## Atypical fibroxanthoma: A malignant tumor of the skin and soft tissue

*To the Editor*: In the most recent edition of the *World Health Organization (WHO) Classification of Tumours of Soft Tissue and Bone*, atypical fibroxanthoma (AFX) is described as a "benign" tumor of the skin and subcutaneous tissue.[1] This has been cited in Bolognia and colleagues' most recent version of *Dermatology*.[2] We disagree with the assertion that AFX is a benign tumor.

AFX is an uncommon fibrohistiocytic tumor that is widely considered a superficial, less aggressive variant of cutaneous undifferentiated pleomorphic sarcoma (UPS) or cutaneous pleomorphic dermal sarcoma (PDS). Clinically, it presents as a pink-to-red exophytic papule, usually on the head and neck of older men. Risk factors include ultraviolet light, radiation, and immunocompromised status.[3] A biopsy, including the full thickness of the dermis, is required to adequately exclude other diagnoses. Superficial sampling of a suspected lesion is insufficient because the tumor can be deep seated.

AFX can grow rapidly and cause local destruction. Tumors can occur at unusual sites, at which detection and removal may be morbid, including the ethmoid sinus, eyelid, and cornea, and even the parietal bone diploic space.[3] Perineural invasion, metastasis, and recurrence are also seen. Perineural invasion, when involving larger nerves, may present as burning, numbness, paralysis, and weakness. Small-nerve perineural invasion can greatly complicate the process of tumor extirpation, thereby increasing the likelihood of tumor persistence or recurrence. Perineural invasion may be relatively common in AFX.[4]

Apart from local tissue destruction, metastases with AFX have been reported at various anatomic sites, including liver and lung, and in pericutaneous tissues such as lymph nodes and the subcutis. The metastatic rate of AFX is 1% to 2% after surgery, but this may be an underestimate due to its rarity and potential misclassification.[5]

Various recurrence rates have been reported depending on the treatment modality and duration of follow-up.[3] Indirect comparisons based on numerous case studies and systematic reviews suggest tumors treated with Mohs micrographic surgery (MMS) may be more likely to be completely removed and have lower recurrence rates than those undergoing wide local excision (WLE). A comparison of WLE and MMS including 914 tumors found a recurrence rate of 2.0% for MMS compared with 8.7% for WLE.[5] This risk increases with immunosuppression.[6] Radiation may be used as adjuvant therapy for recurrent or metastatic disease.

Recognizing that AFX is a superficial variant of cutaneous UPS is necessary to ensure that it is adequately treated and studied as a rare cutaneous malignancy. Because there are no definitive histologic criteria to distinguish AFX from the highly aggressive UPS, classification of AFX as a benign tumor may inadvertently lead to underdiagnosis and undertreatment of UPS. AFX and UPS are on a diagnostic spectrum without clear distinction; therefore, complete removal of both is essential to avoid recurrence and metastasis.

In summary, AFX behaves like a malignant tumor. Local tissue destruction, pathologic features that complicate surgical extirpation, substantial risk of recurrence, and risk of metastasis are seen. The new WHO classification overlooks these known risks and may prompt inadequate treatment. Further research is needed to determine optimal margins for WLE, directly compare different surgical modalities, and compute rates of recurrence-free survival or progression-free survival.

Physicians who treat AFX realize it behaves like other invasive cutaneous malignancies and must be managed similarly. We hope the WHO will reconsider their classification of AFX based on the weight of the evidence.

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