Soft Tissue Tumors of the Skin

Fibrohistiocytic Tumors: Histopathology 2010, 56:148-165

2014-03-18
Clinically important variants and differential diagnoses

DERMATOFIBROMA VARIANTS
# Variants of Dermatofibroma

<table>
<thead>
<tr>
<th>Dermatofibroma Variant</th>
<th>Differential Diagnosis</th>
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<tbody>
<tr>
<td>Deep penetrating DF</td>
<td>DFSP</td>
</tr>
<tr>
<td>Atrophic DF</td>
<td>Atrophic DFSP</td>
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<tr>
<td>Anerysmal DF</td>
<td>Kaposi's Sarcoma</td>
</tr>
<tr>
<td>Lipidised (ankle type) DF</td>
<td>Xanthoma</td>
</tr>
<tr>
<td>Palisading cutaneous DF</td>
<td>Schwannoma</td>
</tr>
<tr>
<td>Clear cell DF</td>
<td>RCC metastasis</td>
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<tr>
<td>Granular cell DF</td>
<td>Granular cell tumor</td>
</tr>
<tr>
<td>Myofibroblastic DF</td>
<td>Myofibroblastoma</td>
</tr>
<tr>
<td>Atypical pseudosarcomatous DF</td>
<td>AFX</td>
</tr>
<tr>
<td>Myxoid DF</td>
<td>Cutaneous myxoma</td>
</tr>
<tr>
<td>Epitheloid cell histiocytoma</td>
<td>Spitz nevus</td>
</tr>
<tr>
<td>Cellular DF</td>
<td>Leiomyosarcoma, DFSP</td>
</tr>
<tr>
<td>Smooth muscle proliferation in DF</td>
<td>Infantile myofibroma(tosis)</td>
</tr>
<tr>
<td>DF of the face</td>
<td>AFX, DFSP, leiomyosarcoma, low grade myofibroblast sarcoma</td>
</tr>
<tr>
<td>Metastasizing DF</td>
<td></td>
</tr>
</tbody>
</table>

**Clinically important variants**

*Histopathology 2000; 36:529-539*

*Histopathology 2010; 56:148-165*
Cellular Dermatofibroma

**Characteristics**
5% of dermatofibromas
Infiltration of superficial subcutis
Cellular spindle cell fascicles
Increased proliferative activity
Central tumor necrosis
More frequently SMA+
Peripheral expression of CD34 and Desmin DD: DFSP

Up to 26% local recurrence

→ Complete excision

*Histopathology 2010; 56:148-165*
Aneurysmal Dermatofibroma

Characteristics
<2% of dermatofibromas
Predilection for lower limbs
Rapid recent growth due to hemorrhage
Large and cellular
Infiltration of deep soft tissues
Blood filled spaces without endothelial lining

19% local recurrence
(usually less than 2% in conventional FH)

→ Complete excision

Histopathology 2010; 56:148-165
Atypical Dermatofibroma

**Characteristics**
- Background of common DF
- Spindled fibroblast/myofibroblast-like cells
- Pleomorphic histiocyte-like cells
- Multinucleated giant cells
- 3 mitoses/10 HPF on average
- Occasional atypical mitoses
- Necrosis and deep infiltration possible

- 14% local recurrence
- Rarely distant metastases

→ Complete excision

Histopathology 2010; 56:148-165
Dermatofibromas of the Face

**Characteristics**

- Very rare localisation
- Rarely suspected by the clinician
- Ill-defined
- Infiltration of deep structures
- More aggressive subtypes
- SMA+ spindle cell fascicles

High rate of recurrence (18.5%)
(usually less than 2% elsewhere)

→ Complete/wide excision

*Am J Dermatopathol 2001; 23:419-426*
Dermatofibrosarcoma Protuberans

**DFSP**
1% of all sarcomas
Low grade, locally aggressive fibroblastic neoplasm
CD34+ S100- Desmin- CK-COL1A1-PDGFB fusion gene

**Important variants**
Myxoid DFSP
Superficial plaque-like
Fibrosarcomatous

*Pathology 2014; 46(2):149-159*
Fibrosarcomatous DFSP

**Characteristics**
- 10-20% of DFSPs
- Metastases ↑
- Abrupt or gradual transformation
- Expansive growth
- Cellular fascicles
- Increased atypia
- Increased proliferation
- Loss of CD34 expression
- Increased p53 expression

Imatinib mesylate as therapeutic option

*J Clin Oncol* 2005;23:866-873

*Pathology* 2014; 46(2):149-159

![Ki-67](image1)

![CD34](image2)
SPINDLE CELL TUMORS IN SUN-DAMAGED SKIN

AFX, pleomorphic dermal sarcoma and DD
Atypical Fibroxanthoma (AFX)

<table>
<thead>
<tr>
<th>Gender</th>
<th>59 male, 7 female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>55-95y, mean 77y</td>
</tr>
<tr>
<td>Site</td>
<td>64 head, 1 neck, 1 forearm</td>
</tr>
<tr>
<td>Location</td>
<td>dermis 38, subcutis expansile 24, subcutis focally infiltrative 4</td>
</tr>
<tr>
<td>Histology</td>
<td>mixed 40, spindle 22, epitheloid 4</td>
</tr>
<tr>
<td>Additional features</td>
<td>hemorrhagic/ pseudoangiomaticous 16, granular cell change 15, keloid-like areas 6, myxoid degeneration 5, osteoclast-like giant cells 4, clear cell change 3</td>
</tr>
</tbody>
</table>

### Atypical Fibroxanthomma (AFX)

<table>
<thead>
<tr>
<th>IHC in spindle cell tumors in sun-damaged skin</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>low- and high mw cytokeratins</strong>&lt;br&gt;pancytokeratin, p63</td>
<td>sarcomatoid squamous cell carcinoma (SCC: may be negative!), melanoma (rare), epitheloid angiosarcoma (50%)</td>
</tr>
<tr>
<td><strong>EMA</strong></td>
<td>SCC &amp; AFX (24%), angiosarcoma (rare) focally positive</td>
</tr>
<tr>
<td><strong>S100</strong></td>
<td>spindle cell melanoma, dispersed dendritic cells in AFX</td>
</tr>
<tr>
<td><strong>HMB45, Melan A</strong></td>
<td>melanoma (more specific but often negative in spindle cell melanoma), AFX (clear cell variant), epitheloid angiosarcoma (rare)</td>
</tr>
<tr>
<td><strong>SMA</strong></td>
<td>leiomyosarcoma (diffuse), AFX (focal myofibroblastic diff. 45%)</td>
</tr>
<tr>
<td><strong>Desmin</strong></td>
<td>leiomyosarcoma</td>
</tr>
<tr>
<td><strong>CD31, Fli-1, F. VIII, ERG</strong></td>
<td>angiosarcoma, epitheloid AFX (focal cytoplasmic CD31, Fli-1)</td>
</tr>
<tr>
<td><strong>CD10, CD99</strong></td>
<td>AFX (but non-specific and non contributory)</td>
</tr>
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</table>

Atypical Fibroxanthomma (AFX)

**Diagnosis of exclusion**

No confirmative IHC
(CD68, CD10, CD99 not helpful!)

**Minimal Panel:**
negative reaction: CK22, CK5/6, ERG, Desmin
S100 (dendritic cells+)

Additional markers depending on morphology
AFX or Pleomorphic Dermal Sarcoma?

AFX versus pleomorphic dermal sarcoma? Superficial biopsy
Atypical Fibroxanthoma (AFX)

AFX versus pleomorphic dermal Sarcoma?

Do not answer this question in a superficial biopsy! Ask for complete excision!

Atypical Fibroxanthoma (AFX):
- Growth into superficial subcutaneous tissue with expansile and well demarcated deepest margin allowed
- Growth around the nerve, but without infiltration of the perineural space

Diagnosis of exclusion

→ Complete local excision is curative
Small risk for local recurrence
No metastases
No Mortality

J Cutan Pathol 2010; 37(3):301-309. Morphological and immunohistochemical characteristics of atypical fibroxanthoma...
Pleomorphic Dermal Sarcoma:

**Invasion of deep subcutis/muscle/fascia > 2cm**

Tumor necrosis 53%
L1 26%, Pn1 29%
→
**Risk for local recurrence**
**Metastasis**
**Rare Mortality**