

# Papillary Glioneuronal Tumor with an Excessive Angiomatous Component in an Elderly Man

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Papillary glioneuronal tumor (PGNT) is a rare tumor categorized as a distinct mixed glioneuronal neoplasm in the World Health Organization (WHO) 2007 classification.<sup>[1]</sup> Here, we report a case of PGNT accompanied by excessive areas with angioma-like features in an elderly man. This report contributes to the diverse spectrum of this rare tumor entity.

A 64-year-old man experienced sudden dizziness for 20 days. He was admitted to Xuanwu Hospital, Capital Medical University, Beijing, China, with the diagnosis of a cerebral tumor. On admission, his hematological tests and physiological functions were in the normal range. A computed tomography (CT) scan revealed a large mass lesion in the left frontal and parietal lobe near the lateral ventricles. Slight calcification was observed in the lesion. Magnetic resonance imaging revealed a cystic tumor with a mural nodule. Perilesional edema was absent, and no mass effect or mid-line shift was noted. The mural nodule exhibited low-signal intensity on T1-weighted images [WI, Figure 1a] and high-signal intensity on T2-WI [Figure 1b] and fluid-attenuated inversion recovery images. The tumor was enhanced with gadolinium, but the cyst wall was not. The patient subsequently underwent surgery with clinical suspicion of choroid plexus papilloma or astrocytoma. During surgery, a cyst containing clear, yellowish fluid and a mural nodule protruding into the cyst were detected. Postoperative CT imaging confirmed radical removal. No radiotherapy was offered to the patient, and no signs of recurrence were noted at the 4-year follow-up.

Macroscopically, the sample was gray-white and soft. Histological examination of the tumor revealed the following different architectural patterns: pseudopapillary and angiomatous areas [Figure 1c and 1d]. In pseudopapillary areas, hyalinized and thickened vessels were covered by single or multiple layers of uniform cuboidal cells [Figure 1d]. Diffuse vascular hyalinization was present throughout the tumor. Interpapillary areas were occasionally populated by sheets of loosely arranged cells with medium-sized,

round nuclei in a background of loose, fibrillar matrix. In focal areas, smaller cells with clear cytoplasm and perinuclear halos reminiscent of oligodendroglia were observed. Angiomatous areas consisting of various sized vessels [Figure 1c] were a unique finding in the present patient. These vessels were lined by a single layer of flattened endothelium lacking smooth muscle in the wall, and there was no evidence of endothelial hyperplasia. Areas of hemosiderin and calcification were noted. These angiomatous areas were scattered among the pseudopapillary areas and occupied approximately two-thirds of the total tumor tissue.

Immunohistochemical examination revealed both neuronal and glial components. The cuboidal cells lining the pseudopapillae were strongly positive for glial fibrillary acidic protein [GFAP, Figure 1e] and vimentin but were negative for neuron-specific enolase (NSE) and neuronal nuclear antigen [NeuN, Figure 1f], suggesting glial differentiation. Conversely, medium-sized cells in the solid areas did not exhibit GFAP immunoreactivity but were positive for NSE and NeuN [Figure 1f], suggesting neuronal components. The Ki-67 labeling index was very low (approximately 2%), and all tumor cells were negative for p53, isocitrate dehydrogenase (IDH1-R132H), and cytokeratin 7. In pseudopapillary and angioma-like areas, endothelial cells were positive for CD34. Desmin and actin were negative around angioma-like areas. All antibodies were obtained from Beijing Zhongshan Golden Bridge Biotechnology Co., Ltd., (Beijing, China). Based on these pathological findings, the patient was diagnosed with PGNT (WHO Grade I).

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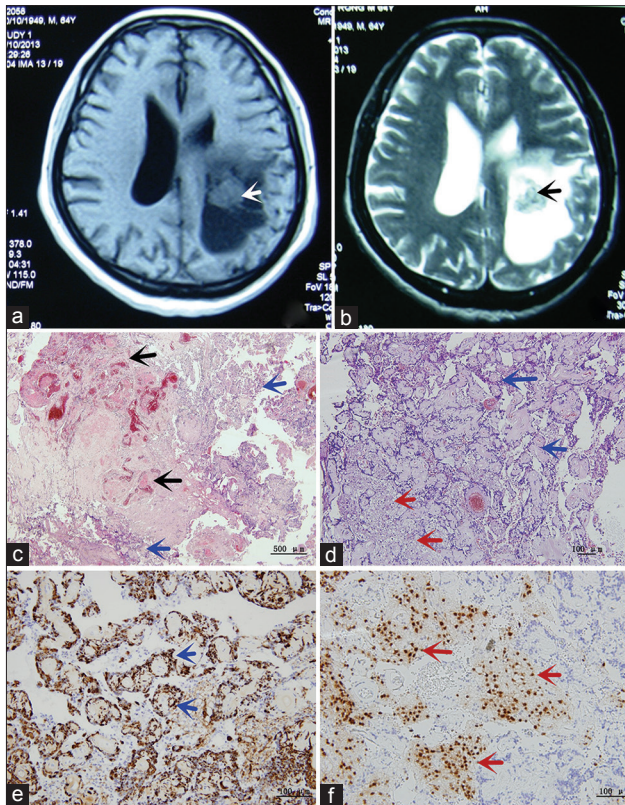
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**Figure 1:** Representative images of the patient. (a and b) Magnetic resonance imaging of the cystic tumor. ([a] T1-weighted image, [b] T2-weighted image). (c and d) Histological features of the tumor revealing the double-architectural patterns, cavernous hemangioma areas (c) black arrow heads, and pseudopapillary areas (c and d) blue arrow heads. In the pseudopapillary areas, aggregates of larger cells with abundant cytoplasm and distinct nucleoli (d) red arrow heads were noted (Hematoxylin and Eosin, [c]  $\times 100$ , [d]  $\times 200$ ). (e) Cuboidal cells covering the hyalinized vessels were positive for glial fibrillary acidic protein (immunohistochemical,  $\times 200$ ). (f) The solid area (red arrow heads) was positive for neuronal nuclear antigen (immunohistochemical,  $\times 300$ ).

PGNTs are rare tumors that predominantly occur in young adults and exhibit an equal gender distribution.<sup>[2]</sup> In this report, the patient was a 64-year-old man. PGNT has also been reported in a 75-year-old woman.<sup>[3]</sup> Both reports suggest that PGNT can clinically manifest in the elderly.

Histologically, PGNT exhibits stereotypic morphology with evidence of both glial and neuronal cell differentiation.<sup>[4]</sup> Glial cells typically line vascular structures, forming pseudopapillae. The neuronal component comprises sheets of cells distributed in the interpapillary zone.<sup>[4]</sup> Classical histomorphological features comprising prominent gliovascular structures and interpapillary sheets of cells exhibiting neuronal differentiation based on morphology and immunohistochemistry were the hallmark findings in the patient.

Diagnostic histological features in this patient included a pseudopapillary lesion with central hyalinized vessels and intervening regions of loosely dispersed sheets of cells with neuronal marker expression. Differential diagnoses for this patient included choroid plexus papilloma, angiomatous

meningioma, and ganglioglioma. Choroid plexus papilloma was excluded based on immunostaining for neuronal markers and a lack of focal cytokeratin expression. In contrast to oligodendrogliomas and astrocytomas, the patient's tumor exhibited no immunoreactivity for mutant IDH1 protein, indicating that IDH1 mutations did not contribute to the pathogenesis of this tumor.

The low proliferation index reported in PGNT is paralleled by a favorable clinical outcome. The presence of pseudopapillae, degenerative changes and a low Ki67 labeling index confirm that PGNTs are slow-growing, low-grade neoplasms. Complete/near complete resection is the treatment of choice for PGNTs.

The patient in this report is unique because he presented with combined vascular proliferation areas mimicking angiomatous areas. Although the presence of angiomatous areas in PGNT has been previously reported,<sup>[1,5]</sup> these areas cover only a small portion of the tumor. However, in the patient, these "angiomatous"-like areas occupied approximately two-thirds of the total tumor tissue. Thus, we initially almost ignored other cellular components and could have misdiagnosed this lesion as cavernous hemangioma. The formation of excessive "angiomatous"-like areas is difficult to interpret. This condition might secondarily result from long-term tumor growth with significant degenerative changes. We hypothesize that long-term tumor growth contributed to angiomatous changes and finally generated cystic changes in the tumor.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name 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.

### REFERENCES

1. Agarwal S, Sharma MC, Singh G, Suri V, Sarkar C, Garg A, *et al*. Papillary glioneuronal tumor – A rare entity: Report of four cases and brief review of literature. *Childs Nerv Syst* 2012;28:1897-904. doi: 10.1007/s00381-012-1860-3.
2. Husain N, Husain M. Endoscopic diagnosis of a pineal papillary glioneuronal tumor with extensive ventricular involvement: Case report with review of literature. *Neurol India* 2009;57:792-5. doi: 10.4103/0028-3886.59481.
3. Tsukayama C, Arakawa Y. A papillary glioneuronal tumor arising in an elderly woman: A case report. *Brain Tumor Pathol* 2002;19:35-9. doi: 10.1007/BF02482454.
4. Celli P, Caroli E, Giangaspero F, Ferrante L. Papillary glioneuronal tumor. Case report and literature review. *J Neurooncol* 2006;80:185-9. doi: 10.1007/s11060-006-9170-9.
5. Radotra BD, Kumar Y, Bhatia A, Mohindra S. Papillary glioneuronal tumor: A new entity awaiting inclusion in WHO classification. *Diagn Pathol* 2007;2:6. doi: 10.1186/1746-1596-2-6.