INTRODUCTION

Brainstem gliomas are characterized by heterogeneous biologic behavior, ranging from low-grade tumors needing little treatment to those that are rapidly fatal despite aggressive therapy [1,2]. Prognosis and treatment depend upon both the clinical symptoms and their duration as well as its location within the brainstem.

Approximately 80 percent of pediatric brainstem gliomas arise within the pons, while the remaining 20 percent arise in the medulla, midbrain, or cervicomedullary junction (figure 1) [3-8]. The majority of pontine tumors are diffuse intrinsic brainstem gliomas, which are usually high-grade, locally infiltrative, and have a uniformly poor prognosis [9]. Histologically, these tumors are usually anaplastic astrocytomas (World Health Organization [WHO] grade III) or glioblastoma multiforme (WHO grade IV) (table 1). However, patients with WHO grade II tumors identified by biopsy do not have an improved prognosis. (See "Classification of gliomas", section on 'Histopathologic and molecular classification'.)

In contrast, most nonpontine tumors involving the cervicomedullary junction and tectum, as well as focal, cystic and dorsal exophytic lesions, are low-grade astrocytomas, mostly grade I pilocytic astrocytomas [6]. These are discrete, well-circumscribed tumors without evidence of locally invasive growth or edema [10]. Approximately 10 to 20 percent of non-pontine gliomas will be high-grade and are treated similar to diffuse intrinsic pontine gliomas.

Diffuse pontine gliomas will be reviewed here. Gliomas arising from other sites within the brainstem are discussed separately. (See "Focal brainstem glioma".)

EPIDEMIOLOGY

Gliomas arising in the brainstem (midbrain, pons, and medulla oblongata) account for 10 to 20 percent of all central nervous system (CNS) tumors in children. Brainstem gliomas are more common in children than adults [3,4,11,12]. In the United States, for example, there are approximately 300 pediatric cases and 100 adult cases reported each year. In children, the median age at diagnosis is five to nine years of age, and the incidence is approximately equal between males and females.
References


