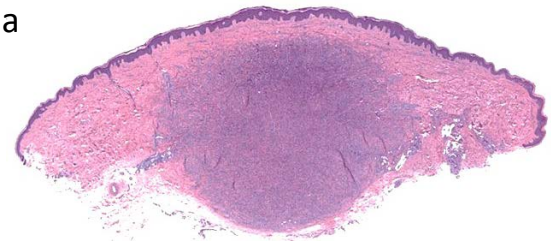
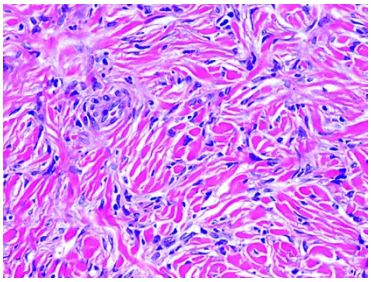
Prepared by Kurt Schaberg

Dermal tumors

Dermatofibroma

aka Benign Fibrous Histiocytoma

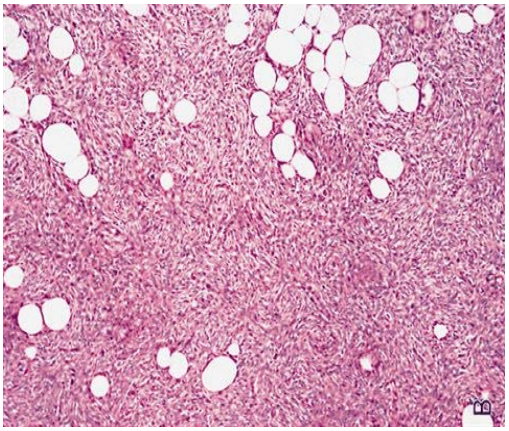




Dermal-based proliferation of typically bland, **spindled to histiocytoid-**appearing cells—can appear like a **blue haze**
Tumors are grossly circumscribed but microscopically have irregular, often jagged borders
Collagen trapping at periphery
Overlying epithelial **basilar induction** with hyperpigmentation (may mimic BCC)
Evidence supports both neoplastic & reactive etiologies
Many variants: Epithelioid, Cellular, with "monster cells" etc…

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

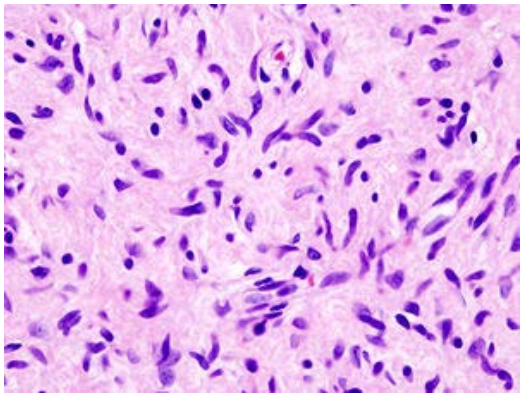
Dermatofibrosarcoma Protuberans (DFSP)



Spindle cell tumor Proliferation of **monomorphic** 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

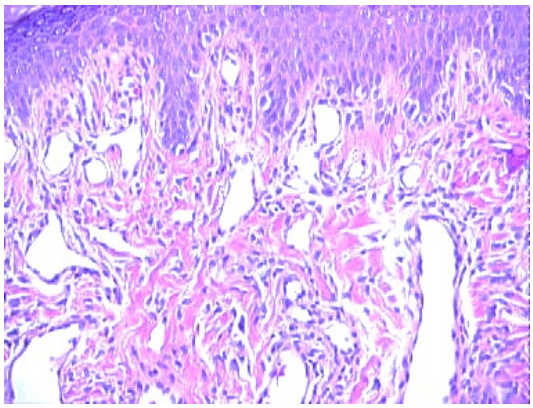


Benign peripheral nerve sheath tumor composed of Schwann cells, fibroblasts, perineurial-like cells, and residual nerve axons within extracellular matrix
Sporadic in ~ 90% of cases; others are syndromic in association with **NF1**
Loosely arranged spindle cells in haphazard arrangement
Poorly defined 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

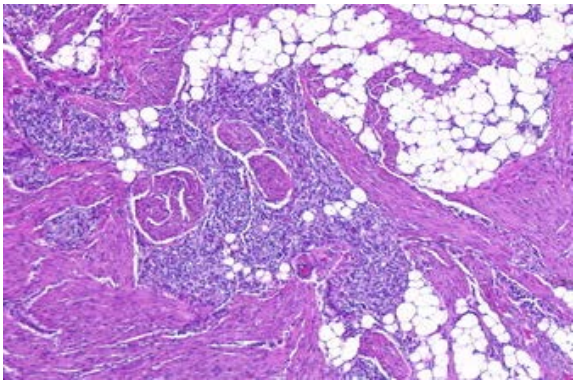
Type of Angiofibroma



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

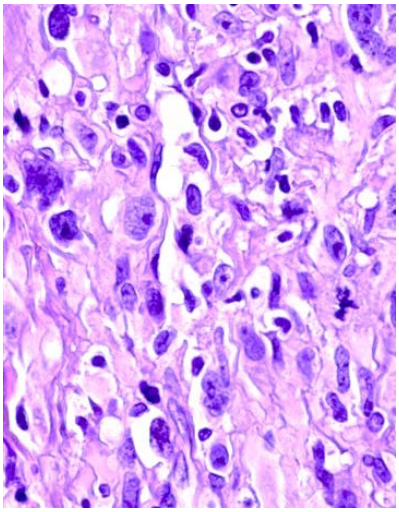
If show enlarged, hyperchromatic-staining nuclei with small nucleoli, scant amounts of eosinophilic cytoplasm → consider **Pleomorphic Fibroma**

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**

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**

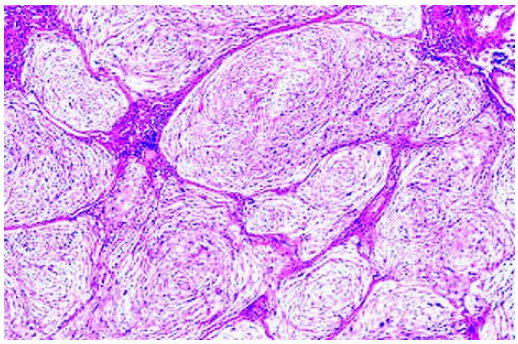
Stains: Essential to exclude more specific diagnoses:
Negative for melanocytic markers, cytokeratins (especially HMWCKs), p63, muscle (except for SMA), and vascular markers
Positive for nonspecific markers like CD10, CD68, CD99, and vimentin

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

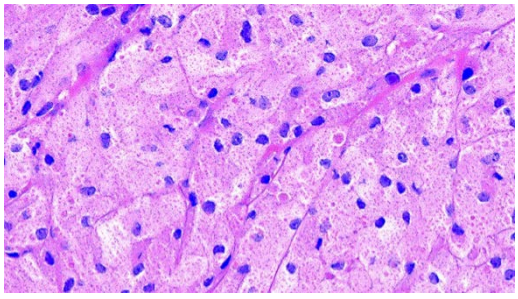
Neurothekeoma



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

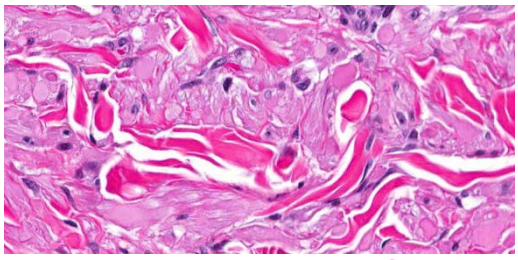
Granular Cell Tumor



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

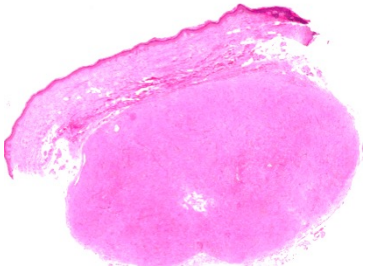
Leiomyoma

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



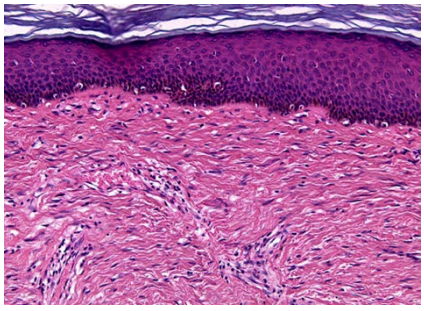
Pilar Leiomyoma
Ill-defined, dermal nodule composed of haphazardly arranged smooth muscle bundles/fascicles
Fascicles often **dissect between dermal collagen**

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

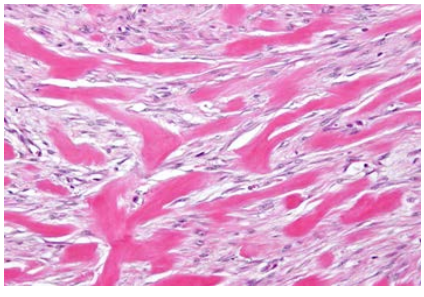


Numerous mitoses, diffuse/marked atypia, necrosis → **Leiomyosarcoma**

Scars



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

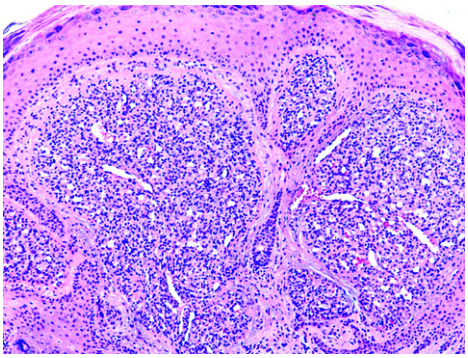


Keloid
Dense proliferation **of thickened, hyalinized collagen** bundles in dermis
Decreased vessels compared to conventional and hypertrophic scars
Classically on ear

Hemangiomas

Benign vascular tumors composed of blood vessels lined by plump to flattened endothelial cells with no atypia

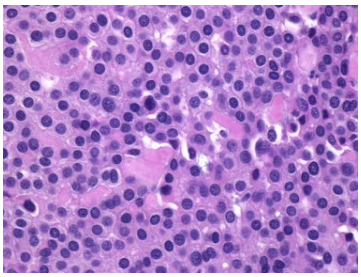
Lobular Capillary Hemangioma (aka Pyogenic Granuloma)
Exophytic with collarette
Numerous small capillaries radiating out from larger central vessels; May be ulcerated



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**

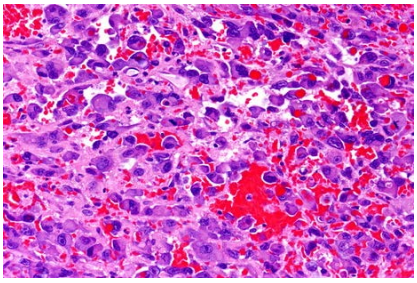
Glomus tumor



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;