Fibrous lesions of soft tissue

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Heterogenous group

• **Reactive lesions**
  – Keloid scars
  – Hypertrophic scars
  – Nodular fasciitis
  – Proliferative fasciitis
  – Proliferative myositis
  – Fibro-osseous pseudotumour
  – Myositis ossificans
  – Ischaemic fasciitis
  – Reactive myoblastic proliferations
  – Elasto-fibroma

• **Hamartomatous lesions**
  – Fibrous hamartoma of infancy
  – Infantile myofibromatosis
  – Fibromatosis colli
  – Juvenile hyaline fibromatosis
• Benign neoplasms
  – Fibroma of tendon sheath
  – Desmoplastic fibroblastoma
  – Storiform collagenoma
  – Calcifying aponeuritic fibroma
  – Solitary fibrous tumour
  – Cellular angiofibroma
  – Giant cell angiofibroma
  – Angiomyofibroblastoma
  – Dermatomyofibroma
  – Solitary myofibroma
  – Nuchal-type fibroma
  – Gardner fibroma
  – Inclusion body fibromatosis

• Fibromatoses
  – Palmar fibromatosis (Dupuytren contracture)
  – Plantar fibromatosis (Ledderhose disease)
  – Penile fibromatosis (Peyronie disease)
  – Desmoid fibromatosis (deep-seated fibromatosis)
  – Inflammatory myofibroblastic tumour
• Fibroblastic Sarcomas
  – Conventional fibrosarcoma
  – Myxofibrosarcoma
  – Low-grade fibromyxoid sarcoma
  – Sclerosing epithelioid fibrosarcoma
  – Acral myxoinflammatory fibroblastic sarcoma

• Fibro- histiocytic tumours (so called)
  – Localized giant cell tumour of tendon sheath
  – Giant cell tumour of soft tissue
  – Deep benign fibrous histiocytoma
  – Plexiform fibrohistiocytic tumour
  – Malignant fibrous histiocytoma (so-called)
Nodular fasciitis

• Clinical features
  – Most prevalent in young adults – peak 40yrs
  – Most common locations
    • Upper extremities
    • Trunk
    • neck
  – History of rapid growth
  – Small size
  – History of preceding trauma in 10-15%
  – Infiltrative margins
  – Grows within fascia or into muscle and subcutis
Myxoid features

Plump immature fibroblasts

Mitoses

NO ATYPICAL CELLS
• Collagen deposition
• Metaplastic bone deposition

• Morphological variants;
  – Cranial fasciitis
  – Intravascular fasciitis
  – Proliferative fasciitis

→ Local marginal excision is adequate
→ Rarely recurs after excision

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44 year old male – rapidly growing palpable mass in proximal forearm – 1.5 cm

Axial T1 weighted image

Axial T2 (fat-suppressed) weighted image
T1 (fat supp) weighted image

Bland spindle cells
Alternating cellularity
M=myxoid degeneration
Fibromatoses

- Group of conditions with common features
  - **Infiltrative** pattern of growth
  - **Locally aggressive** pattern of behaviour
  - Frequently **recur** after incomplete excision
  - Lack features of cytological features of malignancy
  - **Do not metastasise**

Superficial  Deep (desmoid)
Fibromatoses  fibromatoses
Fibromatose

- Often arise in muscular fascia
- Grow diffusely between skeletal muscle fibres

Proliferation of well-differentiated fibroblasts

Active cellular phase increasingly hyalinised
Superficial fibromatoses

- Affect males more frequently
- Genetically distinct from deep type
- Nodules or fascicles
- Mature myo-fibroblasts
- Dense collagen

>>25% of palmar & plantar stabilise.

>>Some recur after excision esp. plantar.
Duputryen contracture
Duputryen contracture

• More common with increasing age
• More common in men
• Some associated with plantar lesions
• Bilaterality is common
• No correlation with occupation or history of trauma

→ Palmar fasciectomy
35 year old male with small palpable painful mass in medial aspect of plantar arch

Short axis T1 image

Firm glistening nodule in aponeurosis
Deep (desmoid) fibromatoses

- Clinically diverse group of neoplasms
- 3 biologic groups:
  - Sporadic
  - Associated with FAP
  - Multi-centric or familial
- 3 anatomic locations:
  - Extra-abdominal 60%
  - Abdominal wall 25%
  - Intra-abdominal 15%
Deep Fibromatoses

• All form large infiltrative masses

• Recur repeatedly if not excised with clear margins

• No metastatic potential

• Surgery +/- radiotherapy.

• (Oestrogen antagonists, NSAIDs, low-dose chemo)

• ??Imatinib (Gleevec)

• Peak incidence is 2\textsuperscript{nd} to 4\textsuperscript{th} decade

• Female predominance 2:1
45 year old Chinese male – previous abdominal surgery for mesenteric trauma and small bowel infarct and recurrent abscesses. Presents one year later with painful mass around midline scar.
Tumour mass adherent to distal ileum

Distal ileum resected

Adjuvant chemo therapy given

Difficult to achieve complete resection with involvement of local structures

Plump fibroblasts in collagenous stroma
Fibroblastic sarcoma

- Strict diagnostic criteria
- Rare
- 2 groups:
  - Adult fibrosarcoma
  - Infantile fibrosarcoma
- Most well-circumscribed masses, less than 10cm

- Can arise in superficial and deep connective tissue.
- Can grow slowly or rapidly
Fibroblastic sarcoma

**Adult**
- 4th to 6th decade
- Male preponderance
- Common sites are thigh and trunk
- Five year survival < 40%
- Most often deep-seated (subfascial)

**Infantile**
- Develop in first 2 years
- Can be congenital
- Male preponderance
- Arise in extremities – distal
- Five year survival >80%
- Most sub-cutaneous
Conventional fibrosarcoma

- Highly cellular
- Fibroblast-myofibroblast cells
- Classical herringbone pattern
- Minimal collagen
- Mitotic rate variable

- Superficial and well-differentiated have better prognosis – generally..

Treatment of choice is radical surgery

Radiotherapy if residual tumour or positive margins

Adjuvant chemo if high grade
Myxofibrosarcoma

• Most common soft tissue sarcoma of older adults
• Affects adults mainly in in 6th - 8th decades
• Slightly more common in males

• Most arise in limbs – in fascia or muscle
• 2/3 of cases arise in subcutaneous locations

• Multi-nodular and diffusely infiltrative
• Large spectrum of histological grades

• Survival and potential to metastasise correlates with grade
• Overall survival 60-70%
Myxofibrosarcoma

• Hypocellular myxoid areas with thin curved vessels – at least 5-10% of total.

• Difficulty in diagnosis in small biopsies as high-grade can appear solid and epithelioid.
61 year old male with lower extremity mass, slow growing over 1 year + mild discomfort. No hx of trauma
Thank you for your attention

References

4. PathConsult http://www.pathconsultddx.com