Malignant Fibrous Histiocytoma

The malignant fibrous histiocytoma (MFH) is the most common high-grade soft tissue tumor. It typically occurs in patients over the age of 50 years and is more common in men. The most common type of MFH is a so-called storiform or pleomorphic malignant fibrous histiocytoma, followed next by the myxoid, giant cell, and inflammatory subtypes. The pleomorphic form is seen in patients between the age of 50 and 70 years, and is usually found deep in a large muscle about the thigh, hip or retroperitoneal area. These tumors usually present with symptoms of local pain along with a tumor mass that has been present for at least three or four months, but sometimes they can be asymptomatic. The gross appearance of the tumor is multi-nodular lesion that has separate satellite lesions located within the same muscle belly, usually at the upper or lower pole of the lesion. On MRI, the lesions may demonstrate central necrosis. The color is a dirty gray to a reddish tan. Because this tumor is usually high grade, is demonstrates a high index of mitotic activity and the cell types usually found are malignant fibroblasts mixed with anaplastic pleomorphic histiocytes. This tumor has the potential to metastasize to the lung in about 40-45 per cent of cases. It goes to regional lymph nodes in 10 per cent of cases. With appropriate surgical and adjuvant therapy, the local recurrence rate is in the neighborhood of 10-15 per cent after a wide surgical resection. When these tumors are located in a superficial, subcutaneous location and present with a size less than 5 cm, the overall prognosis is much better, with only a 20 per cent chance of metastasis to the lung. However, in the more common, deeply located lesions measuring over 5 cm in diameter, the overall survival rate is 55 per cent.

The usual treatment for the high-grade MFH is a wide surgical resection when possible. If the tumor is too large and cannot be resected safely because of major neurovascular invasion or multiple compartmental contamination, aggressive high level amputation is sometimes indicated. Following the surgical resection, patients with the more aggressive lesions are usually treated with adjuvant postoperative radiation therapy, in the neighborhood of 6500 centigray to the tumor site, with a wider field of 5500 centigray to include a wider field outside the surgical site. At some institutions, patients will be treated with preoperative radiation therapy, up to about 5500 centigray, with the additional 100 centigray being delivered two weeks after the surgery. The use of adjuvant chemotherapy for high-grade soft tissue sarcomas is quite controversial because of minimal data to suggest that the prognosis is significantly improved with this adjuvant therapy, and thus, medical oncologists would discourage the use of this

adjuvant, especially in older patients who cannot tolerate the high toxicity levels of current protocols.

The myxoid variant of the MFH is the second most common type of malignant fibrous histiocytoma. It grossly has a myxoid appearance and, because of this, the chance for local contamination at the margins (with local recurrence) is much higher. However, the metastatic rate to the lung is only 25 per cent giving the patient a better prognosis for survival.

The giant cell variant, occurring in large muscle groups in older patients, has a tendency to be a hemorrhagic tumor with a high pulmonary metastatic rate of 50 per cent. The inflammatory MFH is also seen in older patients and is more common in the retroperitoneal area, with a 50 per cent lung metastasis rate.