Atypical fibroxanthoma

By Davis C. Diamond, MD, Silas M. Money, MD, and Matthew D. Belcher, MD, FAAD, FACMS

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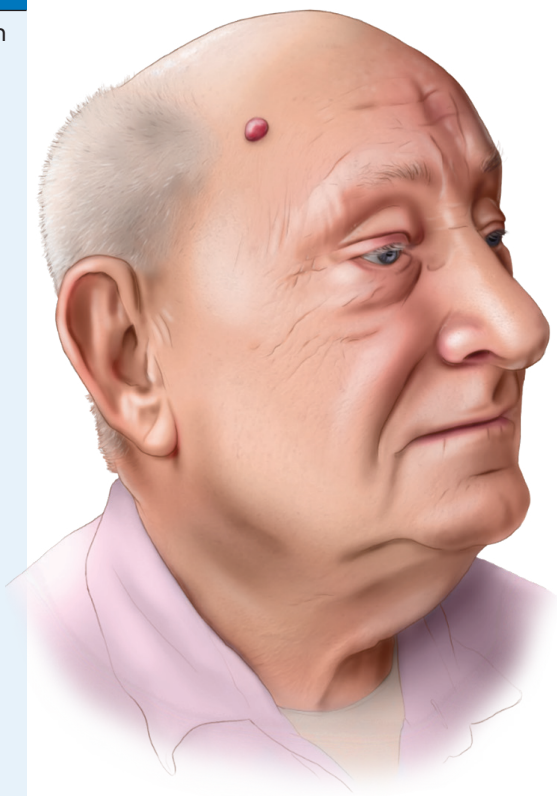
Atypical fibroxanthoma	Population	Location
• Benign dermal-based tumor of uncertain lineage	• Elderly: 70-80 yo • M>F	• Head and neck (most common) • Upper trunk/extremities

Clinical features

Rapidly growing, often ulcerated, dome-shaped red-pink nodule or plaque

Risk factors:
- Cumulative UV exposure
- Advanced age

Differential diagnosis:
- Basal cell carcinoma
- Squamous cell carcinoma
- Amelanotic melanoma
- Merkel cell carcinoma
- Lymphoproliferative disease
- Cutaneous metastasis
- Pleomorphic dermal sarcoma



Histologic features

Proliferation of atypical, monomorphic spindle cells arranged in fascicles, "slamming" up against an ulcerated or atrophic epidermis. Extends down to deep dermis without extensive subcutaneous fat invasion.

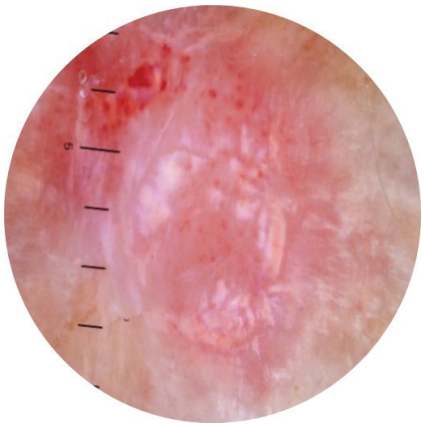
Non-specific staining with: CD10, Procollagen I, SMA (tram-track pattern)

AFX is a diagnosis of exclusion! Other spindle cell neoplasms abutting the epidermis (**SLAM Ddx**) must be ruled out:

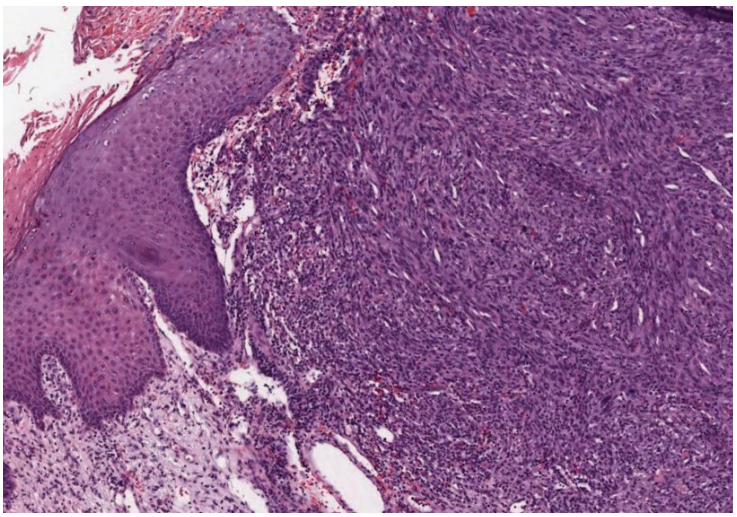
Spindle cell SCCa: CK903, CK5/6, p63, p40
Leiomyosarcoma: Desmin, SMA (diffuse cytoplasmic)
Angiosarcoma: CD31, CD34
Melanoma: S100, Sox-10

Dermoscopic features

Red and white structureless areas with irregular polymorphous vessels



AFX histology



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Management
Primary: • Mohs micrographic surgery (preferred): 3-5% recurrence rate • Wide local excision (1-2 cm margins): 8-10% recurrence rate Recurrent: • Mohs micrographic surgery Postoperative radiation should be considered in all cases where excision with clear surgical margins is not possible

An essential diagnostic distinction: AFX vs. PDS vs. UPS

	Clinical	Pathology	Management/Prognosis
Atypical fibroxanthoma (AFX)	Rapidly enlarging, exophytic, often ulcerated nodules, **typically measuring <2 cm** on head and neck.	Proliferation of atypical, monomorphic spindle cells arranged in fascicles, **slamming** up against an ulcerated or atrophic epidermis. Extends down to deep dermis without extensive fat invasion.	**Mgmt:** Mohs > Excision **Prognosis:** Recurrence: <10% Metastasis: rare
Pleomorphic dermal sarcoma (PDS)	Rapidly growing, large **(median 2.5 cm)**, ulcerated nodules and rarely plaques.	Similar to AFX, but with **deep subcutaneous invasion**, necrosis, lymphovascular or perineural invasion.	**Mgmt:** Excision/Mohs +/- imaging **Prognosis:** Recurrence: 25-30% Metastasis: 5-10%
Undifferentiated pleomorphic sarcoma (UPS) *AKA malignant fibrous histiocytoma*	Similar to PDS, but more likely to be **larger**, deep-seated, ulcerated and on **lower extremity**.	Similar to AFX, but arising in **deep soft tissues** of lower extremity	**Mgmt:** Excision + imaging +/- chemotherapy and/or radiation **Prognosis:** Recurrence: 30-50% Metastasis: 15-40%

** AFX, PDS, and UPS are considered a spectrum by some sources and distinct entities by others*

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