

Review

Neuroblastoma: Tumor Biology and Its Implications for Staging and Treatment

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Abstract: Neuroblastoma, the most common extracranial solid tumor of childhood, has widely variable outcomes dependent on the specific biology of the tumor. In this review, current biologic principles that are used to stratify risk and guide treatment algorithms are discussed. The role for surgical resection in neuroblastoma is also reviewed, including the indications and timing of surgery within the greater treatment plan.

Keywords: neuroblastoma; oncology; pediatric; pathology; surgery

1. Introduction

Neuroblastoma is the most common extracranial solid tumor of childhood and represents a neoplastic expansion of neural crest cells in the developing sympathetic nervous system. The primary tumor originates anywhere along the sympathetic chain but most frequently arises from the adrenal gland. The prognosis for neuroblastoma varies widely, from tumors that spontaneously regress and require no intervention to those that present widely metastatic and resistant to therapy with resulting high mortality. This disparate prognosis is largely dependent on tumor biology, and extensive research has been completed identifying tumor characteristics that associate with aggressive tumor behavior and poor prognosis. Specifically, the expected tumor biology can be predicted by tumor histology and molecular markers, both of which are strongly associated with patient age.

2. Tumor Histology

Neuroblastoma tumor cells show varying degrees of differentiation that help predict patient prognosis. While neuroblastoma primarily contains immature cells, some have a component of fully mature ganglion cells that are typically found in a ganglioneuroma. A tumor with both elements of mature and immature cells is called a ganglioneuroblastoma. Tumors have been classified in detail according to this degree of differentiation by the International Neuroblastoma Pathology Committee [1–3] (Figure 1). Favorable and unfavorable histologic subtypes are based upon the level of Schwannian stroma present in the tumor, then further subclassified based upon the mitosis-karyorrhexis index (MKI) and patient age.

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