

Pleomorphic sarcoma of the neck

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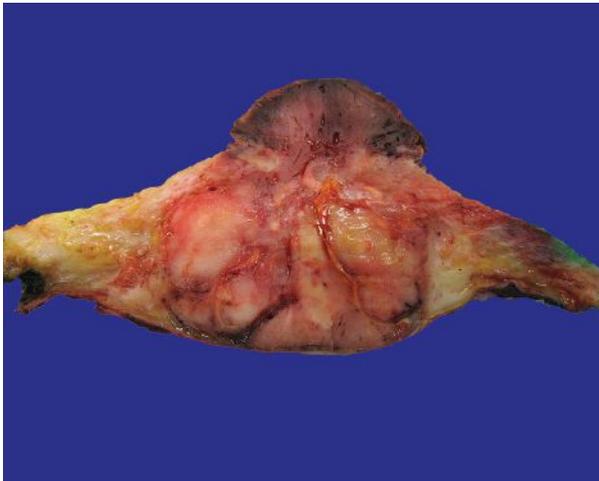


Figure 1. This pleomorphic sarcoma exhibits surface erosion and ulceration. There is a large tumor expanding into the adjacent adipose tissue, present at the deep margin and showing a variegated appearance with hemorrhage and necrosis.

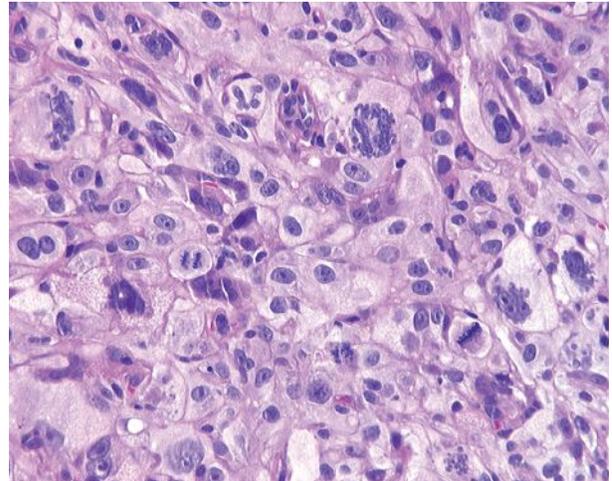


Figure 2. Image shows pleomorphic cells along with multinucleated tumor giant cells. Increased mitoses, including atypical forms, are easily identified in this intermediate-power field.

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 unencapsulated, and they exhibit an infiltrative pattern into adjacent soft tissues, skeletal muscle, nerves, and even vessels. The cells are

profoundly pleomorphic and arranged in a storiform, haphazard architecture. There are short fascicles in a pinwheel or cartwheel configuration. The tumors are generally hypercellular, made up of spindle cells and epithelioid cells. Nearly all tumors are high grade. Neoplastic multinucleated giant cells are common, and reactive foreign-body-type giant cells may also be seen (figure 2).

Several histologic variants are reported, with myxoid, angiomatoid, giant cell, and inflammatory types most common in this location. There are usually easily identified mitoses, atypical mitoses (figure 2), and necrosis and/or hemorrhage.

Xanthoma cells (foamy histiocytes) are usually easy to find in pleomorphic sarcomas. Occasionally, heterologous elements (such as bone or cartilage) are seen. Inflammation may be present; when it is, it usually suggests a better prognosis, as does the presence of a myxoid stroma. The myxoid foci are hypocellular, but they still show pleomorphism. Occasional lipoblast-like cells may be suggested.

In general, immunohistochemical studies show positivity with vimentin, CD68, CD10, and other nonlineage markers. There is an absence of epithelial, melanocytic, myogenic, and hematolymphoid markers.

The differential diagnosis includes high-grade liposarcoma, leiomyosarcoma, rhabdomyosarcoma, melanoma, carcinoma, and lymphoma (anaplastic type).

Suggested reading

- Goldblum JR. An approach to pleomorphic sarcomas: Can we subclassify, and does it matter? *Mod Pathol* 2014;27(Suppl 1):S39-46.
- Matushansky I, Charytonowicz E, Mills J, et al. MFH classification: Differentiating undifferentiated pleomorphic sarcoma in the 21st Century. *Expert Rev Anticancer Ther* 2009;9(8):1135-44.