sympathetic nervous system. Consequently, the majority of the tumors arise from the adrenal gland or from the retroperitoneal sympathetic chain. The primary site is within the abdomen; other sites include the head and neck region, chest, and pelvis.

Clinical Manifestations

The signs and symptoms of neuroblastoma depend on the location and stage of the disease. With abdominal tumors, the most common presenting sign is a firm, nontender, irregular mass in the abdomen that crosses the midline (in contrast to Wilms tumor, which is usually confined to one side). Other primary tumor sites may cause significant clinical effects such as neurologic impairment, respiratory obstruction from a thoracic mass, or varying degrees of paralysis from compression of the spinal cord.

Distant metastasis frequently causes supraorbital ecchymosis, periorbital edema, and proptosis (exophthalmos) from invasion of retrobulbar soft tissue. Lymphadenopathy, hepatomegaly, and skeletal pain are also present in patients with disseminated disease. Vague symptoms of widespread metastasis include pallor, weakness, irritability, anorexia, and weight loss.

Diagnostic Evaluation

Diagnostic evaluation is aimed at locating the primary site and areas of metastasis. A CT scan of the abdomen, pelvis, or chest is the preferred imaging modality to locate the primary tumor. A bone scan and MIBG (iodine-131 metaiodobenzylguanidine) scan should be performed to evaluate for the presence of skeletal metastases. Examination of the bone marrow with bilateral aspirates and biopsies should be performed in all patients. Neuroblastomas, particularly those arising on the adrenal glands or from a sympathetic chain, excrete the catecholamines epinephrine and norepinephrine. Urinary excretion of catecholamines is detected in approximately 95% of children with adrenal or sympathetic tumors.

Staging and Prognosis

Neuroblastoma is a “silent” tumor. In more than 70% of cases, diagnosis is made after metastasis occurs, with the first signs