Spindle cell lesions: The Good, The Bad, and the Ugly

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Objectives

• Refresh and broaden your knowledge of spindle cell lesions
• Help you to establish a histology based approach for classification of spindle cell tumors
• Learn distinctive features of select spindle cell tumors for accurate identification
So you have a spindle cell lesion...now what?
Additional architectural features

• Fasicular
Additional architectural features

• Storiform
Additional architectural features

• Lobulated/plexiform
Additional architectural features

- Palasaded
Stroma

- Myxoid
Stroma

- Collagenous
Nuclear pleomorphism
Prominent inflammatory cells
Adipocytic component
Prominent blood vessels
Fascicular
Nodular Facitits
(and variants)

- Self limited pseudosarcomatous proliferation of fibroblasts/myofibroblasts
- 20-40 yo
- Usually soliar 2-3 cm subcutaneous nodule, develops rapidly
- Upper extremities, head and neck (kids), trunk
- Tends to develop along fibrous septa
- Earlier lesions more myxoid, later fibrotic
- Irregular fascicles/storiform, mitoses can be numerous, may have lymphoid aggregates and capillaries, sometimes centrally hypocellular
Nodular Fasciitis

• Variants: Intravascular, Proliferative, Ischemic
Nodular Fasciitis

- Strongly diffusely positive for SMA, MSA, Calponin, CD68
- Negative for S-100, CD34, B-catenin, only scattered desmin positive cells
- Molecular: rearrangements of the USP6 locus at 17p13 (>90% of cases); MYH9-USP6 fusion gene detected in 65% of cases
Myofibroma/Myopericytomay

• On a continuum (including even glomangiopericytomay)
• Solitary painless cutaneous/subcutaneous/mucosal nodule, head and neck or lower extremity of adults
• Well circumscribed but unencapsulated multinodular, biphasic tumors
• Primitive cellular proliferation of small short spindle cells with thin walled branching blood vessels and whorled nodules/fascicles of plump spindle cells
  • usually the primitive component in center surrounded by myoid whorls
• may have a pseduochondroid appearance to the myoid nodules or prominent hyalinization/calcification
Myofibroma
Myopericytoma

• Ovoid, plump spindle cells around thick and thin walled blood vessels
• Rarely show nuclear atypia and high mitotic rate indicating malignancy
Myofibroma/Myopericytoma

• Positive for SMA and often h-caldesmon
  • Primitive component less positive than the myoid nodules
• Desmin focally positive in a small subset
• Neagative for S-100, EMA, keratins CD34 (except vessels)
Dermatomyofibroma

- Clinically a discolored plaque, few centimeters in size, shoulder girdle/neck/upper arm of young adult females
- Bland spindle cells in a parallel (to the epidermis) fascicular arrangement
Dermatomyofibroma

• SMA and MSA positive, but desmin negative
• CD34 focally positive in a subset (as opposed to diffuse in DFSP)
Leiomyoma

• Solitary or multiple nodules, often painful
• Most commonly shoulder/upper arm
• SMA, MSA and desmin positive
Atypical intradermal smooth muscle tumor

- Leiomyosarcoma confined to the dermis
- Subcutaneous nodule or plaque,
- Extremities/truck most commonly
- Hyperchromasia, increased
- Mitotic index, +/- necrosis
- Keratins can be focally positive
Malignant peripheral nerve sheath tumor

- Usually relatively deep seated malignant tumors
- Extremities or paraspinal most common
- Usually adults, even in NF1 patients (2-10% lifetime risk)
- Usually uniform spindle cells with hyperchromatic nuclei and often some degree of pleomorphism; mitoses (low grade may not show them) and often necrosis
- Any mitotic figures in an otherwise neurofibroma-like lesion with increased cellularity and nuclear atypia warrants the diagnosis of MPNST in a NF1 patient
Usually relatively hypocellular areas alternating with hypercellular areas showing perivascular accentuation; matrix in less cellular areas often myxoid
Pseudomyogenic Hemangioendothelioma

- Soft tissue neoplasm of intermediate biologic potential
- multiple discreet lesions in different tissue planes
- a vascular neoplasm that histologically closely resembles a myoid tumor
- young adults, male predominant
- Extremities, 1-2 cm in size
- histologically irregular infiltrative margins, sometimes plexiform, loose fascicles and sheets of spindle cell with minimal atypia
- Positive AE1/AE3, ERG, FLI1, retained INI1
- Negative CD34, S-100, Desmin
- SERPINE1-FOSB fusion gene
Pseudomyogenic Hemangioendothelioma

About 50% will have a prominent neutrophilic infiltrate

Sometimes a rhabdomyoblast-like appearance
Solitary circumscribed neuroma

- AKA palisaded encapsulated neuroma
- Relatively common tumor of the central face of adults
- Skin colored papule up to 0.5 cm
- Histologically well circumscribed tumor in the dermis composed of spindle cells with schwannian differentiation and intratumoral axons arranged in short fascicles
- Occasional nuclear palisading
- Rare plexiform growth
- Diffusely positive for S100 and intratumoral axons highlighted by neurofilament protein
- Partial capsule is EMA positive
Solitary circumscribed neuroma
Storiform: Benign Fibrous Histiocytoma

• Common
• multiple variants
• usually adult extremity as well as trunk; solitary well-demarcated firm papule or plaque less than 1cm
• often hyperpigmented or red clinically
• histology is a relatively well-circumscribed dermal based tumor composed of short spindle cells in a haphazard arrangement often storiform; collagen trapping at the periphery; may have foamy histiocytes and multi-nucleated giant cells; some have hemosiderin deposition
• Typically have pigmented epidermal hyperplasia and even follicular induction at the surface
• Factor XIIIa positive, SMA and MSA often positive, CD34 positivity is rare
• S100 and desmin negative
Dermatofibrosarcoma protuberans

• Locally aggressive tumor
• Rare tumor of young adults mainly; trunk and extremities
• Clinically usually large and multinodular, several centimeters in diameter with red or bluish discoloration
• Histologically ill-defined and diffusely infiltrative tumor of the dermis and subcutis composed of uniform spindle cells in a monotonous storiform pattern which percolate through the underlying adipose tissue in a honeycomb-like pattern
• Positive for CD34 and may be SMA positive
• S100 and desmin negative
• Fibrosarcomatous variant is fascicular/herringbone with higher mitotic activity and possible necrosis (which loses it’s CD 34 positivity)