Undifferentiated Pleomorphic Sarcoma of Skin: Clinical and histopathologic emulator of atypical fibroxanthoma, distinction imperative

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90 year old Caucasian man with history of several non-melanoma skin cancers, presented with a 4.0 x 2.5 cm ulcerated, friable, exophytic mass on the left mid frontal scalp of two months duration. The patient had previously presented two months earlier with non-healing scalp lesion. At that time, the lesion was a 0.9 cm ulcerated, erythematous, papule. A shave biopsy of the lesion was performed however the histopathology was non-diagnostic and demonstrated marked parakeratosis, fibrosing granulation tissue in the upper dermis, with a massive neutrophilic infiltrate. A repeat biopsy was performed of the exophytic mass for diagnostic and de-bulking purposes. The histopathology of the re-biopsy demonstrated an ulcerated tumor filling the dermis. The tumor cells were pleomorphic, spindled, arranged in vague fascicles, and extended to the deep margin. The cytology was markedly atypical, with large irregular nuclei, prominent nucleoli, and numerous mitoses, including atypical forms. Immunohistochemical analysis was performed. The tumor cells were diffusely positive for CD10 and weakly positive for CD68. Cytokeratin AE1/AE3, desmin, S-100, EMA, cytokeratin 5, p63, Mart-1, smooth muscle myosin, procollagen 1, ERG, CD31, and CD 34 were all negative.

**Histopathology**

The patient subsequently underwent wide local excision with one centimeter margins. The specimen showed atypical spindled cells extending deep into subcutaneous tissue. Due to the depth of invasion with involvement of subcutaneous tissue and the presence of necrosis, the lesion was diagnosed as an undifferentiated pleomorphic sarcoma of skin. The margins were free of tumor however the patient was referred to oncology for further evaluation and consideration of adjuvant therapy. The patient and family declined oncology referral, and as of four months post-excision, there was no evidence of recurrence.

**Discussion**

Undifferentiated pleomorphic sarcoma (UPS) of skin can clinically and histopathologically mimic atypical fibroxanthoma (AFX). Distinguishing between the two is important, as the prognoses of these tumors are vastly different. AFX follows more of a benign course, typically recurs only after incomplete excision, and rarely metastasizes. UPS, previously grouped into the malignant fibrous histiocytoma (MFH) category, is more aggressive in nature, and has a high rate of recurrence along with malignant/metastatic potential.

Clinically, UPS and AFX present as a rapidly growing solitary nodule on sun-damaged, actinic skin of the elderly, usually on the head and neck region. UPS is considered a soft tissue tumor but can occur superficially in the skin, with a presentation mimicking AFX.

Histologically, UPS and AFX consist of spindle shaped cells arranged in a fascicular pattern and can exhibit multinucleation, pleomorphism, and mitotic figures. UPS is distinguished from AFX by deep subcutaneous involvement, perineural and/or lymphovascular invasion, and necrosis. Immunohistochemically, both stain negative for S-100/SOX-10, cytokeratin, CD31/CD34, and desmin/myosin allowing differentiation from other pleomorphic tumors in the skin, such as melanoma, squamous cell carcinoma, angiosarcoma, and leiomyosarcoma. AFX and UPS are diagnoses of exclusion, requiring broad lineage-specific immunohistochemical analysis to exclude other poorly differentiated tumors.

**References**