Selected Spindle Cell Tumors

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STORIFORM COLLAGENOMA
(Sclerotic Fibroma, Plywood Fibroma)

- Cutaneous & sometimes mucosal papules
- Single or multifocal (with Cowden’s disease)
- Well-circumscribed, unencapsulated thickened and sclerotic collagen bundles
  - whorled and “plywood-like” pattern
  - paucicellular with few spindled fibroblasts and some accumulation of mucin
- May represent a final common endpoint
Sclerotic fibroma-like change in various neoplastic and inflammatory skin lesions: is sclerotic fibroma a distinct entity?

Abstract: Sclerotic fibroma was first described in association with Cowden’s disease by Weary et al. in 1972. In 1989, Rapini and Golitz detailed 11 cases of solitary sclerotic fibroma (SFS) in the absence of Cowden’s disease, suggesting the term SFS of the skin. Classic histological features include hypocellular, hyalinized bands of collagen sharply demarcated from the surrounding skin. Numerous authors have described sclerotic fibroma-like changes in other entities including melanocytic nevi, dermatofibromas, lipomas, tendon sheath fibromas, giant cell collagenomas, neurofibromas, angiofibromas, erythema elevatum diutinum, and folliculitis. Dissension has arisen, with some dermatopathologists asserting that sclerotic fibroma is just an evolutionary end-point of a previous lesion. Others contend that SFS is a distinct lesion and cite recurrent cases and/or proliferation marker studies to corroborate this view. We detail the histopathological findings of lesions consistent with the classic description of SFS and compare these to sclerotic changes observed in an intradermal nevus, blue nevus, erythema elevatum diutinum, neurofollicular hamartoma, angiofibroma, neurofibroma, accessory nipple, and dermatofibromas. Sclerotic fibroma-like change may be seen in a variety of lesions and may represent a common reaction pattern in the skin.

High WA, Stewart D, Essary LR, Kageyama NP, Hoang MP, Cockerell CJ. Sclerotic fibroma-like change in various neoplastic and inflammatory skin lesions: is sclerotic fibroma a distinct entity?

FIGURE 1 – Comparison of a classic sclerotic fibroma of the skin at low (A) and medium power (B) with a dermatofibroma with prominent central sclerotic change at low (C) and medium power (D).
DERMATOMYOFIBROMA

- Composed of fibroblasts/myofibroblasts
- Most common in adolescents & young adults, predilection for women
- Location: upper back, shoulder, axilla, upper arms, abdomen
- Primary lesion: asymptomatic, firm, skin-colored, tan/yellow plaque/nodule
DERMATOMYOFIBROMA

• Low Power:
  – well-circumscribed proliferation of spindle cells in reticular dermis, may involve fat
  – intersecting fascicles, resemble smooth muscle, oriented parallel to the epidermis

• High Power:
  – spindle cells with uniform nuclei, rounded/pointed ends but may be wavy
  – Embedded in a fine, collagenous stroma

• Immunohistochemistry
  – POSITIVE = vimentin, MSA, SMA (+/-)
  – NEGATIVE = desmin, factor XIIIa, S100
DESMOID TUMOR

• Extraabdominal desmoid
  – Age - 10-40 years
  – Location - shoulder, chest wall, back, thigh
  – Deep-seated, firm, poorly defined mass

• Abdominal desmoid
  – Typically adult women
  – Location- fascial coverings of rectus and internal abdominal oblique muscles
DESMOID TUMOR

- Low power
  - poorly circumscribed tumor, ill defined fascicles
  - infiltrative growth pattern, often into muscle
  - necrotic muscle fibers common
- Cells - uniform, wavy, pointed
- Stroma
  - delicate, rarely sclerotic
  - mucoid stroma - focal, rare
  - chondroid or osseous metaplasia is rare
Wavy Nuclei with Pointed Tips

Collagen Thin Tibers
Necrotic Striated Muscle
INFANTILE DIGITAL FIBROMA

- Neonates & infants, rarely children and adolescents
- Skin - colored or erythematous nodule
- Location - digits of hands, occasionally feet
- May recur locally after excision
- Regress spontaneously over years
FIBROUS HAMARTOMA OF INFANCY

- Poorly circumscribed tumor in subcutis
- TRIPHASIC - three different components:
  - well-defined interlacing fascicles with abundant collagen
  - foci of immature ovoid cells with myxoid stroma
  - interspersed mature fat
- Less commonly fibrosis resembling neurofibroma
Fibrous Hamartoma of Infancy
MYOFIBROMA

• Congenital variant (at birth or in early life)
  – 50% solitary, 50% multiple
  – usually SQ nodules
  – spontaneous regression (for pt with few lesions)
• Adult variant
  – single or rarely multiple
  – non-descript nodule, resembles DF or scar
• BIPHASIC histological pattern
  – spindled fascicles that resemble smooth muscle
  – hemangiopericytoma-like areas (vascular spaces)
NODULAR FASCIITIS

- Patients typically young adults
- Primary lesion – usually solitary, subcutaneous nodule, 1-3 cm
- Usually a history of rapid growth
- Most common locations
  - Adults - forearm
  - Children - head and neck (cranial fasciitis)
Histology

- Mass in the fibrous septa
- Cell arranged in fascicles
- Mucin/myxoid
- Chronic inflammation
- Extravasated RBCs
- Rare MNCs
- “Fibroblasts in tissue culture”
DERMATOFIBROMA

- Ubiquitous fibrohistiocytic tumor
- Typically arises in adults
- Women > men
- Most common location – legs > arms > trunk
- Primary lesion - firm nodule, exophytic or endophytic, positive dimple sign
- Eruptive lesions associated with SLE
DERMATOFIBROMA VARIANTS

• Histiocytic (lipidized) variant - ↑ lipid, +/- Touton multinucleated giant cells
• Hemosiderotic variant - hemorrhage, increased hemosiderin
• Epithelioid cell histiocytoma - exophytic with stellate epithelioid cells
• Sclerotic fibroma - distinct tumor versus histologic variant controversy
SOLITARY FIBROUS TUMOR

• Mesenchymal tumor originally described in pleural space
  – numerous extrapleural sites reported
• Rare tumor
• Typically middle-aged adults
• Site - primarily head & neck, extremities
• Slow-growing subcutaneous mass
• Malignant transformation in 10%
SOLITARY FIBROUS TUMOR

• Well-circumscribed, not encapsulated
• “Patternless Pattern Architecture”
  – alternating hypo and hypercellular areas
  – hyalinized collagen and keloidal fibers
  – myxoid changes & fibrosis variably present
• Branching vessels, round to spindle cells
• Variable numbers of plasma cells, mast cells, lipocytes, MNGC, CD34+
DERMATOFIBROSARCOMA PROTUBERANS

- Rare tumor (0.1% of all malignant neoplasms), but #1 soft tissue sarcoma
- Middle-aged persons, trunk and extremities
- Only 40% of cases are protuberant
- Atrophic variants can be particularly vexing
- Dense spindled cell proliferation with generally strong CD34+
- >90% with t(17;22) COL1A/PDGFR translocation
DERMATOFIBROSARCOMA PROTUBERANS
ATYPICAL FIBROXANTHOMA

• Coined at AFIP in 1963
• Heavily sundamaged skin of elderly
• Some low grade malignant behavior observed
• Atypically atypical (“too malignant to be malignant”)
• Immunostains employed
  – CD68, CD10, Procollagen I, P53
Atypical Fibroxanthoma
P53 is also of utility...

- Eroded nodule
- Ear of 89 y/o woman
Atypical spindled cells...

PanCK, P63, 34Beta12 all neg

S100 was similarly negative.
Combination of CD68/P53 has been useful as confirmation of AFX
EPITHELIOID SARCOMA

• Rare soft tissue tumor on extremities of young adults
• Confused clinically and histologically with granuloma annulare
• Aggressive (metastasis in 35-40% cases)
• Regional LN and lungs are most affected
• Disputed histiogenesis – synovial?
Epithelioid Sarcoma
Staining Pattern

• Classic pattern:

  POSITIVE = vimentin, PanCK, EMA

  NEGATIVE = S100, CK20, CK5/6

  Variable = 34Beta12, CK7