

Alveolar: Second most common type; most often seen in deep tissues of the extremities and trunk

Pleomorphic: Rare in children (adult form); most often occurs in soft parts of extremities and trunk

Clinical Manifestations

The initial signs and symptoms are related to the site of the tumor and compression of adjacent organs. Some tumor locations, such as the orbit, manifest early in the course of the illness. Other tumors, such as those of the retroperitoneal area, only produce symptoms when they are relatively big and compress adjacent organs. Unfortunately, many of the signs and symptoms attributable to rhabdomyosarcoma are vague and frequently suggest a common childhood illness, such as “earache” or “runny nose.” Often the site of the primary tumor site is never identified.

Diagnostic Evaluation

Diagnosis begins with a careful history and physical examination. Radiographic studies to delineate the primary tumor site should include PET/CT or MRI scans. Metastatic evaluation should include a CT of the chest, bone scan, and bilateral bone marrow aspirates and biopsies. For patients with tumors in the parameningeal area, an LP is performed to examine the spinal fluid. An excisional biopsy or surgical resection of the tumor, when possible, is done to confirm the diagnosis.

Staging and Prognosis

Careful staging is extremely important for planning treatment and determining the prognosis. The Intergroup Rhabdomyosarcoma Study has developed a surgicopathologic staging system, which includes four stage classifications depending on disease involvement.

With the use of contemporary multimodal therapy, more than 60% of patients with nonmetastatic disease are expected to survive, and if diagnosed in the early stage, the survival rate increases to 80% ([Davenport, Blanco, and Sandler, 2012](#)). If relapse occurs, the prognosis for long-term survival is poor.