Pediatric Malignancies

Neuroblastoma, Wilm's Tumor, Hepatoblastoma, Rhabdomyosarcoma, and Sacroccygeal Teratoma

Katherine P. Davenport, MD[a], Felix C. Blanco, MD[a],
Anthony D. Sandler, MD[b,*]

KEYWORDS

• Pediatric malignancies • Wilm's tumor • Neuroblastoma • Hepatoblastoma
• Sacrococcygeal teratoma • Rhabdomyosarcoma

KEY POINTS

- The aggressive behavior of high-risk neuroblastoma results in incomplete response to multimodality treatment.
- Multimodality therapies have led to improved prognosis and excellent cure rates for patients with Wilms tumor.
- Prognosis of hepatoblastoma is improved with combination of surgery and neoadjuvant therapy.
- The survival rate of children with rhabdomyosarcoma is improved with combination therapy.
- Sacrococcygeal teratoma is associated with significant perinatal concerns. Following complete en bloc excision, the overall prognosis is good.

NEUROBLASTOMA

Neuroblastoma is the most common extracranial solid tumor in children. Despite the excellent prognosis of neuroblastoma when diagnosed early, the aggressive behavior of advanced-stage neuroblastoma results in a very poor prognosis, despite the use of multimodal therapy.

Incidence

The incidence of neuroblastoma varies in different regions of the world, with a greater prevalence in developed countries of 1 in 7000 children.[1] The most common age at

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[a] Sheikh Zayed Institute for Pediatric Surgical Innovation, Children's National Medical Center, 111 Michigan Avenue NW, Washington, DC 20010, USA; [b] Department of Surgery, Sheikh Zayed Institute for Pediatric Surgical Innovation, Children's National Medical Center, 111 Michigan Avenue NW, Washington, DC 20010, USA
* Corresponding author.
E-mail address: ASandler@cnmc.org

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presentation is 18 months and equally affects males and females. Neuroblastoma is more prevalent in children younger than 4 years (85%), declining to 8% at 9 years and 1.5% after 15 years of age.[2]

Etiology

Through embryologic development, neuroblastic cells migrate from the neural crest to peripheral organs in the neck, chest, and abdomen. During this process, physiologic forces drive neuroblasts to differentiate into adult neuronal tissue that forms the sympathetic chain and adrenal medulla. Neuroblastoma is neuronal tissue derived from the neural crest that remains or becomes (de-)undifferentiated after reaching its peripheral location, adopting the appearance of small, round blue cells with neuritic processes.[3] Evidence of this embryologic development is seen in vestigial remnants of neuroblastoma tissue occasionally observed in the adrenal gland and sympathetic chains of otherwise normal newborns.[4]

No theory clearly explains the origin of neuroblastoma. Abnormalities in the rearrangement of chromosome 1p and 11q are reported.[5] Similarly, neuroblastoma was linked to neurofibromatosis and Hirschsprung disease that could explain its association with defects of neural crest development.[6]

Distribution

Primary neuroblastoma tumors mainly originate in the following body compartments: craniofacial and neck in 1% to 3% of patients; mediastinum in 16% of patients; and retroperitoneum, with adrenal (48%) and extraadrenal (25%) locations.

Classification and Staging of Neuroblastoma

The classification of neuroblastoma was for a long time based solely on the extension of the primary tumor, according to the International Neuroblastoma Staging System (INSS). Although useful and widely accepted, the INSS was unable to clearly assess the patient's risk status or to stage the patient in the preoperative setting, because the system was mainly based on operative findings. Furthermore, clinical trials using the INSS could not uniformly compare or assess the status of extraregional nodal disease.[7,8]

Briefly, the INSS includes[9]

- Stage 1: localized, unilateral tumor with negative lymph nodes, completely excised
- Stage 2A: localized, unilateral tumor with negative lymph nodes, incompletely excised
- Stage 2B: localized, unilateral tumor with ipsilateral positive lymph nodes, completely or incompletely excised
- Stage 3: unresectable, unilateral tumor infiltrating across the midline, with or without regional node involvement
- Stage 4: tumor with metastasis to distant lymph nodes, bone marrow, liver, skin, and other organs
- Stage 4S: localized tumor with metastasis to skin, liver, and bone marrow without involvement of cortical bone in infants younger than 1 year (<10% of cells are malignant).

To overcome some of the limitations of the INSS, the International Neuroblastoma Risk Group designed a staging system (INRGSS) to provide information about tumor resectability based on diagnostic imaging using CT scan or MRI.[7] The INRGSS