Rhabdomyosarcoma

The rhabdomyosarcoma accounts for 20 per cent of all soft tissue sarcomas. The embryonal and alveolar types are seen in the pediatric age group while the less common pleomorphic rhabdomyosarcoma is seen in adults.

The embryonal rhabdomyosarcoma occurs in children between birth and the age of 15 years, more often in boys than girls, with the head and neck area being the most common location for this tumor. Histologically, it is a round cell tumor similar to Ewing's sarcoma but in 50 per cent of cases there will be evidence of cross striations in associated spindle cells and the tumor will have immunohistochemical markers for desmin, myoglobin, and actin. Prior to the advent of chemotherapy, this tumor was fatal in about 90 per cent of cases but with current chemotherapy protocols over 80 per cent of patients survive this tumor. Treatment consists of local surgical resection when indicated, along with postoperative radiation therapy if the surgical margins are considered contaminated.

The alveolar form of rhabdomyosarcoma affects children and young adults between the ages of 10 and 25 years, and is more common in males than females. It occurs in the head and neck area, but can also develop in the extremities, especially the thigh and calf. Histologically, this variant has a typical alveolar pattern of round cells that gives it its clinical name and only rarely will rhabdomyoblasts be present in histological specimens. Treatment consists of surgical resection followed by radiation therapy if the margins are positive, and adjuvant perioperative chemotherapy. The prognosis for survival is much worse than with the embryonal rhabdomyosarcoma.

The pleomorphic rhabdomyosarcoma is the rarest variant of the group, consisting of 5 per cent of all cases. It occurs in middle-aged or older patients and is most commonly located in large muscle groups in the proximal extremities, usually the lower extremities. In the 1940's, the pleomorphic rhabdomyosarcoma was a common histological diagnosis that included many cases of malignant fibrous histiocytoma (MFH). At that time, MFH was an unpopular histological diagnosis but is now a common one by current pathological criteria. Microscopically, this variant consists of large, bizarre-appearing giant cells with very atypical nuclei. The cells stain positive for glycogen and cross striations appear in the so-called "candy ribbon" cellular patterns with tandem nuclei on the surface of these rectangular cells. This variant carries a very poor prognosis for survival and the use of adjuvant

chemotherapy is not nearly as effective as it is for the pediatric variants. The mainstay of treatment is wide local resection, when possible, followed by radiation therapy, even with negative surgical margins.