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

### Atypical fibroxanthoma
- **Population**
  - Elderly: 70-80 yo
  - M>F

### Location
- 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

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<th>Clinical</th>
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| Atypical fibroxanthoma  | Rapidly enlarging, exophytic, often ulcerated nodules, **typically measuring <2 cm** on head and neck. | Proliferation of atypical, monomorphic spindle cells arranged in fascicles, **slaming** 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) | 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

### Acknowledgements:

Special thanks to Dr. Harold Rabinovitz, professor of dermatology at the Medical College of Georgia at Augusta University, and Dr. Matt Powell, assistant professor of pathology at the Medical College of Georgia at Augusta University, for providing original dermoscopic and histologic images, respectively. Medical illustration by Alicia Berry, Augusta University.

### References: