Fibrous and Fibrohistiocyctic Tumors of the Skin

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Fibrohistiocytic Tumors

- Fibroblasts
- Myofibroblasts
- Dermal dendrocytes
- Histiocytes
- Collagen fibers
- Elastic fibers
- Mucin (hyaluronic acid)
Fibrohistiocytic Proliferations

**Immunohistochemistry:**

- Vimentin
- Factor XIII a
- CD34
- Histiocytic markers
  - KP-1 (CD68)
  - CD10
  - Alpha-1-antichymotrypsin
- Muscle specific actin (HHF 35)
Soft Fibroma
Acrochordon, fibroepithelial polyp, skin tag

CLINICAL PRESENTATION
• Young adults to elderly patients
• Neck, axilla, and groin
• Few mm in diameter
• Skin color soft polypoid lesion
Soft Fibroma
Acrochodon, fibroepithelial polyp, skin tag

**HISTOPATHOLOGY**
- Polypoid pattern
- Mammillated epidermal hyperplasia
- Core of loose connective tissue
- Sometimes increased small blood vessels or mature adipose tissue
- Absence of adnexal structures
Angiofibroma

• Solitary
  – Fibrous papule

• Multiple
  – Tuberous sclerosis
  – Multiple endocrine neoplasia type I
• Dome shaped dermal nodule
• Proliferation of fibroblasts
• Increased small blood vessels

• Fibroblasts with vesicular nuclei, some binucleated
Dermatofibroma

- Second most common fibrohistiocytic tumor of the skin
- Synonyms
  - Benign fibrous histiocytoma
  - Nodular subepidermal fibrosis
  - Histiocytoma
• Dermal nodule
• Epidermal hyperplasia
• Small follicular or sebaceous structures
Proliferation of fibroblasts arranged as intersecting fascicles
Fibroblasts with plump oval nuclei and small nucleoli
Mono- or multinucleated histiocytes with vacuolated cytoplasm with hemosiderin deposits

Thick hyalinized “keloidal” collagen bundles at the periphery
Immunohistochemistry

- Factor XIIIa (+)
- Histiocytic markers (+)
- CD34 (-)
Variants of Dermatofibroma

• DF with atypical cells (pseudosarcomatous DF, DF with “monster” cells)
• DF extending into the subcutaneous tissue
Dermatofibroma with atypical cells

Pseudosarcomatous dermatofibroma
Dermatofibroma with “monster” cells
DERMATOFIBROMA EXTENDING INTO THE SUBCUTANEOUS TISSUE

Clinical Features:
• Young adults
• Back and proximal extremities; sometimes head and neck
• Deep nodule 1 to 2 cm in diameter
• May appear clinically attached to the subcutaneous tissue
There are two main patterns of extension into the subcutaneous tissue:

In 72% of cases, vertical or radial fashion, predominantly along the septae, which have a wedge-shaped appearance.

In 28% of cases, well circumscribed deep rounded margin with a pushing border.

[Images of radial and rounded patterns]
DERMATOMYOFIBROMA

Clinical Presentation

- Most commonly in young adults
  some cases in children
- Predominantly in women
- Around the shoulder area, axilla, upper arm, and neck
- Well-circumscribed plaque measuring 1 to 2 cm in diameter
- Tan to erythematous color
- No recurrence after simple excision
DERMATOMYOFIBROMA

Histopathologic Findings

- Well-circumscribed plaque involving the reticular dermis and the upper portion of the subcutaneous septae
- Well-defined, elongated, intersecting fascicles parallel to the skin surface
- Adnexal structures are mostly preserved
• Spindle-shaped cells with a very uniform appearance separated by thin collagen fibers and elastic fibers
• Elongated nuclei with rounded or pointed ends and one or two small nucleoli
• No evidence of nuclear atypia
• Mitotic figures are rare
Immunohistochemistry

• Vimentin (+)
• Muscle specific actin (+)
• Smooth muscle actin (-/+)
• Desmin (-)
• CD34 (-)
Sclerotic Fibroma of the Skin
Clinical Features

- Multiple in Cowden’s disease (multiple hamartoma syndrome)
- Skin and/or mucous membranes
- Pearly papules or nodules
- Less than 1 cm in diameter
- Solitary, sporadic
Histopathologic Findings

- Well-circumscribed dermal nodule
- Hypocellular lesion
- Sclerotic thick collagen bundles
- Hyalinized collagen bundles in a "plywood-like" pattern
Differential Diagnosis

• Pleomorphic fibroma
• Keloid
Epithelioid Cell Histiocytoma

Clinical presentation:

- Adults in the fifth decade of life
- Slight predominance in women
- Extremities, the thigh is the most frequent location
- Firm, sessile or polypoid papules or nodules
- 0.5 to 1.5 cm
- No evidence of recurrence after excision
Epithelioid Cell Histiocytoma
Histopathology
Xanthogranuloma

- Juvenile
- Adult
Clinical Presentation

- 20% at birth
- Predominantly in infants in the first year of life
- Seen also in adults
- Single or multiple
- Red to yellow papules or nodules
- 0.5 to 1.0 cm
Xanthogranuloma

Systemic Manifestations:

- Usually in children
- Ocular most common
  - Glaucoma
  - Bleeding into the anterior chamber
- Oral
- CNS, Lung, Liver
Multinucleated histiocytes with nuclei in Christmas wreath arrangement (Touton cells)

Mono and multinucleated histiocytes with vacuolated cytoplasm
Lymphocytes
Eosinophils
Two variants:

- Giant cell reticulohistiocytoma
- Multicentric reticulohistiocytosis
• Giant cell reticulohistocytoma
  – usually single nodule
  – adults
  – head and neck
• **Multicentric reticulohistiocytosis**
  - adults (5th to 6th decade)
  - female
  - disseminated nodules
  - extremities commonest site
  - face, mucosa
  - polyarthritis may be destructive and mutilating
  - xanthelasma in one fourth of cases
  - hyperlipidemia, internal malignancy, autoimmune disorders
Histiocytes with abundant eosinophilic “ground glass” cytoplasm
Multinucleated histiocytes
IMMUNOHISTOCHEMISTRY

- KP-1 (+)
- Xllla (+)
- S-100 (-) occasional (+)
- CD1a (-)
GIANT CELL TUMOR OF TENDON SHEATH

CLINICAL PRESENTATION

Firm nodule on the hands, fingers, toes and other periarticular sites
- Slow-growing and fixed to subcutaneous structures
- Usually asymptomatic, but pain, numbness, or stiffness can occur
- Usually in adults between the ages of thirty to fifty
- More common in females than in males
Dermal and subcutaneous nodule
Multinucleated giant cells
Histiocytes with vacuolated cytoplasm
Hemosiderin deposits
Giant multinucleated “osteoclast”-like cells
Nodular Fasciitis

- Pseudosarcomatous fasciitis
Clinical Presentation

- Middle age
- Upper extremity most common site in adults
- Head and neck in children
- Rapidly growing, self limited
- 1 to 5 cm
- Subcutaneous, fascial or intramuscular nodule
- May be tender
Subcutaneous nodule with stellated appearance
Proliferation of fibroblasts and small vessels
Edematous stroma with mucin deposits
Spindle and stellated fibroblasts
Mitoses
Extravasated erythrocytes
Fibromatoses

• Fascial
• Deep musculoaponeurotic
Fascial

- Palmar fibromatosis (Dupuytren’s disease)
- Plantar fibromatosis (Ledderhose’s disease)
- Penile fibromatosis (Peyronie’s disease)
- Knuckle pads
CLINICAL PRESENTATION

- Slowly growing
- May measure up to several cm in greatest diameter
- Firm nodules or cord-like tumors
- After pregnancy in abdominal wall
- Abdominal scars
- Gardner's syndrome
- Trauma maybe a trigger
Fibromatosis

Large dermal and subcutaneous poorly circumscribed lesion
• Fascicles of monomorphous spindle shaped fibroblasts and myofibroblasts

• Elongated nuclei with fine chromatin

• Rare mitoses

• Collagen fibers between the cells
Dermatofibrosarcoma Protuberans
Clinical Presentation

- Young adults
- Trunk or proximal extremities
- Indurated plaques and nodules, measuring one to several centimeters
- Slowly growing
• Flat to elevated surface
• Plaque and nodular areas in the dermis and subcutaneous tissue
Slender to plump spindle-shaped cells
Intersecting fascicles in a "storiform" pattern
"Honeycomb" infiltration in the subcutaneous tissue
Atypical cells between fat cells
Dermatofibrosarcoma Protuberans
Immunohistochemistry: CD 34(+) Factor XIIIa (-)
Atypical Fibroxanthomma
Clinical Features

- Elderly patients
- Head and neck
- Sun-exposed skin
- Measures from 1 to 2 cm in diameter
- Rapid growth
- Low grade sarcoma that usually is cured by conservative excision
Dome shaped nodule covered by a thin epidermis
• Atypical spindle-shaped cells and multi-nucleated histiocyte-like cells with large, pleomorphic, and hyperchromatic nuclei and pale staining vacuolated cytoplasm
• Numerous typical and atypical mitotic figures
Differential Diagnosis

- Spindle cell squamous-cell carcinoma
- Spindle cell malignant melanoma
Immunohistochemistry

- Vimentin (+)
- Alpha-1-antichymotrypsin (+)
- CD 68 (+)
- CD 10 (+)
- Muscle specific actin (+) (spindle-shaped cells)
- Cytokeratins (-)
- S-100 (-)
FIBROSARCOMA
Clinical Features

• 4\textsuperscript{th} and 5\textsuperscript{th} decades of life
• Most commonly located on the lower extremities
• Predominantly deep soft tissues
• May involve the overlying dermis
• Solitary tumor 3 to 10 cm in greatest diameter
More Clinical Features:

- Usually diagnosed when it becomes a palpable painful nodule
- May appear in old burn scars
- Areas of chronic radiation dermatitis
- Tends to recur
- Hematogenous metastases
Fibrosarcoma

- Usually the dermis and subcutaneous tissue are secondarily involved from a fibrosarcoma of the underlying soft tissues
- Monomorphous atypical spindle-shaped cells arranged as intersecting fascicles in a "herring-bone" pattern
- Thin collagen bundles between the cells
- Cells have scanty cytoplasm and elongated hyperchromatic nuclei
Differential Diagnosis

• Nodular fasciitis
• Fibromatosis
• Dermatofibrosarcoma protuberans
• Atypical fibroxanthoma (malignant fibrous histiocytoma)
• Malignant peripheral nerve sheath tumor