Surgical indication and technical considerations in the management of benign brain stem gliomas

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Summary

The treatment of brain stem gliomas has evolved over the last few decades, reflecting advances in imaging (MR), microsurgical techniques and biological understanding. The aim of this chapter is to provide a preoperative classification for intrinsic brain stem lesions that will predict histopathology and biological behavior from the clinical syndrome and the MR appearance. Such a classification system may help selecting children with brain stem tumors that can benefit from surgery. Technical considerations, potential surgical complications, and the ways to avoid them are discussed.

Introduction

Brain stem (BS) gliomas represent between 6 and 15\% of pediatric intracranial tumors [1–11]. As a group, these tumors offer great resistance to therapeutic intervention and, overall, are associated with a 30\% five year actuarial survival rate [9].

Bailey, based on Bucy's experience, stated that gliomas of the brain stem are a 'hopeless problem for treatment'. Six of eight patients died immediately after surgery and the other two within months of their operation [12]. Dandy wrote pertaining to medullary lesions that '... there is little indication for attempting any enucleation of a tumor in this region' [13]. Matson considered these lesions malignant irrespective of the histopathology, given their location, and favored a non-interventional approach [14]. Recognition of the true heterogeneity of these tumors emerged slowly. In 1964, Baker reported on a series of patients with 'subependymal gliomas' arising just below the ependyma of the fourth ventricle. He described three subtypes, based on the postoperative neurological sequelae [15]. In 1968, Pool described a small group of patients with brain stem lesions that could be resected with some of them having long-term survivals [16].

In the past fifteen years, there has been a resurgence of interest in these tumors, largely stimulated by more information on long-term disease-free survivals. The concept of heterogeneity of brain stem tumors (BST) was further explored and some groups have established the presence of prognostic factors that appear to influence outcome in patients with brain stem gliomas. Other groups have identified sub-categories of tumors that appear to have low-grade pathologies and offer a better prognosis [17–22]. It is clear, therefore, that all BST's should not be grouped together and that with modern imaging and technology, some of these lesions should be treated surgically [7, 20, 22–32].

The most important technological advancement encouraging the pursuit of operative treatment for brain stem gliomas has been the availability of the MR scan. This diagnostic modality has made it possible to identify the precise localization of a neoplasm within the brain stem and perhaps more im-
portantly to suggest, together with the clinical picture, the microscopic pathology with a relatively high degree of probability. An MR-based surgical classification of brain stem gliomas has been developed to facilitate decision-making regarding these lesions [20, 31–38].

While serious potential complications are inherent to this surgery, they become ‘inevitable’ if it is undertaken in inappropriate cases. The ‘key to success’ in brain stem surgery is to operate on low-grade localized neoplasms before there is significant disability. It is the purpose of this chapter to first address the issues which relate to patient selection and, following this, to discuss specifics of surgical technique and intraoperative management.

**Classification**

It is now recognized that there is no single neoplasm which may be appropriately described as a ‘brain stem tumor’. Classifying BST’s serves several purposes. First, it allows pathological prediction from the preoperative imaging and clinical syndrome. Second, it helps in planning the surgical procedure, if surgery is indicated. Finally, it helps in predicting tumor behavior and prognosis. Several attempts have been made over the past 10 years to look into the relationship between anatomy, the clinical syndrome, histopathology, and outcome. These attempts will be summarized in the following paragraphs.

The first and most important MR-based differentiation of BST’s is between diffuse lesions (DL) and all other tumors. Diffuse lesions are typically

**Fig. 1.** Axial T2-weighted images of a focal pontine lesion (A) and a diffuse pontine tumor (B). Note that the diffuse tumor grows across the entire cross-section of the brain stem. In the patient with the focal lesion, there is preserved brain stem tissue anterior to the tumor.