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Case Series

Brainstem glioma clinical features and treatment outcomes: a case series

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ABSTRACT

A diverse category of gliomas that mostly affect youngsters are known as brainstem gliomas (BGs). The diffuse intrinsic pontine glioma (DIPG), exophytic medullary glioma, and tectal glioma can be divided into categories based on architecture and clinical behaviour. The most frequent BG is DIPG. The median age at start is 6.5 years, and the median survival is less than a year. The fact that adults with DIPG live longer suggests that their tumours are less aggressive and have physiologically distinct origins than those in children. Patients may appear with one or more of the following symptoms: ataxia, long tract signs, or malfunction of the cranial nerves. The majority of the pons is occupied by an infiltrative lesion on magnetic resonance imaging, and contrast enhancement is typically not noticeable. Fractionated radiation is the norm in medicine.

Keywords: BGs, Radiation, Temozolomide

INTRODUCTION

A malignant glioma tumor in the brainstem is referred to as a brainstem glioma. Though they have been found to afflict older persons as well, over 75% of diagnoses are made in children and young adults under the age of twenty. BGs often spread throughout the nervous system after beginning in the brain or spinal cord tissue.[1] Less than 2% of adult gliomas are these tumors, while they account for roughly 20% of all pediatric primary brain tumors.[2]

Due to problems in important areas of the brain, BGs are less frequently treated with neurosurgery than other types of brain tumors. Chemotherapy and/or radiation therapy are the most frequent forms of treatment.[3] A deadly and aggressive cancer, brainstem glioma. The life expectancy without therapy is usually only a few months after the diagnosis. With the right care, 37% make it past the first year, 20% make it to the second year, and 13% make it to the third. This statistic only applies to DIPG, not all BGs. Other BGs exist. Another is localized gliomas, which develop in several brainstem regions. While the median survival for adult brainstem glioma is in the bleak but not quite as grim range of 30-40 months, the prognosis for pediatric DIPG is a dismal 10 months, with just 10% of pediatric patients living >2 years beyond diagnosis.[4] Furthermore, predicting the prognosis for a specific patient at diagnosis can be challenging due to the considerable variation in the aggressiveness of adult BGs. Standard of care treatment for juvenile DIPG is extensive field radiation therapy, usually at a dose of about 54-60 Gy. Alternative pediatric DIPG fractionation plans, such as hyperfractionated, accelerated, and hypofractionated regimens, do not increase survival and considerably raise radiation toxicity concerns at doses greater than 64 Gy.[5] As with juvenile DIPG, extensive field radiation therapy, usually at a dose of 54-60 Gy, is regarded as standard upfront therapy for adults with BGs.

CASE SERIES

This was a retrospective study related to brainstem glioma, conducted in our hospital in which clinical and epidemiological features of five patients with brainstem glioma were analysed from the period of January 2019 till December 2021. The mean (standard deviation) of the continuous measurements was reported. The Mann-Whitney 'U' test was employed for non-parametric data, and the paired student t test was used for statistical comparisons of parametric quantitative variables. All statistical analyses were performed using the social science statistical system (SPSS version 20, SPSS Inc, Chicago, IL, USA) and a p=0.05 or lower was regarded as statistically significant.

A total of five cases of BGs were analysed, 3 (60%) were children and 2 (40%) adults. There were 1 (20%) female patient and 4 (80%) male patients. Headache, vomiting, seizures, neurological deficit were the presenting symptoms. All the five patients were having lesion >3 cm. Biopsy was available in none; only radiological diagnosis was available. As a result, all patients received treatment following an informed consent. Radiation therapy was administered to all five patients concurrently with temozolomide (75 mg/m) at a dosage of 54 Gy over the course of 27-30 fractions over a period of 6 weeks. Three patients received adjuvant temozolomide (150-200 mg/m^2) on days 1-5 every 28 days for a maximum of six cycles. There was no surgery on any patient. Due to hydrocephalus, the VPMP shunt was only administered to one patient. The median survival time was 22 months. The OS rates at 1 and 3 years after diagnosis were 71.4% and 53.6%, respectively.

DISCUSSION

The main glial tumour known as brainstem glioma develops within the brainstem and is thought to represent a diverse collection of gliomas. BGs have been split into two types by certain writers. The other 80% of tumours originate in and take up most of the pons, are diffuse in form, and have a bad prognosis.[6-8] Twenty percent are thought to be localised low-grade lesions with a favourable prognosis. Based on the information given, we were unable to determine whether the tumour started in the midbrain, medulla, or pons, or if it was localised or diffuse. According to the report from the central brain tumor registry of the United States, in children and adolescents, brainstem tumors account for 10.8% of all primary central nervous system tumors.[9] BGs were also reported to account for up to 20% or more of primary brain tumors.[10] However, we found that BGs could be diagnosed at every age. We believe that timely diagnosis and treatment are essential for patients. Focal radiation therapy is the current standard of care for children with DIPG.[11] We also found that radiation therapy was chosen more frequently than surgery and chemotherapy.

Dellaretti and colleagues investigated the correlation between magnetic resonance imaging findings and histological diagnosis of intrinsic brainstem lesions in adults in a series of 96 patients. Stereotactic biopsy established a precise histological diagnosis in 92 patients which consisted of 63 diffuse BGs, 19 other neoplastic diseases (lymphomas, metastases, pilocytic astrocytomas, craniopharyngioma, ganglioma) and 10 non-neoplastic lesions (inflammatory disease, ischemic lesion, fungal abscess, gliosis). Overall morbidity rate was 9% and one patient died from exacerbated peritumoral edema. With regard to neuroradiological features the diagnostic effect of stereotactic biopsy was greater in patients with focal or enhancing lesions shown by MRI in whom the diagnosis of a diffuse gliomas was less frequent.[12]

The value of additional imaging modalities to improve non-invasive diagnostic accuracy by MR spectroscopy or positron emission tomography is currently under investigation. However, massager showed recently in a series of 30 BGs that the integration of PET imaging cannot replace histological analysis as MRI combined with PET data was only concordant with histological findings in 63% of cases.[13] Reports about the use of antiangiogenic substances in the literature are rare. Besides two case reports a small series of 3 patients showed the effectiveness of bevacizumab as a salvage therapy for progressive BGs with improvement of clinical condition, reduction of daily dexamethasone dosage and radiological response.[14-16]

CONCLUSION

At any age, BGs can be identified. For patients, prompt diagnosis and appropriate treatment are crucial. Radiation is still the mainstay of treatment, but it is also important to investigate other options like interstitial radiosurgery, chemotherapy, and antiangiogenic drugs, ideally in the context of molecular profiling for common alterations like 1p/19q codeletion, MGMT promoter methylation, and IDH mutation.

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