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CASE REPORT OPEN ACCESS

CEREBRAL GANGLIOGLIOMA: A CASE REPORT

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ABSTRACT

Ganglioglioma (GG) is a benign and rare tumor of the central nervous system, typically located supratentorially in the temporal lobe. Subtentorial localization, particularly in the cerebellum, is uncommon.It represents 0.4% to 7.6% of all central nervous system tumors, 1.3% of brain tumors, and only 1% of intramedullary tumors. Clinically, supratentorial GG causes epilepsy, while subtentorial GG leads to focal neurological deficits due to local mass effect and signs of increased intracranial pressure. A benign brain tumor, called a ganglioglioma (GG), can sometimes be found in the cerebrum. We describe an unusual pediatric two cases of supra and subtentorially GG with a spinal cord localisation in this article. Our patient suffered from headache and epilepsy. Magnetic resonance imaging of the skull revealed a giant cyst with a mural nodule in the right cerebellar hemisphere that flattened the fourth ventricle. Pilocytic astrocytoma was the provisional diagnosis on the basis of clinical and imaging details. After biopsy and immunohistochemistry of the extramedullary intra dural process revealed the presence of a GG. Although GG is an uncommon tumor, it should be included in the differential diagnosis of a cerebellar mass with cystic and solid components in children.

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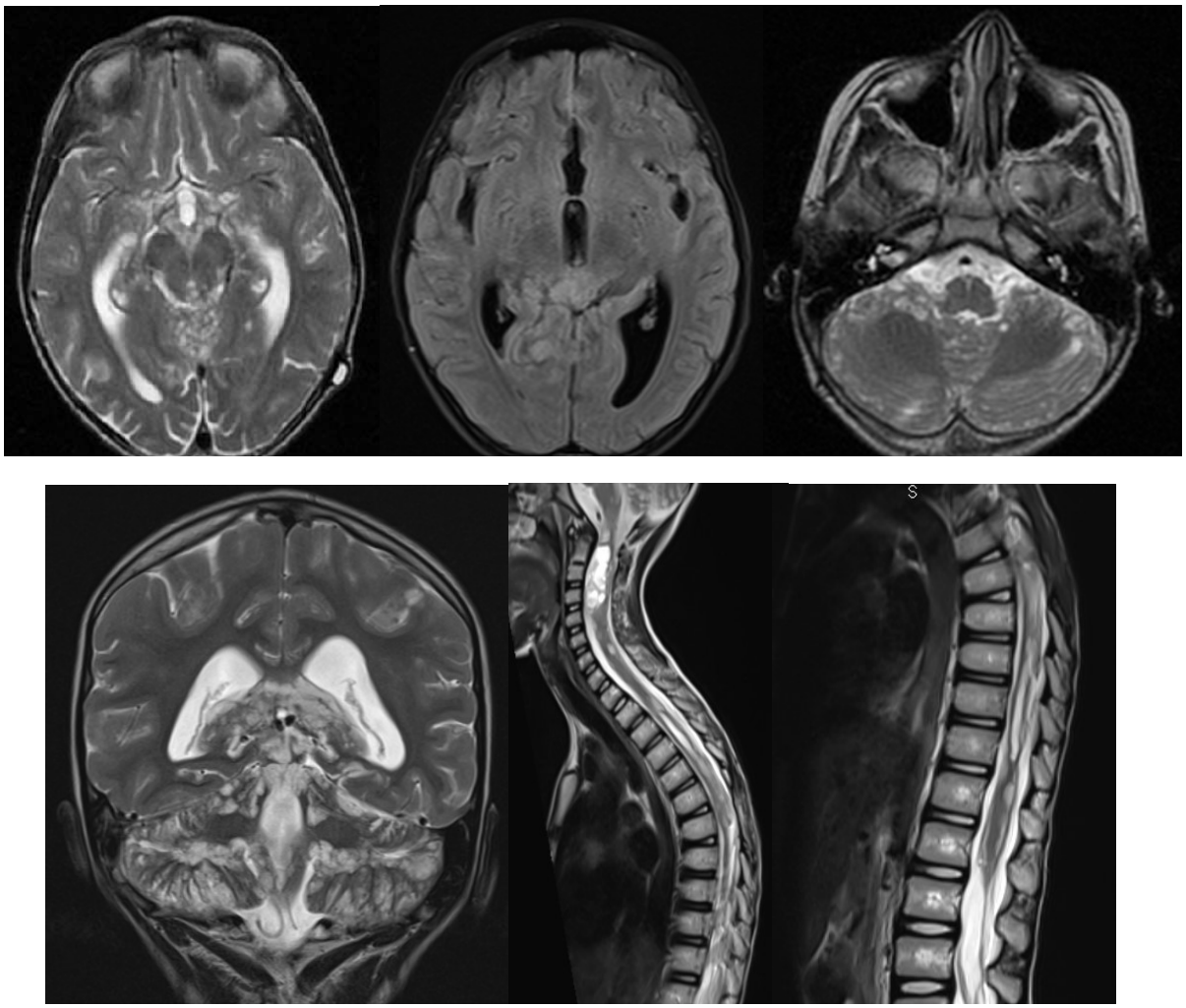
INTRODUCTION

Ganglioglioma (GG) is a benign and rare tumor of the central nervous system, typically located supratentorially in the temporal lobe. Subtentorial localization, particularly in the cerebellum, is uncommon. Histopathologically, GG consists of both glial and ganglion cells. It is mainly observed in children. Seizures and epilepsy are the most common symptoms (1). Since their description and histological study in 1930, several studies have been dedicated to them. Despite their slow clinical progression, these tumors are relatively benign, but several series have shown that they can be aggressive or malignant due to their glial components (2). In this article, we describe an unusual case of GG that radiologically presented as metastatic astrocytoma with diffuse leptomeningeal glioneuronal tumor. Complete resection is the preferred treatment with a 5-year survival rate of over 90%.

CASE REPORT

Over a 5-year period, from 2015 to 2020, a 12-year-old child with no previous medical history was received at our hospital structure, who presented to the emergency department at the age of 9 with complicated symptoms of intracranial hypertension, including seizures, an ataxic gait and coordination disorders. Magnetic resonance imaging (MRI) of the brain and spinal cord revealed multiple supratentorial and subtentorial lesions with triventricular hydrocephalus, as well as diffuse and disseminated spinal cord involvement. Biopsy and immunohistochemistry of the extramedullary intra dural process confirmed the diagnosis in favor of a diffuse leptomeningeal glial tumor with secondary staged medullary localization (Figures). A chemotherapy protocol has been considered, along with the possibility of regular follow-up.



Figures Brain MRI (axial and coronal) and sagittal spinal MRI in T2-weighted sequence ; showing a supratentorial and infratentorial cerebral ganglioglioma with a medullary process resembling an astrocytoma. After spinal biopsy, a GG was demonstrated through histopathological analysis with leptomeningeal dissemination

DISCUSSION

Epidemiologically, the term "ganglioglioma" was first used by Loretz in 1870 [4], and Perkins popularized it in 1926 (3). With slow growth and a peak incidence between 10 and 20 years of age, GG consists of astrocytic glial cells and neuro-ganglion cells. It represents 0.4% to 7.6% of all central nervous system tumors, 1.3% of brain tumors, and only 1% of intramedullary tumors (4). It can develop anywhere in the central nervous system, with the temporal lobe, cerebellum, parieto-occipital region, frontal lobe, and spinal cord being the most common locations. It can also be found in the brainstem, thalamus, hypothalamus, V3, V4, trigone, pineal region, and optic nerve. However, subtentorial localization, especially in the cerebellum, is extremely rare. (5) Clinically, supratentorial GG causes epilepsy, while subtentorial GG leads to focal neurological deficits due to local mass effect and signs of increased ICP, such as headaches, nausea, and vomiting. However, there are no specific signs that suggest a cerebellar GG and differentiate it from other cerebellar neoplasms that occur in children (6). Medical imaging reveals, on CT scan, an isodense, hypodense, or cystic lesion with a enhancing mural nodule after contrast injection, without perilesional edema. Calcifications within the mass have been reported in 6% to 30% of cases. Several series suggest possible confusion with arachnoid cysts and porencephalic cavities (3). The positive diagnosis of gangliogliomas relies on MRI, which is not specific in the absence of spectroscopy. MRI shows a well-circumscribed lesion with hypo-intensity on T1 and hyperintensity on T2, with little or no mass effect. However, low-grade gliomas and juvenile pilocytic astrocytomas can pose a differential diagnostic problem. Furthermore, the presence of calcifications in one-third of cases may raise the possibility of oligodendroglioma. Spectroscopy can be significantly helpful in establishing the diagnosis by differentiating gliomas based on the choline/creatine ratio, which is lower in gangliogliomas compared to gliomas, while the N-acetyl aspartate/creatine ratio is higher. Diffusion-weighted imaging shows elevated apparent diffusion coefficient (ADC), higher than that of low-grade gliomas, indicating low tumor cellularity (7). Perfusion imaging shows elevated relative cerebral blood volume (rCBV) without changes in vascular permeability.

In terms of treatment, complete resection is the preferred treatment with a 5-year survival rate of over 90%. However, complete resection of cerebellar GG or GG in the brainstem is challenging due to adjacent structures. In these cases, partial resection is recommended to minimize neurological sequelae, and radiotherapy is suggested to reduce the risk of recurrence and malignant transformation (7, 8).

CONCLUSION

Cerebellar GG is extremely rare. The symptoms are nonspecific and may include convulsive seizures and signs of increased ICP. Radiologically, the manifestations of GG and pilocytic astrocytoma can overlap. Radical neurosurgery, the optimal treatment of choice, improves prognosis but may not be feasible in all patients. The use of adjuvant chemoradiotherapy remains controversial.

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