Pleomorphic sarcoma of the neck

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Pleomorphic sarcoma is the World Health Organization’s preferred term for malignant fibrous histiocytoma. This high-grade pleomorphic malignant mesenchymal neoplasm is a diagnosis of exclusion after other sarcomas and pleomorphic neoplasms have been excluded by histochemistry, immunohistochemistry, electron microscopy, and/or molecular evaluation. Most of these tumors arise de novo, but postradiation tumors are not uncommon. To be considered as a postradiation tumor, the tumor must be located in the radiation field and it must develop at least 3 years after radiation in an area that was free of tumor before radiation.

Pleomorphic sarcoma is an uncommon neoplasm in the head and neck now that refinements in diagnostic techniques have more accurately classified tumors that used to be placed in this category. Pleomorphic sarcoma usually develops in older adults (the sixth and seventh decades of life), and men are affected more often than women. Within the head and neck, the two most common sites are the sinonasal tract and the neck. Most patients present with a mass with or without pain. Complete surgical excision is the treatment of choice, with adjuvant therapy being employed in many patients. Lymph node metastasis is seen in approximately 15% of cases; metastatic disease is more commonly seen in the lung, liver, and bone. Tumor depth, size, and stage are strongly associated with the prognosis.

Macroscopically, pleomorphic sarcomas are nodular or multinodular, tan-white to gray tumors that may be associated with necrosis and hemorrhage (figure 1). The myxoid lesions are translucent or gelatinous. These tumors are usually uncircumscribed, and they exhibit an infiltrative pattern into adjacent soft tissues, skeletal muscle, nerves, and even vessels. The cells are

Figure 1. This pleomorphic sarcoma exhibits surface erosion and ulceration. There is a large tumor expanding into the adjacent adipose tissue, present at the deep margin and showing a variegated appearance with hemorrhage and necrosis.

Figure 2. Image shows pleomorphic cells along with multinucleated tumor giant cells. Increased mitoses, including atypical forms, are easily identified in this intermediate-power field.