

Case Report

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Cerebellar Ganglioglioma

The location of ganglioglioma (GG) within the infratentorial compartment is unusual. The authors report a rare case of GG in the cerebellar hemisphere. A 12-year-old boy suffered from headache and gait disturbance. Neuroimaging studies demonstrated a large enhancing cerebellar mass with cystic components compressing the fourth ventricle. After complete resection of the tumor, the patient became symptom free. Histological examination on the tumor disclosed glial cells and dysplastic ganglion cells. Although it is a rare tumor, in the appropriate clinical setting, a GG should be considered in the presence of a cerebellar mass with both solid and cystic components on magnetic resonance images in children.

KEY WORDS : Ganglioglioma · Cerebellum · Brain neoplasm · Children.

INTRODUCTION

Gangliogliomas (GGs), histologically defined by atypical ganglion cells and neoplastic glial cells, are found predominantly after a seizure in child or young adult⁷⁾. The major location for GG is the temporal lobe. Uncommonly, these lesions do occur in the cerebellum, basal ganglia, pineal gland, hypothalamus, optic nerve, spinal cord, brain stem, and pituitary gland^{2,8,9)}. In the literature, only a few cases of GG arising from the cerebellum have been reported^{4,15)} with being even rarer in Korean⁶⁾. The authors present herein a rare case of GG in the cerebellar hemisphere which radiologically diagnosed as a pilocystic astrocytoma (PA).

CASE REPORT

A 12-year-old boy complained of occipital headache for 4 months. There was no familial history about medical illness. On neurological examination, there were signs of cerebellar dysfunction including gait disturbance and dysmetria. Brain magnetic resonance (MR) images revealed a 5.2 cm mass in the right cerebellum which displaced brain stem (Fig. 1A, B). The mass was associated with obstructive hydrocephalus. The cystic portion of tumor showed the cerebrospinal fluid-like signal intensity on MR images. The tumor showed heterogenous ring enhancement with gadolinium administration (Fig. 1C). There was no calcification within the tumor on the computed tomography (CT) scan. Preoperative tentative diagnosis of the lesion was a PA.

We performed the operation via a right suboccipital craniotomy. After opening of tensed dura, a cyst was noted and 20 ml of xanthochromic fluid was then aspirated. The solid tumor part was yellow-grayish, slightly mucoid, rather readily bled, and was well demarcated from adjacent folia (Fig. 2). The tumor was totally removed macroscopically. Histologically, the tumor was composed of ganglion cells and astrocytic cells, compatible with the GG (Fig. 3A, B). Postoperatively, there was an uneventful recovery without any neurological deficits. No further treatment was given. There has been no signs of tumor recurrence on repeated MRI (Fig. 1D). At last follow-up, 20 months after surgery, this boy had no problems in daily school life.

DISCUSSION

Ganglioglioma is the most commonly encountered glial-neuronal neoplasm of the central nervous system, with its incidence of 0.4% to 4.3%¹⁴⁾. This tumor is usually seen in children and young adults and there is no gender preponderance³⁾. GGs may arise throughout the neural axis, although the majority of the lesion found in the temporal lobe. As a possible reason for this, it has been suggested that the subgranular zone of the dentate gyrus has a potential to produce

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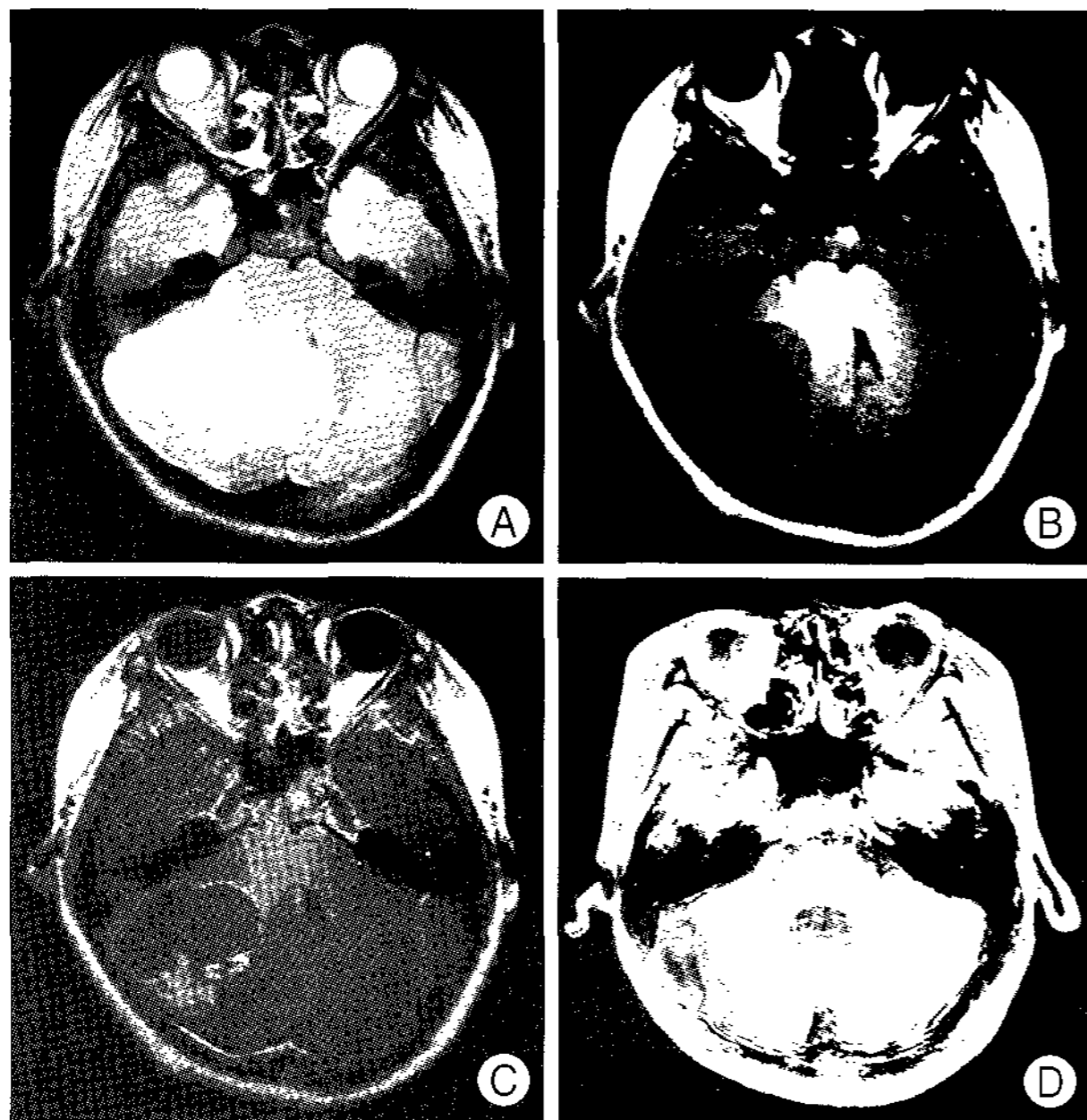


Fig. 1. Axial magnetic resonance images before and after surgery in 12-year-old boy having a ganglioglioma in the cerebellum. This tumor appears hyperintense on T2-weighted image (A) and hypointense on T1-weighted sequence (B). The cyst wall and intracystic solid nodule are enhanced uniformly and strongly (C). One year postsurgical scan shows no evidence of lesion recurrence (D).

the granular neurons postnatally. Those neurons have a greater susceptibility to a neuronal-glial neoplastic transformation resulting in a GG^{13,16}.

The supratentorial cortical lesions often present with long history of seizures, whereas posterior fossa GGs have presented with focal neurological deficits, cranial nerve palsy, hydrocephalus, increased intracranial pressure, speech or gait changes, and myoclonus^{3,5,7}. In addition, the shorter interval from symptom onset to the diagnosis was conspicuous as compared with supratentorial GGs because of the anatomically narrow space of the infratentorial compartment^{1,4}. Interestingly, an exceptional case with infratentorial GG who presented with cerebellar seizure was also reported¹⁰. Nevertheless, there are no specific clinical findings to indicate cerebellar GG and to discriminate that from other cerebellar lesions that are more common in pediatric patients.

Radiographically, there is no single "diagnostic" appearance for supratentorial and cerebellar GGs, because cyst with an enhancing mural nodule is classic, but not specific for GGs. The tumor is often isodense or hypodense in the precontrast CT, with calcification present in 6% to 30%^{3,12}. In our case, there was no definite calcification. On MR imaging studies, GGs are usually hyperintense on T2-weighted images and isointense to slightly hypointense on T1-weighted images¹⁴. The variable signal in cystic portion depends on whether the contents are proteinaceous, hemorrhagic, or contain cerebrospinal fluid. Gadolinium enhancement of the solid com-

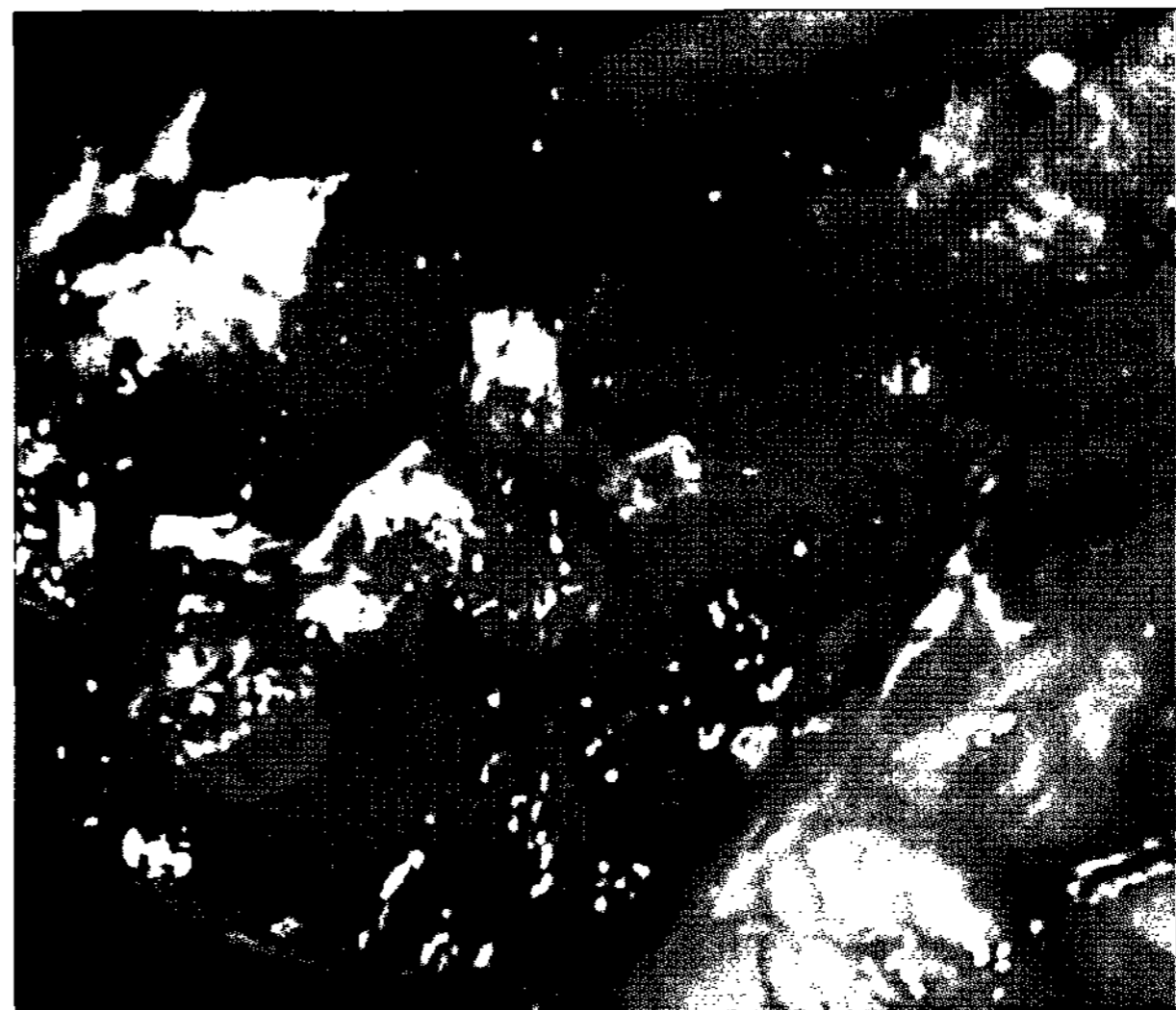


Fig. 2. Operative photograph of cerebellar ganglioglioma. Sequential dissection of the tumor from the surrounding cerebellar cortex is being performed. The solid component is yellowish, slightly viscous, and well-defined.

ponents of the GGs is observed in about half of the cases, although the pattern varies from intensely homogenous to heterogenous^{11,16}. Although there is considerable variability in the radiographic appearances of the cerebellar GGs, almost all tumors appear as a discrete, solid/cystic lesion with mild mass effect and little or no surrounding edema^{1,15}. The differential diagnosis of cerebellar GGs in children or young adults includes cerebellar astrocytomas, ependymomas, desmoplastic medulloblastomas, and hemangioblastomas. Each of these of tumors may demonstrate cyst formation as well as a solid portion. Among them, the PA, the most common primary cerebellar tumor in children, can be indistinguishable from cerebellar GG radiologically. In this case, neuroimaging studies showed characteristic "cyst with mural nodule" appearance of PA in posterior fossa and somewhat heterogenous enhancement including wall of cyst, making us strongly suspect a PA preoperatively. Such features are similar to those of hemangioblastoma and metastasis, but these differential diagnostic possibilities could easily be excluded by the patient's age and its association with systemic manifestation^{2,3}. An uniform hyperdensity on CT scans, midline location, and filling and expanding of the 4th ventricle are the hallmarks of medulloblastoma. On the contrary, cerebellar GGs arise from hemisphere and compress the 4th ventricle, as demonstrated in this case. Ependymoma extends out the 4th ventricle foramina, and are usually heterogenous with calcification and hemorrhage, but those are not typical for GGs on CT and MR images⁸. Additionally, other rare disease processes known to involve the posterior fossa in child are gangliocytoma, oligodendroglioma, neurocysticercosis, dysembryoplastic neuroepithelial tumor, atypical teratoid-rhabdoid tumors, pleomorphic xanthoastrocytoma, schwa-

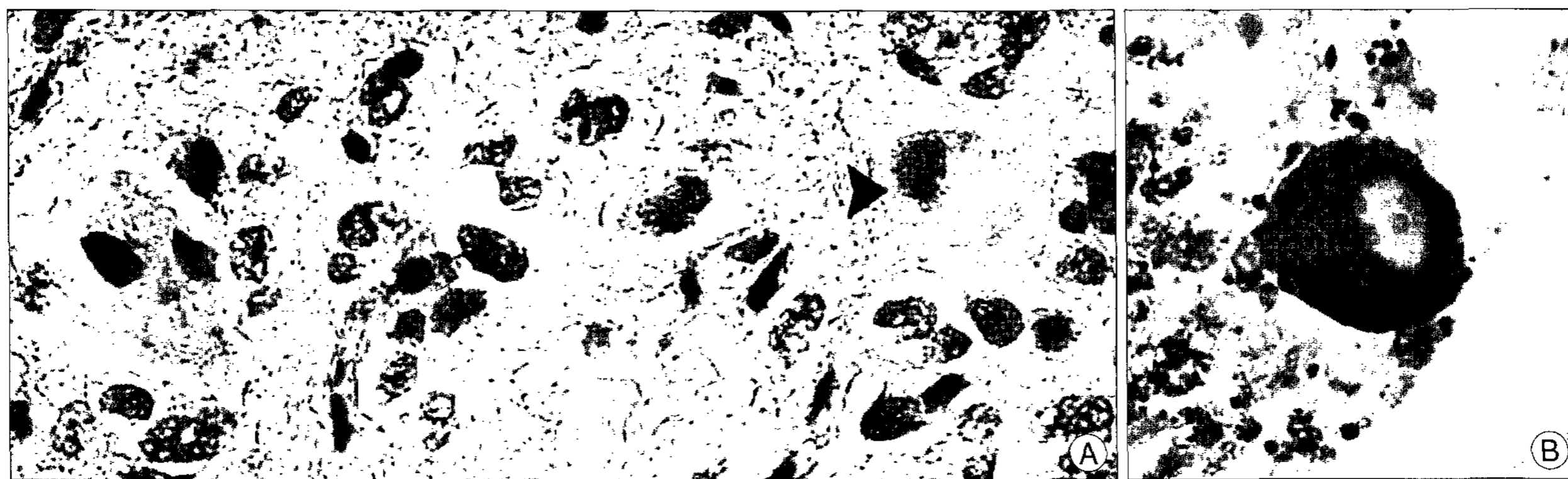


Fig. 3. Photomicrographs of the tumor pathology. A : The H & E staining reveals diffuse proliferation of astrocytes together with ganglion-type neurons (arrowhead) (original magnification, $\times 200$). B : Immunohistochemistry for synaptophysin highlights the dysplastic ganglion cells (synaptophysin, $\times 400$).

nnoma, hemangioma, and meningioma. In this setting, imaging characteristics such as disease dissemination, extension into the cerebellopontine angle, tumor demarcation, pattern and degree of contrast enhancement, tumor necrosis, meningeal involvement, multiplicity, invasion into brain stem, and cortex expansion are also clues in the radiological differentiation of cerebellar GGs from other entities^{2,4,12}.

Accordingly, as our presumption on diagnosis before surgery, an enhancing intra-axial tumor with cystic change in a "middle-age" child who present with cerebellar signs and/or symptoms of hydrocephalus, the cerebellar mass is more likely to be a PA than anything else. However, even intraoperatively, cerebellar GG can be difficult to differentiate from the cerebellar astrocytoma, because there is a quite similarity between both tumors during operation, such as color, consistency, vascularity, and demarcation^{6,15}. Consequently, correct diagnosis of cerebellar GG requires elaborate histological examinations. The identification of atypical-appearing neuronal or ganglion cells in the tumor is the first step. Commonly, the neuronal cell component is unevenly distributed throughout the neoplasm and, therefore, appropriate sampling of the tumor is important for an accurate diagnosis¹¹. The highlighting the neuronal populations using Nissl's method and synaptophysin immunohistochemistry provides a valuable aid for the diagnosis of small and fragmented tumor samples¹³. The second important point on the histopathologic confirmation of GG is that the glial component of the tumor may demonstrate variable degrees of hypercellularity and nuclear pleomorphism¹⁶.

Surgery is the treatment of choice for this histologically benign neoplasm. The tumor is often well circumscribed and, therefore, complete resection would be feasible, which results in greater than 90% of 5-year survival^{5,7}. Unlike this case, found to be slightly vascular at surgery, cerebellar GGs are usually avascular and be visibly calcified at times. The location of the lesion dictates the surgical approach. Because the majority of these tumors are located in the temporal lobe, a

temporal or pterional craniotomy is used to approach the lesion. Given the history of long-standing seizure associated with GGs, intraoperative electrocorticography is an important surgical adjunct and often guides the extent of resection beyond the radiographic or visuotactile boundaries of the tumor¹⁰. If the lesion is served in the suboccipital approach, similarly for other lesions in posterior fossa, local and regional anatomy must be appreciated. Whereas aggressive resection is the goal, the cerebellar vessels, dentate nucleus, cerebellar peduncle, fourth ventricle, and cranial nerve and its nucleus should be protected during the surgery of infratentorial GGs¹¹. Cerebellar GGs are also indolent, but they retain the potential to grow. With regard to recurrent or progressive lesions and tumors with anaplastic features, radiation therapy has been recommended, but it remains uncertain whether or not this improves outcomes¹⁴.

CONCLUSION

The authors report a rare case of GG that occupied the cerebellar hemisphere in a pediatric patient. In spite of its rarity, GG should be included in the differential diagnosis of a cystic infratentorial tumor in the children. With complete tumor resection during surgery, cerebellar GG could be cured and no additional therapy is required.

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