to the parietal cortex consistent with CSF (Fig. 1). Systemic workup was negative for metastatic disease. The patient underwent a transcortical resection of the intraventricular tumor through the CSF-filled parenchymal cysts. Intraoperatively the tumor appeared exophytic from the atrial choroid plexus. Histologic examination revealed a neuroepithelial neoplasm composed of atypical ganglion cells and a population of smaller neurocytic cells (Fig. 2). The tumor revealed mineralization; numerous hyalinized vessels and pseudopapillary structures. By immunohistochemistry, the gangliocytic population showed staining for neurofilament and synaptophysin, and was negative for nuclear NeuN. A subset of the gangliocytic cells expressed GFAP. The small neurocytic cell population was weakly/moderately positive for synaptophysin and displayed variable positivity for Olig2 and GFAP. The tumor cells were CD34 and TTF-1 negative. High-grade morphologic features were absent, although cytologic atypia was focally increased. The Ki67 proliferation index was less than 1%. Next-generation sequencing studies identified a classic SLC44A1::PRKCA fusion, confirming diagnosis of PGNT. The tumor failed to classify on DNA methylation array analysis using the Heidelberg central nervous system classifier v12.5; the significance of this is unknown. Initially, his NPH-like symptoms resolved, as the resection corridor acted as a ventriculostomy around the aqueductal obstruction; however, this ultimately closed off and symptoms returned. His residual hydrocephalus was subsequently managed via third ventriculostomy with resolution of gait and cognitive symptoms.

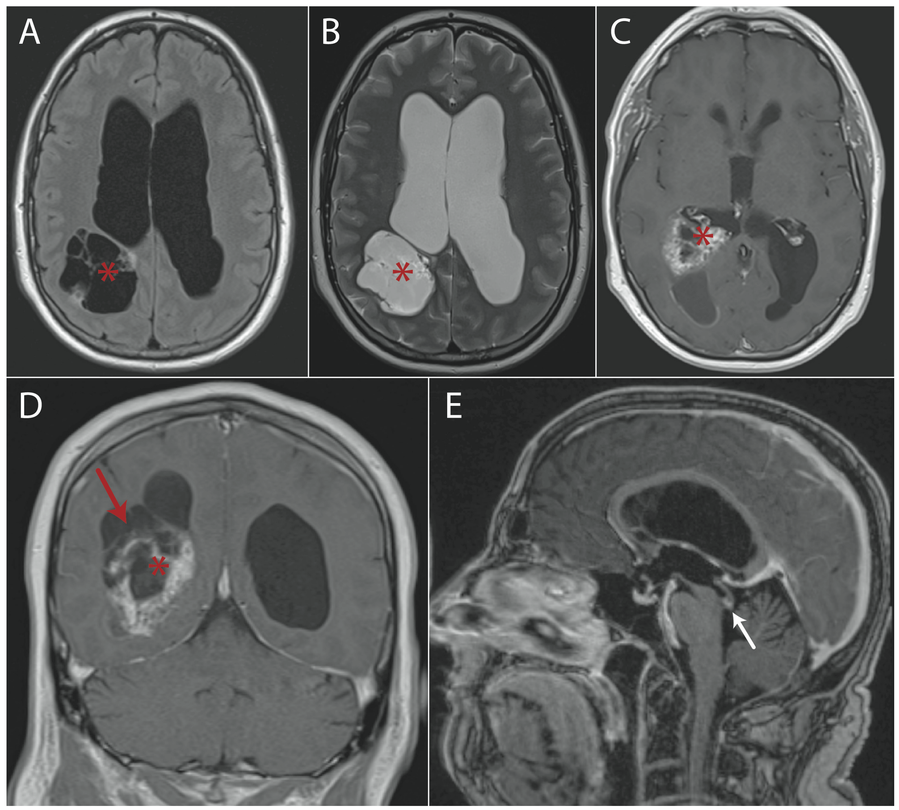


Fig. 1 – Preoperative MRI. Axial FLAIR and T2 (A and B) images showing CSF-consistency parenchymal cysts extending from the tumor to the subcortical region (asterisk). Contrasted T1 images (C axial and D coronal) show the atrial tumor (asterisk) and the surgical corridor through the parenchymal cysts (red arrow). Figure E shows the satellite lesion (white arrow) in the distal aqueduct causing the clinical hydrocephalic presentation. MRI: magnetic resonance imaging

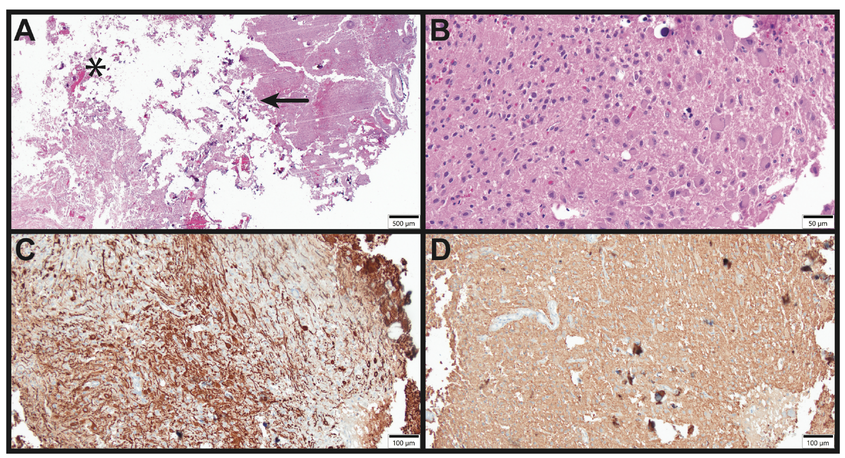


Fig. 2 – Histologic features. (A) Low-power view of hematoxylin and eosin (H&E) stained section. The tumor was heavily fragmented and mineralized. Large areas were hypocellular and showed numerous hyalinized vessels (asterisk) while other areas were more cellular (arrow). The adjacent brain parenchyma showed reactive piloid gliosis (not shown). (B) Higher-power view. The tumor was composed of a gangliocytic component and a smaller neurocytic component. Mitotic activity was not prominent and there was no endothelial proliferation or necrosis. Cytologic atypia was focally increased (not shown). (C and D) Immunohistochemical staining for neurofilament and synaptophysin, respectively. The gangliocytic cells showed variable somal positivity for neurofilament. Synaptophysin highlights a neuropil background and was weakly to moderately positive in both the gangliocytic and smaller neurocytic components.

## Extraordinary Care for an Extra-Ordinary Brain Tumor

## Conclusion:

The unusual tumor presented with multifocal disease from presumed intraventricular spread, which has never been described, with remote disease obstructing the cerebral aqueduct. The ventricular system decompressed through the tumor, creating intraparenchymal CSF cavities and thus presenting as a lower-pressure, chronic hydrocephalus while providing surgical access for resection without deficit. This case describes the unique presentation of a multifocal intraventricular tumor and its subsequent management.

## Affiliations:

a) College of Osteopathic Medicine, Marian University, 3200 Cold Spring Rd, Indianapolis, IN, 46222, USA

b) Department of Pathology, University of Pittsburgh Medical Center, Pittsburgh, PA, USA

c) Goodman Campbell Brain and Spine, Carmel, IN, USA

d) Ascension St. Vincent Hospital, Indianapolis, IN, USA

Images from the published report in Radiology Case Reports
Volume 18, Issue 10, October 2023, Pages 3632-3635

GOODMAN CAMPBELL
BRAIN AND SPINE