# Atypical Fibroxanthoma

# 30

## Definition

Atypical fibroxanthoma (AFX) is a tumor of intermediate malignant potential classified among the soft tissue tumors of uncertain differentiation. The term "superficial malignant fibrous histiocytoma" should no longer be utilized.

It belongs to a group of atypical cutaneous spindle cell neoplasms referred to as the *SLAM*, which stands for *s*pindled squamous cell carcinoma, *l*eiomyosarcoma, *A*FX, and spindled *m*elanoma.

## Epidemiology and Presentation

AFX is a dermal-based lesion that typically presents as a red or pink rapid-growing nodule or papule (sometimes ulcerated) or plaque arising on sun-damaged skin of elderly patients (men are affected much more frequently than women), with special regard to head and neck region (90% of cases). Usually the nodule measures less than two centimeters and is asymptomatic.

Dermoscopy reveals polymorphic vessels (linear, dotted, hairpin, arborescent, and/or highly tortuous vessels) radiating to the center of the lesion with intervening white areas.

The differential diagnosis includes basal cell carcinoma, squamous cell carcinoma, Merkel cell carcinoma, amelanotic melanoma, leiomyosarcoma, atypical dermatofibroma, pleomorphic dermal sarcoma, and metastasis from other malignancies.



S. Mocellin, *Soft Tissue Tumors*, https://doi.org/10.1007/978-3-030-58710-9_30

## Etiology and Predisposition

AFX etiology is poorly understood. Ultraviolet light is believed to play an important role since most lesions appear on the sun-exposed head and neck area of Caucasian patients. The incidence is increased in immunosuppressed populations.

## Pathology

AFX presents as a circumscribed dermal nodule usually within sun-damaged skin (elastosis). It is composed of spindled to round or epithelioid tumor cells organized in a fascicular pattern. Bizarre multinucleated pleomorphic cells are present. High mitotic rate and many atypical mitotic figures are also present. It closely resembles undifferentiated pleomorphic sarcoma (which is also called pleomorphic dermal sarcoma when arising from the dermis, → see dedicated section) but centered in dermis.

The tumor should not extensively involve the subcutaneous tissue or deeper structures (i.e., muscle or fascia): if so, the lesion may represent a pleomorphic dermal sarcoma.

Several variants have been described, such as the following: angiomatoid, chondroid, clear cell, granular cell, keloidal, myxoid, osteoclastic, osteoid, and pigmented AFX.

**Differential diagnosis** may be needed with the following: angiosarcoma spindle cell variant (prominent vascular spaces; positive for vascular biomarkers such as CD31 and ERG); atypical fibrous histiocytoma (→ see dedicated section); leiomyosarcoma pleomorphic type (usually more fascicular growth pattern, desmin positive); pleomorphic dermal sarcoma (though morphologically very similar, more deeply infiltrative lesion with a worse prognosis; complete excision may be needed to correctly classify the neoplasm as superficial biopsy of both tumors may look histologically identical); spindled or desmoplastic melanoma (S100 positive); and squamous cell carcinoma spindle cell type (cytokeratin positive).

## Biomarkers

The staining pattern of AFX is nonspecific, and the diagnosis is largely made by exclusion, also based on the negative staining for markers such as S100, cytokeratins, CD31, ERG, CD34, desmin, and h-caldesmon.

## Prognosis

Overall, AFX has a good prognosis after complete excision: its outcome has been compared to that of cutaneous leiomyosarcoma, while it is much better than that of pleomorphic dermal sarcoma. However, it can recur (in about 10% of cases) and

rarely metastasize. Metastatic cases have been mainly observed in tumors with necrosis, invasion into deep subcutis and beyond, as well as lymphovascular invasion and perineural infiltration: actually these features should cast doubts on the diagnosis of AFX and rather lead to the diagnosis of pleomorphic dermal sarcoma.

## Therapy

Surgery (wide excision) is the treatment of choice.

## Suggested Readings

Chapman (2019) Atypical Fibroxanthoma. Semin Cutan Med Surg 38(1):E65–E66
Phan (2019) Time to recurrence after surgical excision of atypical fibroxanthoma-updated systematic review and meta-analysis. Australas J Dermatol 60(3):e220–e222
Sandhu (2019) Cutaneous Leiomyosarcoma: A SEER Database Analysis. Dermatol Surg [Epub ahead of print]
Soleeymani (2019) Atypical Fibroxanthoma and Pleomorphic Dermal Sarcoma: Updates on Classification and Management. Dermatol Clin 37(3):253–259
Kohlmeyer (2017) Cutaneous sarcomas. J Dtsch Dermatol Ges 15(6):630–648
Mentzel (2017) Atypical Fibroxanthoma Revisited. Surg Pathol Clin 10(2):319–335
Fletcher (2013). WHO classification of tumours of soft tissue and bone (4th edition)