Brain Stem Gliomas

Introduction

Brain stem gliomas are primary tumors occurring in the brain stem, the deep central/ posterior part of the brain that connects the rest of the brain to the top of the spinal cord. The brain stem consists of the medulla, pons, and midbrain. This critical area of the brain controls many of the vital functions of life such as heart rate and breathing, as well as nerves that control vision, hearing, speech, swallowing, and movement of both arms and legs. The term "brain stem glioma" most commonly refers to a malignant, diffuse (spreading) tumor that involves the entire pons (also called a diffuse pontine glioma). However, it is also used to describe focal, low-grade (slow-growing) tumors that can originate anywhere within the brain stem. Cervicomedullary gliomas and tectal gliomas are particular subtypes of low-grade brainstem gliomas whose epicenters are either in the medulla/upper cervical spinal cord junction or the posterior portion of the midbrain (the tectum), respectively.

Incidence

Approximately 75% of brain stem gliomas occur in patients younger than 20 years of age, with the peak incidence occurring between the ages of 5 and 10 years. Brain stem gliomas account 10-15% of pediatric brain tumors and less than 1% of adult brain tumors. There is an increased incidence of low-grade brain stem gliomas in patients with neurofibromatosis type 1. Brain stem tumors in older adults are more likely to be metastases from other sites than primary brain stem gliomas.

Diagnosis

Symptoms of a brain stem tumor frequently include cranial nerve dysfunction (double vision, drooping of one eyelid or the whole side of the face, difficulty swallowing), weakness in the arms and legs, impaired ability to walk, and blockage of the flow of cerebrospinal fluid in the brain (hydrocephalus) that leads to headache, vomiting and fatigue. Behavioral changes and seizures may also be seen in patients with brain stem gliomas.

Brain stem gliomas are one of the few types of brain tumors whose diagnosis is commonly made by physical examination and brain MRI only, without a biopsy. A diagnosis of diffuse pontine glioma can be made when a patient presents with a non-enhancing mass that diffusely expands the pons on MRI, has one or more cranial nerve findings on exam, and identifies symptoms present for less then 6 months (usually less than 3 months). Focal tumors of the tectum or cervicalmedullary junction that typically enhance with gadolinineum on MRI tumors are also classic and do not usually have to be biopsied. Brain stem tumors that appear atypical, protrude out from the brainstem, or do not meet any of the above criteria are often biopsied to confirm the diagnosis.

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Treatment
Due to the vital importance of this area of the brain and the frequently infiltrative nature of these tumors, surgical resection or biopsy of most brain stem gliomas is usually not possible. Radiation therapy and/or chemotherapy are the current mainstays of treatment for most brain stem gliomas.

Patients with diffuse pontine gliomas are often enrolled in an open clinical trial and treated with a combination of focal radiation therapy and experimental chemotherapy.

Focal brain stem tumors, particularly tumors that occur in the midbrain or protrude from the medulla or pons are sometimes biopsied or partially resected. For these tumors, following any surgery, focal radiation therapy and/or standard low-grade glioma chemotherapy is used. Prognosis for this group of patients is significantly better than those diagnosed with diffuse pontine gliomas, but not a good as those diagnosed with low-grade gliomas in other parts of the brain.

The tectal gliomas are a particular sub-set of low-grade brainstem gliomas whose growth is usually so slow and indolent that these patients are frequently observed with no treatment for long periods of time. If any treatment is necessary, it is usually involves surgically relieving any hydrocephalus by cerebrospinal shunting or a third ventriculostomy.

About the Brain Tumor Society
Resources and supportive services are always available at the Brain Tumor Society. We offer a wealth of information for brain tumor patients, long-term survivors and families coping with a brain tumor diagnosis.