Dermal tumors

Dermatofibroma

Dermal-based proliferation of typically bland, **spindled to histiocytoid**-appearing cells—can appear like a **blue haze**

Tumors are grossly circumscribed but microscopically have irregular, often jagged borders

**Collagen trapping** at periphery

Overlying epithelial **basilar induction** with hyperpigmentation (may mimic BCC)

Evidence supports both neoplastic & reactive etiologies

Many variants: Epithelioid, Cellular, with “monster cells” etc...

**Stains:** FXIIIA(+), CD163(+), CD68(+), CD34(-)

Dermatofibrosarcoma Protuberans (DFSP)

Spindle cell tumor

Proliferation of **monomorph**ic spindle-shaped cells with **deep dermal and subcutaneous** involvement

Arrayed in **storiform** or cartwheel patterns

Lesional cells typically lack significant atypia and pleomorphism

Subcutaneous areas typically show honeycombing fat entrapment (“pearls on a string”)

Defined by t(17;22): Rearrangement of COL1A1 with PDGFB

If loses storiform pattern → herringbone pattern → consider malignant transformation to **fibrosarcoma**

**Stains:** Strong, diffuse CD34, Factor XIIIa (-)

Neurofibroma

**Benign peripheral nerve sheath tumor** composed of

Schwann cells, fibroblasts, perineurial-like cells, and residual nerve axons within extracellular matrix

**Sporadic** in ~ 90% of cases; others are syndromic in association with **NF1**

**Loosely arranged spindle cells in haphazard arrangement**

Poorly defined cytoplasmic borders/processes—Small, hyperchromatic, **wavy or buckled nuclei**

**Stains:** S100(+) in ~ 50% of total cells (Schwann cells); CD34(+) admixed spindled fibroblasts; Neurofilament protein highlights intratumoral axons
**Fibrous Papule**

Solitary, dome-shaped, **flesh-colored papules** on *nose* or central face

Scattered bland, spindled to stellate, and multinucleated fibroblasts

Dense collagenous stroma

**Ectatic thin-walled blood vessels**

If show enlarged, hyperchromatic-staining nuclei with small nucleoli, scant amounts of eosinophilic cytoplasm → consider **Pleomorphic Fibroma**

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**Fibrous Hamartoma of Infancy**

Benign superficial fibrous lesion occurring during first 2 years of life

3 components in organoid growth pattern

1) Intersecting bands of **mature fibrous tissue**, comprising spindle-shaped myofibroblasts and fibroblasts

2) Nests of **immature round, ovoid, or spindle cells** within loose stroma

3) Interspersed **mature fat**

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**Atypical Fibroxanthoma (AFX)**

Mesenchymal neoplasm showing no specific lineage of differentiation

**Highly atypical and pleomorphic** dermal-based proliferation of spindled to epithelioid-appearing cells

Scattered large, bizarre-appearing multinucleated cells often seen

**Numerous mitoses**, including highly atypical forms, easily found

Subcutaneous invasion, PNI, LVI, and tumor necrosis implies more aggressive behavior, and such cases are typically diagnosed as **pleomorphic dermal sarcoma**

**Stains**: Essential to exclude more specific diagnoses:

Negative for melanocytic markers, cytokeratins (especially HMWCKs), p63, muscle (except for SMA), and vascular markers

Positive for nonspecific markers like CD10, CD68, CD99, and vimentin

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**Additional DX:**

**Fibroepithelial polyp (Acrochordon)** → aka Skin Tag

Fibrovascular core is composed of loose to dense connective tissue devoid of adnexal structures

**Sclerotic Fibroma** → Circumscribed, unencapsulated dermal nodule composed of thickened, hyalinized-appearing collagen bundles in storiform/whorled pattern with prominent clefts
**Rare dermal tumor of uncertain histogenesis** composed of epithelioid cells in multiple **nests divided by fibrous septa**
Epithelioid to spindled cells with **abundant pale eosinophilic cytoplasm** arranged in nests **divided by dense fibrous septa**

**Stains:** Often positive for variety of nonspecific markers, including NKI/C3, NSE, PGP9.5

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**Granular Cell Tumor**
Benign tumor of putative schwannian origin composed of cells with abundant **granular cytoplasm**
Overlying **pseudoepitheliomatous hyperplasia**

**Stains:** PAS-D(+) granules; **Strong, diffuse S100(+)**, SOX10(+), Calretinin, CD68

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**Leiomyoma**
Benign Smooth Muscle Tumors;
Both often painful (esp. pilar)

**Pilar Leiomyoma**
**Ill-defined,** dermal nodule composed of haphazardly arranged smooth muscle bundles/fascicles
Fascicles often **dissect between dermal collagen**

**Angioleiomyoma**
**Well-circumscribed** neoplasm composed of mature smooth muscle cells **arranged around prominent blood vessels**

Numerous mitoses, diffuse/marked atypia, necrosis → **Leiomyosarcoma**

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**Scars**

**Scar**
Dense collagen fibers run parallel to the surface
Small, **perpendicularly oriented vessels**
Loss of adnexal structures

**Keloid**
Dense proliferation of **thickened, hyalinized collagen** bundles in dermis
Decreased vessels compared to conventional and hypertrophic scars
Classically on ear
Benign vascular tumors composed of blood vessels lined by plump to flattened endothelial cells with no atypia

**Hemangiomas**

- **Lobular Capillary Hemangioma** (aka Pyogenic Granuloma)
  - Exophytic with collarette
  - Numerous small capillaries radiating out from larger central vessels; May be ulcerated

- **Cavernous Hemangioma**
  - Non-lobular, poorly demarcated proliferation of large, cystically dilated vessels filled with blood

- **Infantile (Juvenile) Hemangioma**
  - Characterized by onset during infancy, rapid growth, and spontaneous involution
  - Appearance changes over time; Tightly packed small- to medium-sized vessels; Unique immunoprofile of placental vasculature → Glut 1 expression

**Glomus tumor**

- Solid nests of round cells with round, uniform, central nuclei closely associated with variably sized blood vessels
- Most common in distal extremities, particularly nail bed
- Typically small, red, painful nodule

  **Stains:** SMA (+)

**Angiosarcoma**

- Malignant neoplasm showing morphologic and/or immunophenotypic evidence of vascular/endothelial differentiation
- Most often scalp/face in elderly (sun exposed) or breast s/p radiation
- Aggressive tumor treated surgically

- **Infiltrative, poorly circumscribed**
  - Variable vascular formation
  - Often cytologic atypia (hyperchromasia, nuclear pleomorphism) and mitoses

  **Stains:** CD31 (+); CD34 (+); ERG (+); FlI-1 (+); Epithelioid angiosarcomas may be CK (+)!

**Additional DX:**

- **Kaposi Sarcoma** → Vascular neoplasm caused by HHV8; often AIDS-associated; Jagged interconnected vascular channels in reticular dermis; grows into normal vessels (promontory sign)

- **Papillary Endothelial Hyperplasia (Mason’s Tumor)** → Reactive intravascular endothelial proliferation; circumscribed, intravascular; Fibrin thrombus with associated papillary structures lined by endothelial cells in single layer; may form anastomosing network;