

# Myxoid Soft Tissue Tumours

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## WHO classification of soft tissue tumours

- Adipocytic tumours
- Fibroblastic/myofibroblastic tumours
- So-called fibrohistiocytic tumours
- Smooth muscle tumours
- Pericytic (perivascular) tumours
- Skeletal muscle tumours
- Vascular tumours
- Chondro-osseous tumours
- Tumours of uncertain differentiation

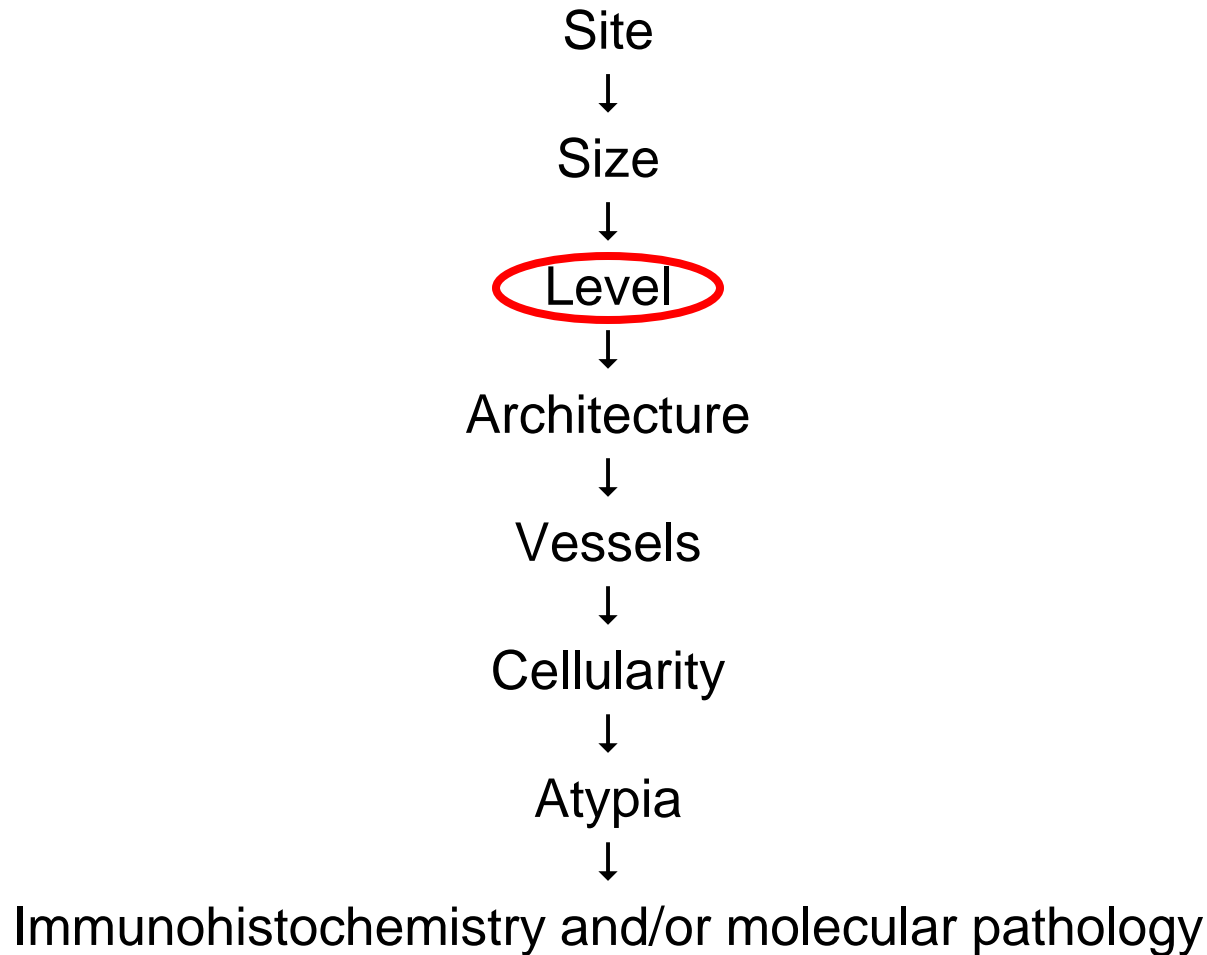
## My approach to soft tissue tumours

- Spindle cell pattern
  - *Nodular fasciitis pattern*
  - *Fibromatosis pattern*
  - *Fibrosarcoma pattern*
  - *Storiform pattern*
  - *Patternless pattern*
- Epithelioid cell pattern
- Small round blue cell pattern
- Pleomorphic cell pattern
- Myxoid pattern

***Al-Nafussi A.***

***Practical morphological approach to diagnosis and differential diagnosis of soft tissue sarcomas.  
Current Diagnostic Pathology 2002, 8, 395-411.***

# My approach to myxoid soft tissue tumours



***Van Roggen J, Hogendoorn P, Fletcher C.  
Myxoid tumours of soft tissue.  
Histopathology 1999, 35, 291–312.***

<b>Dermal</b>				
<b>Sub- cutaneous</b>				
<b>Intra- muscular</b>				

<b>Dermal</b>	<b>Dermal nerve sheath myxoma</b>	<b>Superficial angiomyxoma</b>	<b>Digital myxoma</b>	<b>Digital fibromyxoma</b>
<b>Sub- cutaneous</b>				
<b>Intra- muscular</b>				

## Dermal nerve sheath myxoma (myxoid neurothekeoma)

- **Site:** *Anywhere*
- **Size:** *<3 cm*
- **Level:** *Dermis*
- **Architecture:** *Lobulated*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Hypocellular*
- **Cytology:** *Non-atypical*
- **IHC:** *S100 +ve*

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- **IHC:** *S100 +ve*

### *Myxoid neurothekeoma*

- *Spindle cell + myxoid*
  - *Schwannian*
  - *S100 +ve*

### *Cellular neurothekeoma*

- *Epithelioid cell + non-myxoid/myxoid*
  - *Fibroblastic/fibrohistiocytic*
  - *CD63 (NKI/C3) +ve*

## Superficial angiomyxoma (cutaneous myxoma)

- **Site:** *Anywhere*
- **Size:** *<4 cm*
- **Level:** *Dermis/subcutis*
- **Architecture:** *Lobulated*
- **Vessels:** *Round*
- **Cellularity:** *Hypocellular*
- **Cytology:** *Non-atypical*
- **IHC:** *CD34 +ve*

## Superficial angiomyxoma (cutaneous myxoma)

- **Site:** *Anywhere*
- **Size:** *<4 cm*
- **Level:** *Dermis/subcutis*
- **Architecture:** *Lobulated*
- **Vessels:** *Round*
- **Cellularity:** *Hypocellular*
- **Cytology:** *Non-atypical*
- **IHC:** *CD34 +ve*

## Superficial angiomyxoma (cutaneous myxoma)

- **Site:** *Anywhere*
- **Size:** *<4 cm*
- **Level:** *Dermis/subcutis*
- **Architecture:** *Lobulated*
- **Vessels:** *Round*
- **Cellularity:** *Hypocellular*
- **Cytology:** *Non-atypical*
- **IHC:** *CD34 +ve*

### Carney complex

- **Lentiginos (65%)**
- **Cutaneous myxomas (45%)**
  - **Cardiac myxomas (72%)**
  - **Mammary myxomas (42%)**
    - **Cushing's (45%)**

## Digital myxoma (digital mucous cyst)

- **Site:** *Fingers*
- **Size:** *<2 cm*
- **Level:** *Dermis*
- **Architecture:** *None*
- **Vessels:** *Avascular*
- **Cellularity:** *Hypocellular*
- **Cytology:** *Non-atypical*
- **IHC:** *N/A*

# Digital fibromyxoma (superficial acral fibromyxoma)

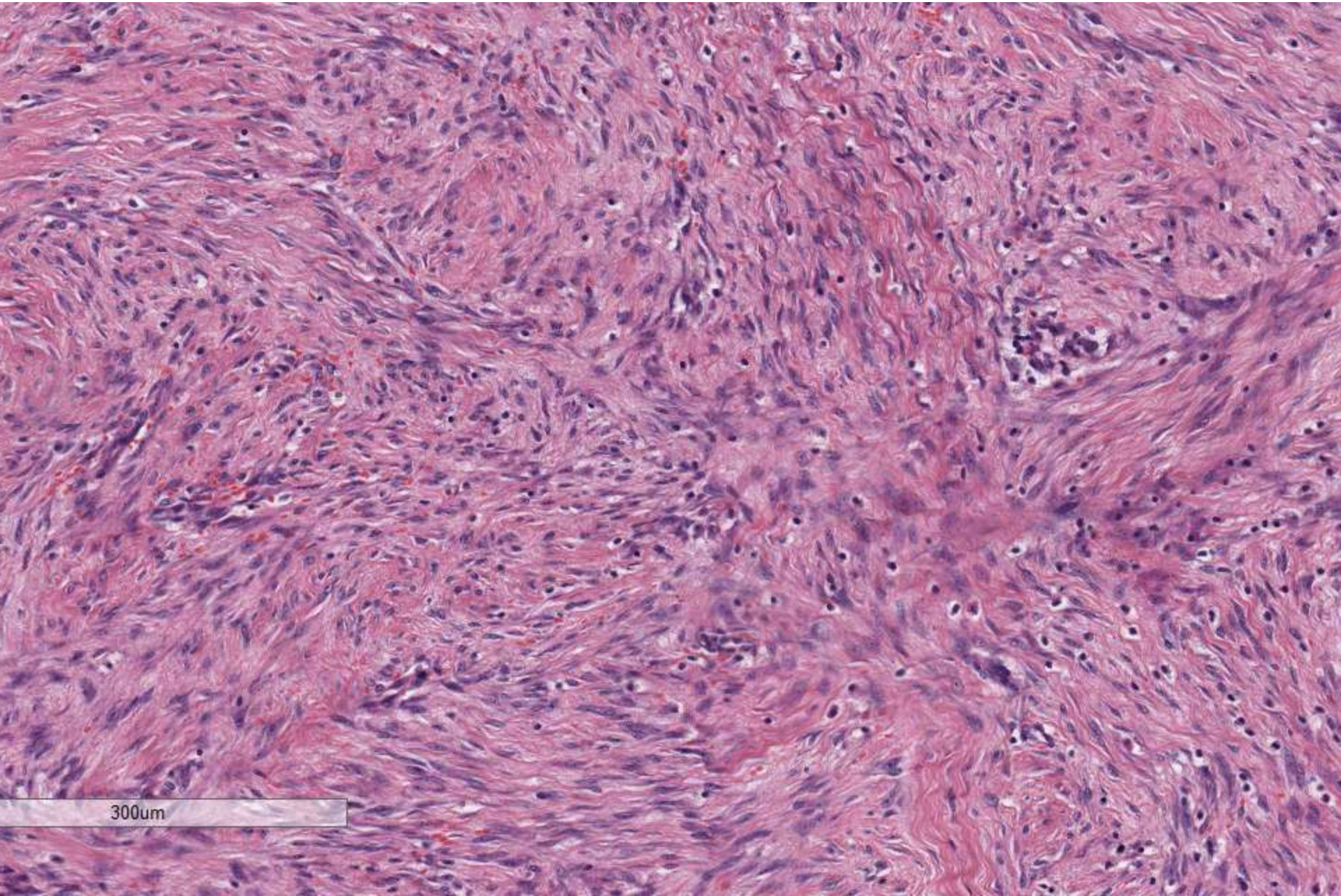
- **Site:** *Fingers*
- **Size:** *<2 cm*
- **Level:** *Dermis*
- **Architecture:** *Mixed collagenous and myxoid stroma*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular*
- **Cytology:** *Non-atypical*
- **IHC:** *CD34 +ve*

<b>Dermal</b>	<b>Dermal nerve sheath myxoma</b>	<b>Superficial angiomyxoma</b>	<b>Digital myxoma</b>	<b>Digital fibromyxoma</b>
<b>Sub-cutaneous</b>				
<b>Intra-muscular</b>				

<b>Dermal</b>	<b>Dermal nerve sheath myxoma</b>	<b>Superficial angiomyxoma</b>	<b>Digital myxoma</b>	<b>Digital fibromyxoma</b>
<b>Sub-cutaneous</b>	<b>Nodular fasciitis, myxoid</b>	<b>Myxofibrosarcoma</b>	<b>Ossifying fibromyxoid tumour</b>	<b>(Acral) Myxoinflammatory fibroblastic sarcoma</b>
<b>Intra-muscular</b>				

## Nodular fasciitis, myxoid type

- **Site:** *Anywhere, but rare on hand and feet*
- **Size:** *<3 cm*
- **Level:** *Subcutis*
- **Architecture:** *Short fascicles*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular to hypercellular*
- **Cytology:** *Non-atypical*
- **IHC:** *SMA +ve*



300µm

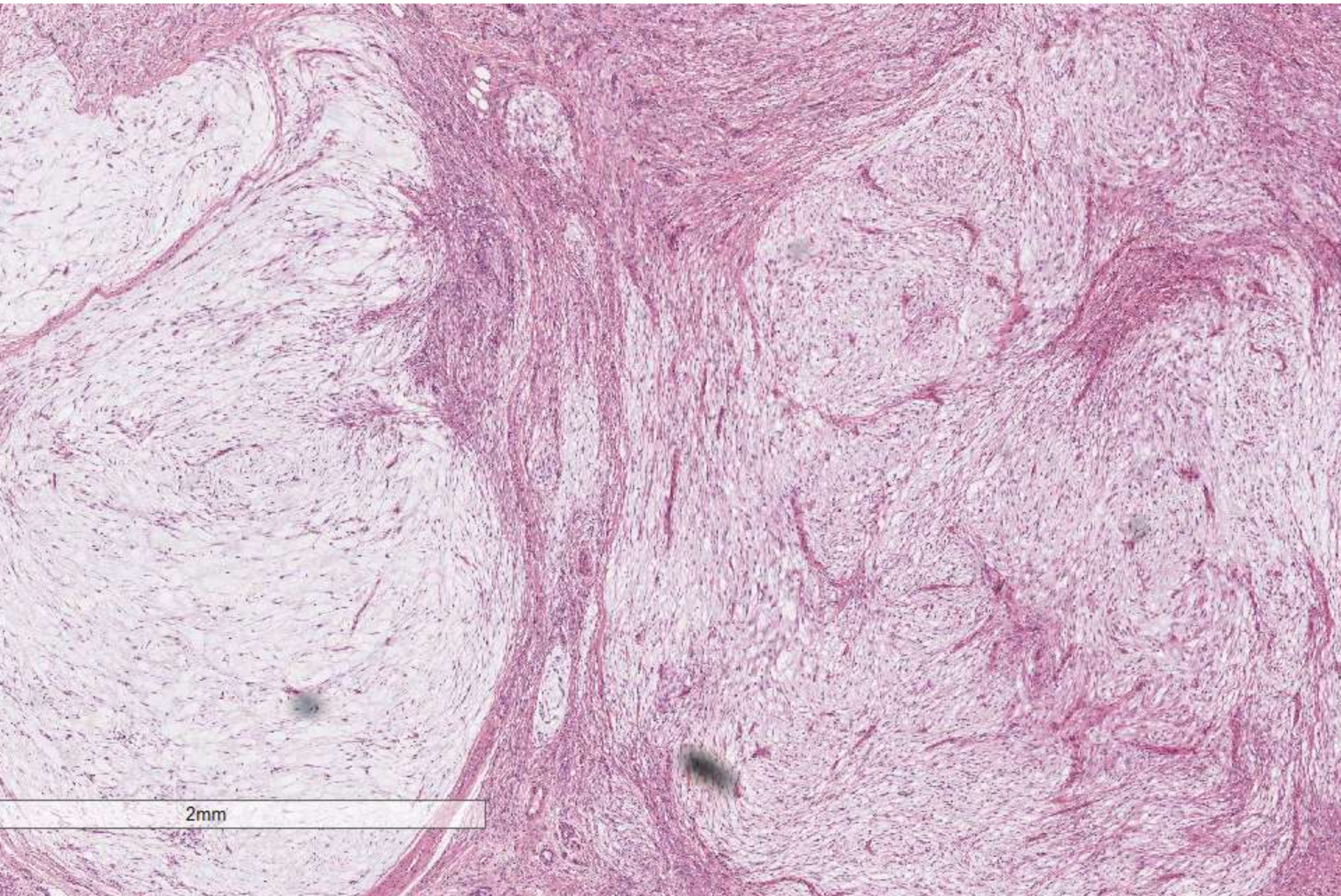
## Myxofibrosarcoma, low- and intermediate-grade (myxoid MFH)

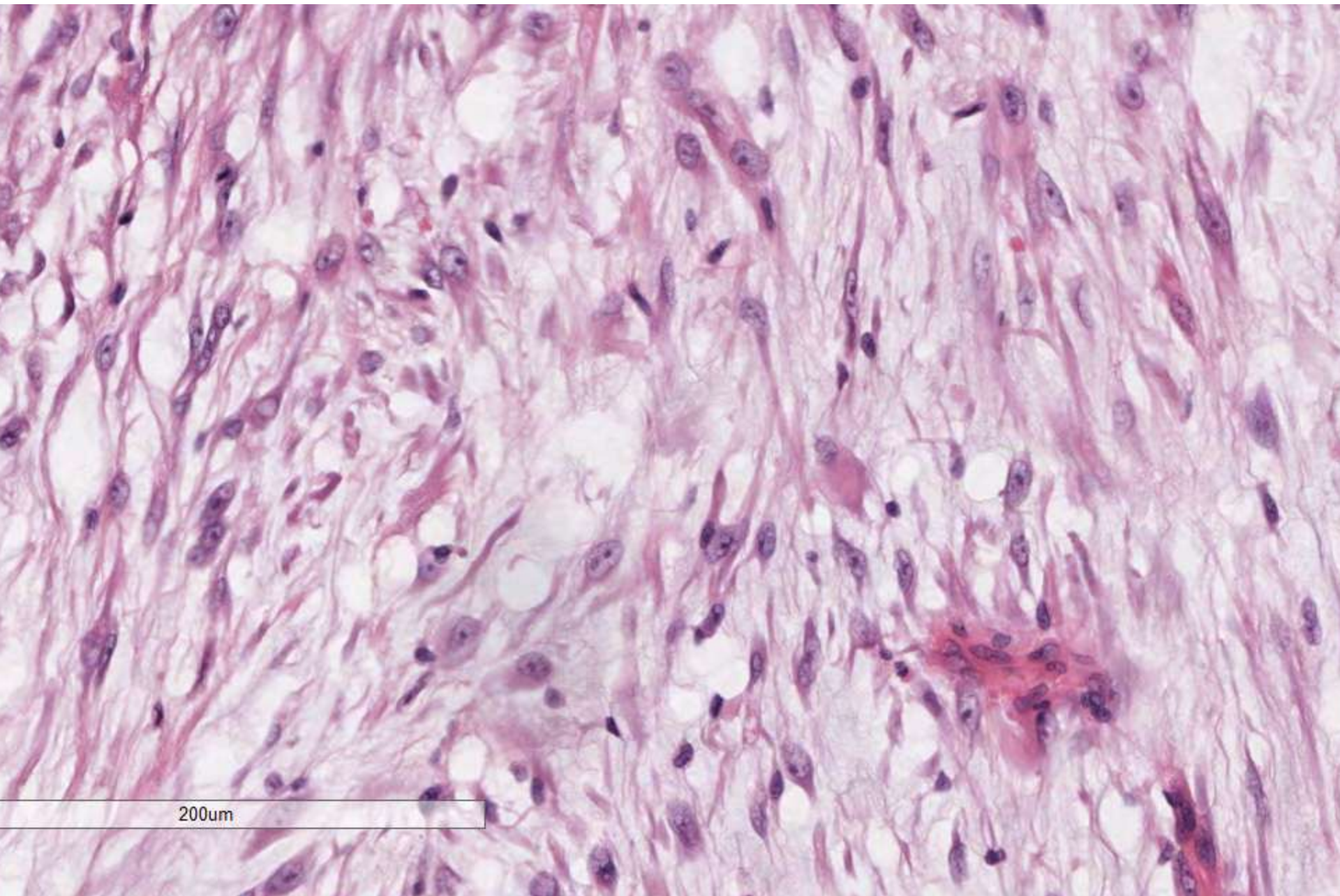
- **Site:** *Anywhere*
- **Size:** *5-10 cm*
- **Level:** *Subcutis (70%)/intramuscular (30%)*
- **Architecture:** *Lobulated*
- **Vessels:** *Curvilinear*
- **Cellularity:** *Hypocellular to moderately cellular*
- **Cytology:** *Mildly to moderately atypical*
- **IHC:** *N/A*

# Myxofibrosarcoma, low- and intermediate-grade (myxoid MFH)

- **Site:** *Anywhere*
- **Size:** *5-10 cm*
- **Level:** *Subcutis (70%)/intramuscular (30%)*
- **Architecture:** *Lobulated*
- **Vessels:** *Curvilinear*
- **Cellularity:** *Hypocellular to moderately cellular*
- **Cytology:** *Mildly to moderately atypical*
- **IHC:** *N/A*

	<b>Myxofibrosarcoma</b>	<b>Low-grade fibromyxoid sarcoma</b>
Age	Elderly	Young to middle age
Level	Subcutaneous	Intramuscular
Stroma	Myxoid	Changeable collagenous and myxoid
Atypia	Mildly to moderately atypical	Non-atypical





200um

## Ossifying fibromyxoid tumour

- **Site:** *Anywhere*
- **Size:** *5-10 cm*
- **Level:** *Subcutis*
- **Architecture:** *Boney shell; lobulated; net-like*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular*
- **Cytology:** *Non-atypical*
- **IHC:** S100 (70%) and desmin (50%)

# (Acral) Myxoinflammatory fibroblastic sarcoma

[Inflammatory myxohyaline tumor of distal extremities with virocyte or Reed-Sternberg-like cells]

- **Site:** *Hands/wrists and feet/ankles*
- **Size:** *1-8 cm*
- **Level:** *Subcutis*
- **Architecture:** *Changeable myxoid, inflammatory and collagenous*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular to hypercellular*
- **Cytology:** *Mildly-to-severely atypical, including RS-like cells*
- **IHC:** *t(1;10)*

**Montgomery E, et al.**

***Inflammatory myxohyaline tumor of distal extremities with virocyte or Reed-Sternberg-like cells.***

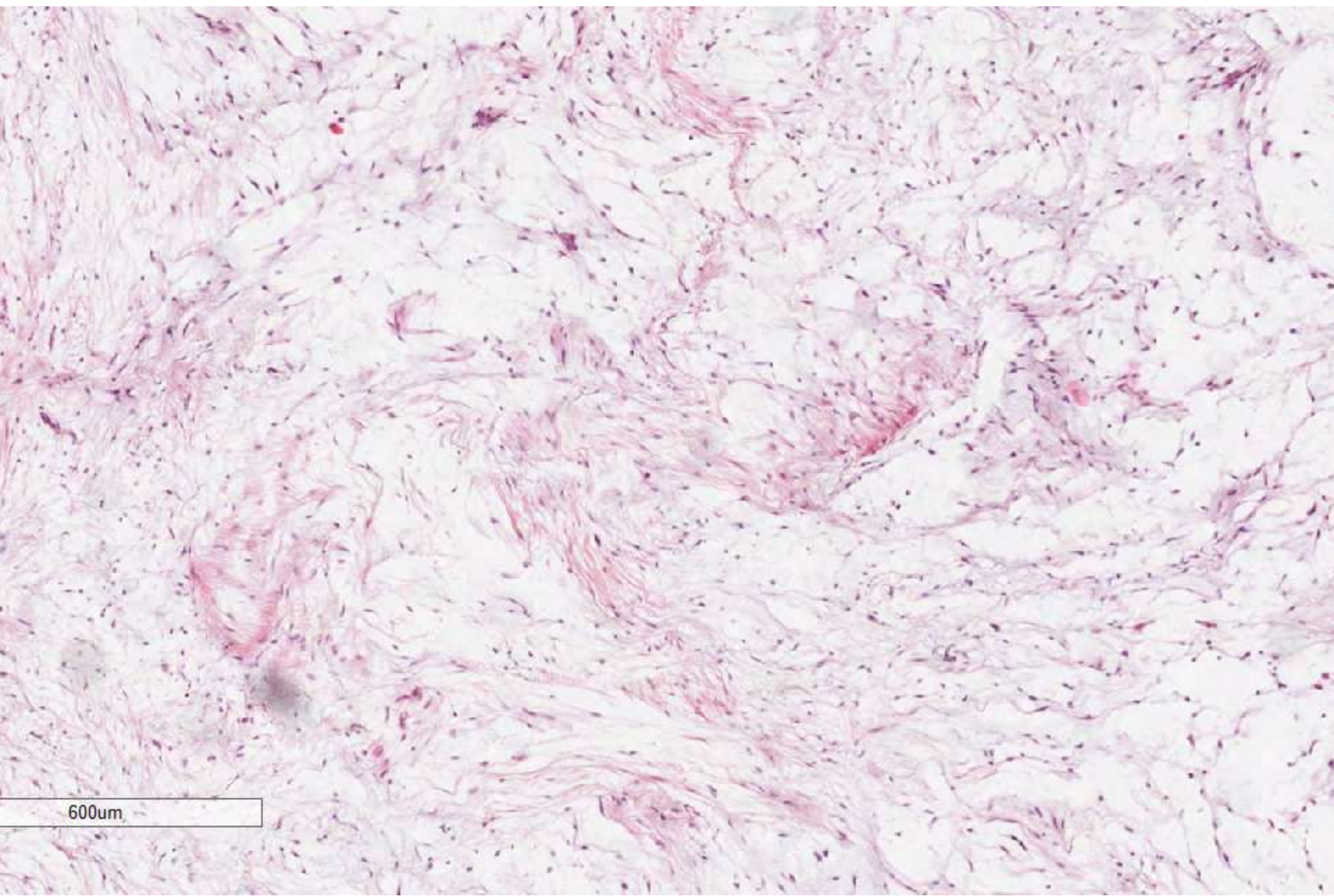
***Mod Patho 1998, 11, 384-391.***

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<b>Intra-muscular</b>				

<b>Dermal</b>	<b>Dermal nerve sheath myxoma</b>	<b>Superficial angiomyxoma</b>	<b>Digital myxoma</b>	<b>Digital fibromyxoma</b>
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<b>Intra-muscular</b>	<b>Intramuscular myxoma</b>	<b>Low-grade fibromyxoid sarcoma</b>	<b>Myxoid liposarcoma</b>	<b>Deep angiomyxoma</b>

## Intramuscular myxoma

- **Site:** *Anywhere*
- **Size:** *5-10 cm*
- **Level:** *Intramuscular*
- **Architecture:** *None*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Hypocellular*
- **Cytology:** *Non-atypical*
- **IHC:** *N/A*



600um

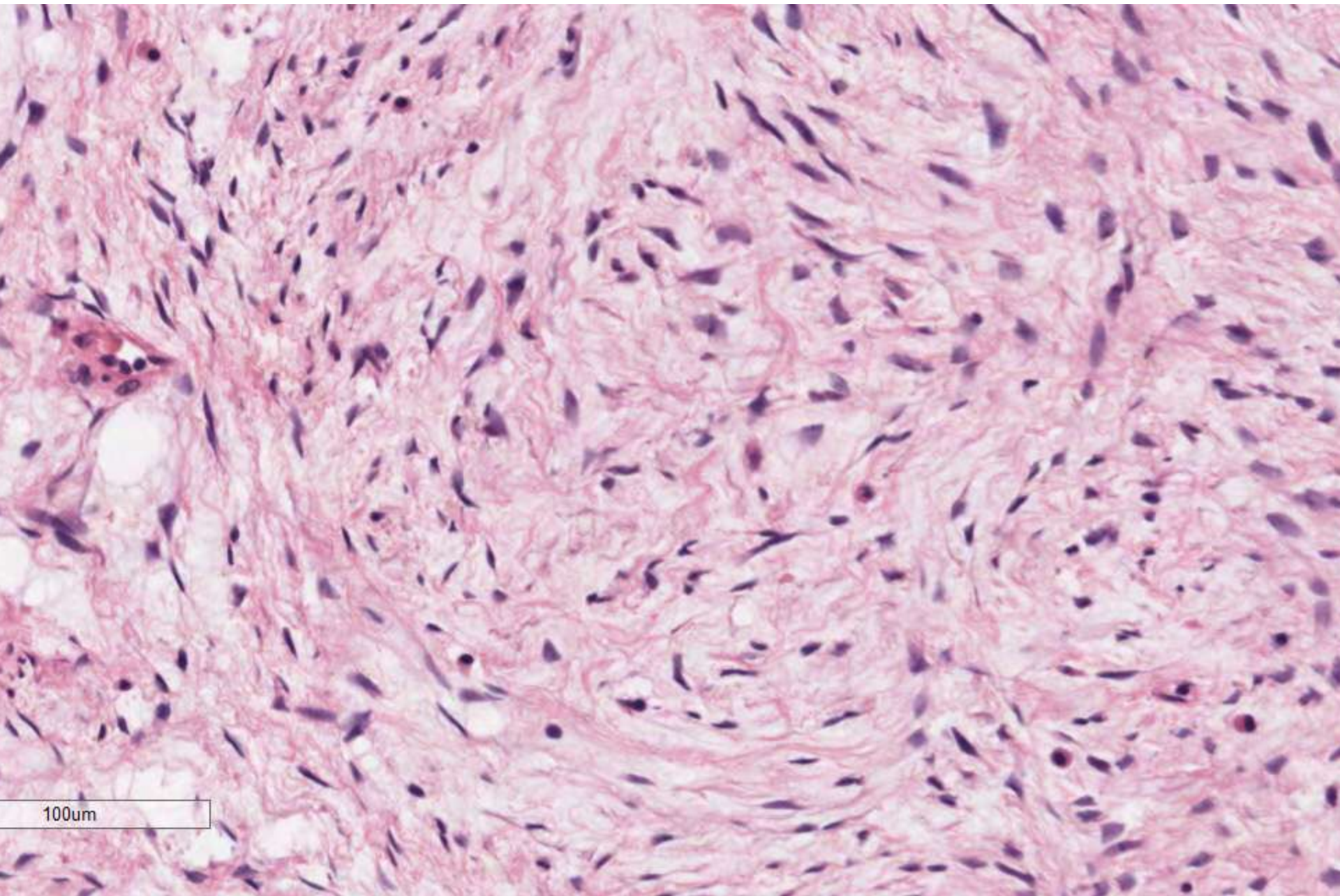
## Low-grade fibromyxoid sarcoma

- **Site:** *Anywhere*
- **Size:** *8-10 cm*
- **Level:** *Intramuscular*
- **Architecture:** *Changeable collagenous and myxoid*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular*
- **Cytology:** *Non-atypical*
- **IHC:** *MUC4 and t(7;16)*

## Low-grade fibromyxoid sarcoma

- **Site:** *Anywhere*
- **Size:** *8-10 cm*
- **Level:** *Intramuscular*
- **Architecture:** *Changeable collagenous and myxoid; whorled*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular*
- **Cytology:** *Non-atypical*
- **IHC:** *MUC4 and t(7;16)*

	<b>Myxofibrosarcoma</b>	<b>Low-grade fibromyxoid sarcoma</b>
Age	Elderly	Young to middle age
Level	Subcutaneous	Intramuscular
Stroma	Myxoid	Changeable collagenous and myxoid
Atypia	Mildly to moderately atypical	Non-atypical



100um

## Low-grade fibromyxoid sarcoma

- **Site:** *Anywhere*
- **Size:** *8-10 cm*
- **Level:** *Intramuscular*
- **Architecture:** *Changeable collagenous and myxoid; whorled*
- **Vessels:** *Inconspicuous*
- **Cellularity:** *Moderately cellular*
- **Cytology:** *Non-atypical*
- **IHC:** *MUC4 and t(7;16)*

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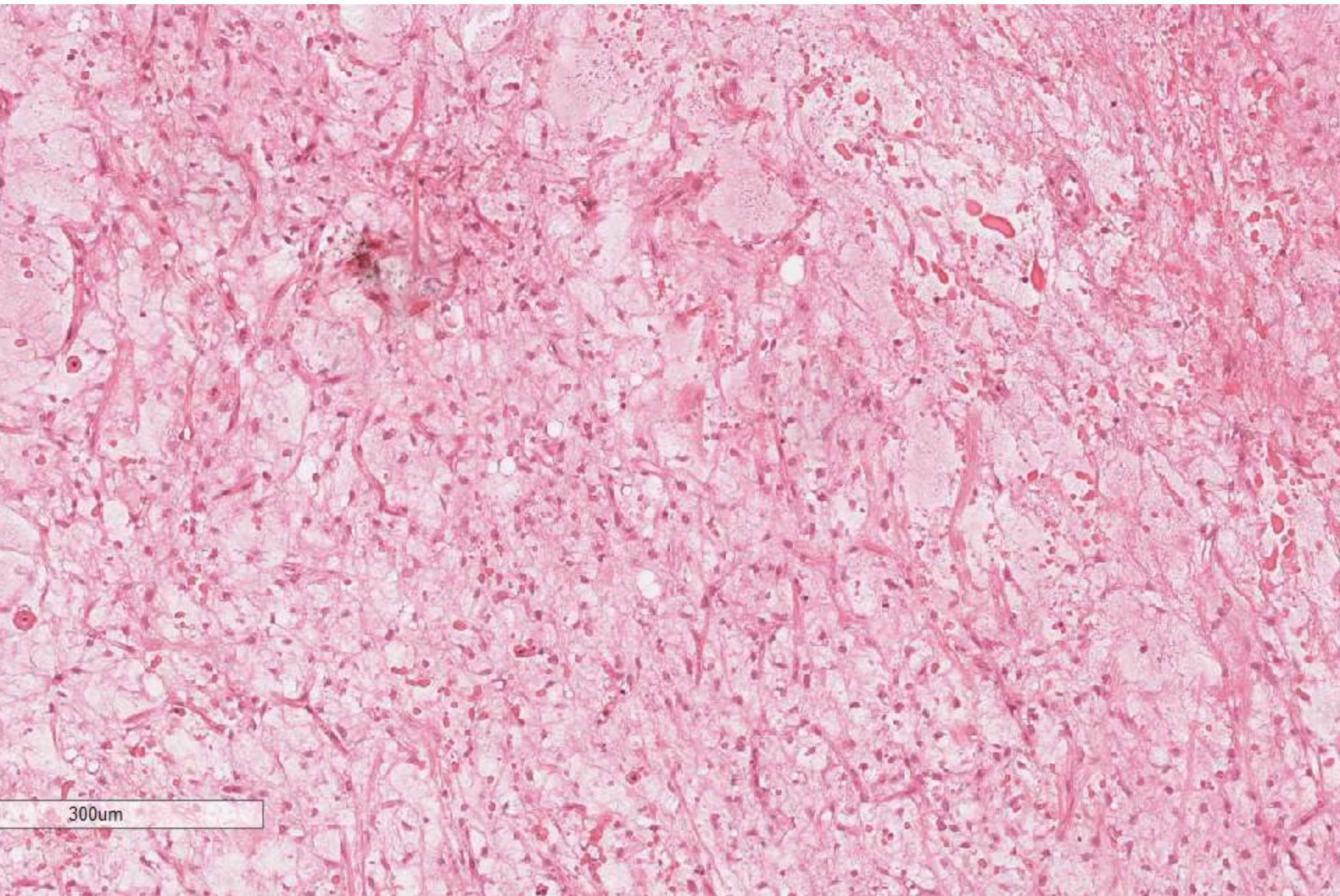
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- **Size:** *8-10 cm*
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- **Architecture:** *Changeable collagenous and myxoid; whorled*
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	<b>Myxofibrosarcoma</b>	<b>Low-grade fibromyxoid sarcoma</b>
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Level	Subcutaneous	Intramuscular
Stroma	Myxoid	Changeable collagenous and myxoid
Atypia	Mildly to moderately atypical	Non-atypical

\* **Variant - Hyalinising spindle cell tumor with giant rosettes.**

## Myxoid/round cell liposarcoma

- **Site:** *Anywhere*
- **Size:** *>5 cm*
- **Level:** *Intramuscular*
- **Architecture:** *None*
- **Vessels:** *Chicken wire or crow's feet*
- **Cellularity:** *Moderately cellular*
- **Cytology:** *Non-atypical + lipoblasts*
- **IHC:** *S100 +ve (40%) and t(12;16)*



300um

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<b>Intra-muscular</b>	<b>Intramuscular myxoma</b>	<b>Low-grade fibromyxoid sarcoma</b>	<b>Myxoid liposarcoma</b>	<b>Deep angiomyxoma</b>

## Honorable mentions

- Extraskeletal myxoid chondrosarcoma
- Chordoma