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Abstract

Gangliogliomas are rare neoplasms, histologically defined by the presence of both atypical ganglion cells and neoplastic glial cells. Usually located in the supratentorial space, they occur frequently during childhood and adolescence. Clinical and radiological features of GGs are not specific and are very similar to those of cystic cerebellar astrocytoma. Surgery is the treatment of choice of GGs and the role of radiotherapy and chemotherapy remains unclear. We report the case of an 8-year-old child who presented with inoperable cerebellar GG and treated exclusively by chemotherapy followed by radiotherapy.

Introduction

Gangliogliomas (GGs) are uncommon slow-growing tumors of the central nervous system histologically defined by the presence of both atypical ganglion cells and neoplastic glial cells (1). These tumors are usually located in the supratentorial space, primarily the temporal lobe, but they can arise in virtually any part of the brain (2-6). GGs occur frequently during childhood and adolescence (7,8). Clinical and radiological features of GGs are not specific and are very similar to those of cystic cerebellar astrocytomas (3,9). Surgery is the treatment of choice of GGs and the role of radiotherapy and chemotherapy remains unclear. (4,5,8) In the literature, only a few cases of GGs arising from the cerebellum have been reported. Because of their localisation complete resection of infratentorial GG is often not possible. In this work we report an inoperable cerebellar GG in an 8-year-old child treated exclusively by chemotherapy followed by radiotherapy.

Case Report

An 8-year-old girl presented at our institution with headache and right hemiparesis of 4-months duration. On admission, neurological examination revealed right hemiparesis associated with gait disturbance. Computed tomography (CT) and Magnetic resonance imaging (MR) scans showed a cystic and solid lesion in the right lateral cerebellar hemisphere extending to the bulbar region and the bulbo-medullary junction (Figures 1,2,3). A stereotactic biopsy was performed, which confirmed the diagnosis of benign ganglioglioma (WHO grade I). Because of the localisation of the tumor, surgery was not possible, so the patient received a first-line chemotherapy with carboplatin and vincristine. After six cycles, a new MRI shows a stability of the lesion whereas the patient develops a left hemiparesis. Radiotherapy was given to the patient who received a dose of 45 Gy in 25 fractions delivered over 5 weeks. The entire regimen of radiation was well tolerated by the patient.

Discussion

GG is a relatively uncommon tumour of the central nervous system that was first described in 1926 as a distinct clinicopathologic entity containing both mature neuronal and glial neoplastic elements (10). They represent between 0.4% and 7.6% of all paediatric central nervous system (CNS) neoplasms and 1.3% of those in adults (4-6, 11). This tumor is usually seen in children and young adults during the first three decades of life, but they can occur at any age; extremes of a 3-day-old neonate and 80-year-old adults have been documented (7-8, 12-14). Ganglioglioma are more commonly located in the supratentorial area, mainly in the temporal lobe, but it can occur anywhere in the CNS. Less frequent locations within the cerebral hemisphere are (in order of decreasing frequency): the frontal, parietal, and occipital lobes (3-6, 8,15,16). Gangliogliomas occur rarely in the cerebellum with 34 cases reported previously (17-23). Clinical symptoms depend of the location of the tumors: for supratentorial GGs, seizure occurs with an incidence rate of 72-100% (3,4, 24,25). Posterior fossa GGs are more presented with focal neurological deficits, cranial nerve palsy, hydrocephalus, increased intracranial pressure, speech or gait disturbance, and myoclonus (1, 17, 18, 21, 26, 27). There are no specific radiological findings that could be discriminate GGs from other cerebellar lesions that are more common in pediatric patients such as cerebellar astrocytomas, ependymomas,

desmoplastic medulloblastomas, and hemangioblastomas. These tumors may also demonstrate cyst formation as well as a solid portion, CT-scan shows an isodense or hypodense tumor, with calcification present in 6% to 30% (26). Mass effect and oedema are minimal (3,28,29). MR findings also are not characteristic. GGs appears as a well-defined lesion with variable mass effect. They usually have low signal on T1-weighted images but some are isointense. On T2-weighted images, solid lesions show increased signal. The variable signal in cystic components depends on whether the contents are proteinaceous, haemorrhagic, or contain cerebrospinal fluid (3,28,29). Correct diagnosis of cerebellar GGs requires adequate sampling of the tumor and elaborate histological examinations. This is determined by identification of atypical-appearing neuronal or ganglion cells in the tumor with the glial component of the tumor that may demonstrate variable degrees of hyper cellularity and nuclear pleomorphism (4,30,31).Synaptophysin, neurofilament protein glial and fibrillary acidic protein may also help to the diagnosis (32). GGs can be categorized further as either low-grade or high-grade tumors. Low-grade GGs are slow-growing tumors that generally have a favourable prognosis unless occurring in a non-resectable location. If the tumor has anaplastic features or necrosis of the glial components or an elevated MIB-1 labelling index ≥10%, it is considered a high-grade glioma and tends to behave in a more aggressive fashion (33). Gross total resection is the recommended treatment of choice for GGs and complete tumor resection is associated with good prognosis (4,5). Long-term survival is achieved if gross total resection is possible, ranging between 7 and 17 years (24,26,34,35). The benefit of adjuvant radiotherapy has not been proven. In fact, many physicians are hesitant to administer radiotherapy because of the potential morbidity in long-term survivors (34,35). Furthermore, it have been suggested that postoperative radiation may predispose GGs to malignant degeneration (36,37). As GGs are quite uncommon, no prospective studies have been performed or can be expected in the near future to assess the role of radiotherapy. In the review of Rades et al that includes 402 cases of GGs, local control (but not overall survival) was improved significantly with the addition of RT after subtotal resection in both the low grade and high-grade subgroups. After gross total resection, RT conferred no improvement in outcome for patients with either low-grade or high-grade tumors (8). Because of their location complete resection of infratentorial GG is often not possible without producing severe deficits or even death. In our case, the resection was not possible so the patient was treated with chemotherapy, followed by radiotherapy after progression. Cerebellar GGs are uncommon tumors that occur frequently during childhood and adolescence. CT scan and MR imaging could suggest the diagnosis but it's rarely made prior to surgery. With complete tumor resection, cerebellar GGs could be cured and no additional therapy is required. For unresecable GGs, or after subtotal resection, radiotherapy and/or chemotherapy should be considered.

Conclusion

Cerebellar GGs are uncommon tumors that occur frequently during childhood and adolescence. CT scan and MR imaging could suggest the diagnosis but it's rarely made prior to surgery. With complete tumor resection, cerebellar GGs could be cured and no additional therapy is required. For unresecable GGs, or after subtotal resection, radiotherapy and/or chemotherapy should be considered.

Abbreviations

GGs: Gangliogiomas

MRI: Magnetic resonance imagine

CT scan: computed tomography scanner

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Illustrations

Illustration 1

MRI (Axial) showing a cystic and solid lesion in the right lateral cerebellar hemisphere extending to the bulbar region and the bulbo-medullary junction

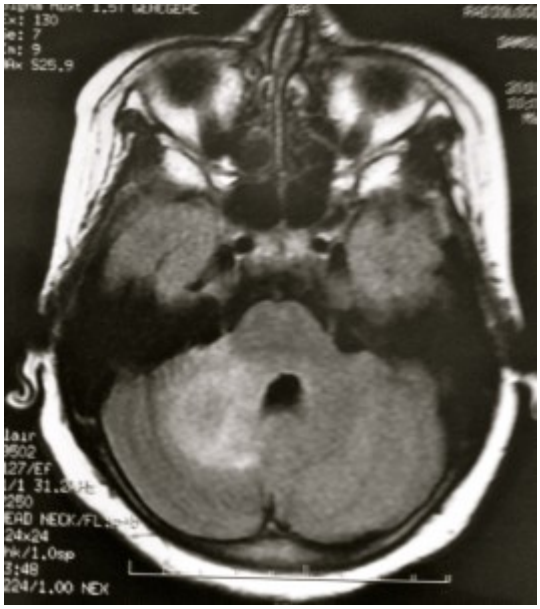


Illustration 2

MRI (Coronal) showing a cystic and solid lesion in the right lateral cerebellar hemisphere extending to the bulbar region and the bulbo-medullary junction

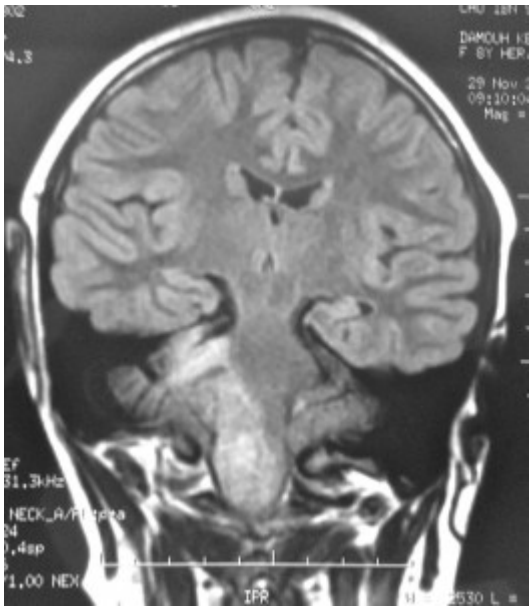
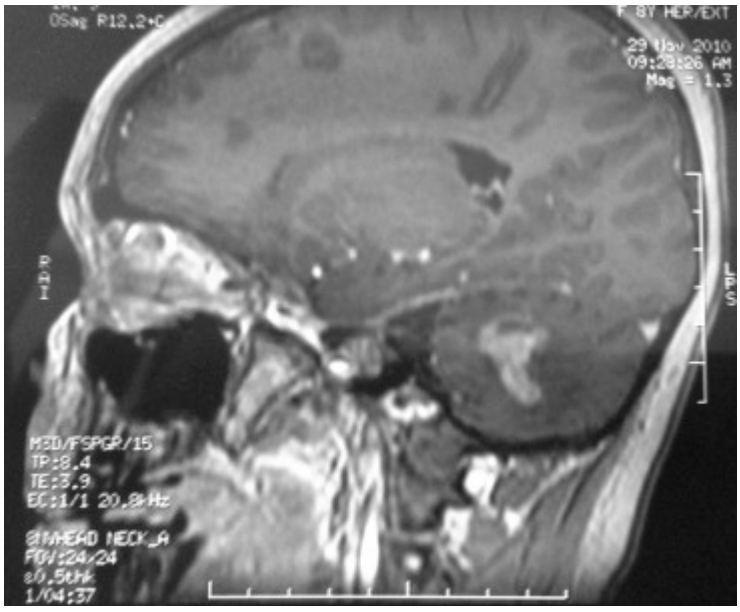


Illustration 3

MRI (Sagittal) showing a cystic and solid lesion in the right lateral cerebellar hemisphere extending to the bulbar region and the bulbo-medullary junction



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