Adipose and Myxoid Tumors in Young Patients

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The Pathologist’s Challenges

• Almost any soft tissue or mesenchymal tumor can sometimes have a myxoid appearance
• Some adipose tumors can be predominantly myxoid
• Non-neoplastic conditions and non-mesenchymal neoplasms can have myxoid features
Objectives

• To review the more common and/or problematic adipose and myxoid tumors that occur in children and adolescents
• To provide a systematic approach to pathologic evaluation, diagnosis, and differential diagnosis
Adipose Tumors in Children and Adolescents *

- Lipoma, all types: 64%
- Lipoblastoma: 30%
- Hibernoma: 2%
- Liposarcoma: 4%

*Lipomatoses not included; Minnesota series*
Lipoblastoma

- Prototypical benign adipose tumor of childhood
- Infants and young children (90% before 3 years of age)
- Extremities and trunk most frequent sites
- May be associated with developmental delay or abnormalities, malformations, seizures
- Risk of recurrence with incomplete excision
- Rearrangement or amplification of *PLAG1* gene on 8q13
Lipoblastoma: desmin, S100
Lipoblastoma with t(7;8)
Lipoblastoma with maturation
Lipoblastoma with maturation
Lipoma
Myxoid Liposarcoma

- Most common liposarcoma of childhood
- Peak age 3rd-6th decades
- Deep soft tissue of extremities or retroperitoneum, head and neck
- Favorable histologic-prognostic type unless significant round cell component
- Rearrangement of \textit{CHOP} gene on 12q13 with \textit{FUS}(16p11) or \textit{EWS} (22p11) as partners
Myxoid liposarcoma
Myxoid liposarcoma
Myxoid liposarcoma
Myxoid/ round cell liposarcoma
Other Liposarcoma Subtypes in Young Patients

• Pleomorphic-myxoid: very aggressive, poor prognosis
• Spindle cell-myxoid: frequent local recurrence
• Other LPS subtypes: similar to adults
Pleomorphic-myxoid liposarcoma
Pleomorphic and round cell liposarcoma
Inflammatory liposarcoma
Myxoid Soft Tissue Tumors in Young Patients

- Extensive differential diagnosis: almost any mesenchymal neoplasm (and also other types of neoplasms) can have a myxoid variant
- Myxoid variants of specific sarcomas and other soft tissue neoplasms are a particular challenge
- True myxoid and angiomyxoid tumors are uncommon
- Non-neoplastic conditions can form myxoid masses (i.e., cutaneous mucinosis)
Differential Diagnosis: Points for Consideration

- Clinical features
- Location: superficial or deep
- Pathologic themes:
  - Architectural pattern
  - Cellularity
  - Vascularity
  - Atypia
  - Cytodifferentiation
- Diagnostic adjuncts
Aggressive Angiomyxoma

- Infiltrative mass in deep soft tissue of perineum, pelvis, and genitalia
- Female predominance
- Rare pediatric and adolescent cases
- Irregular clusters of blood vessels in a hypocellular myxoid stroma
- High risk of local recurrence (70%)
Aggressive angiomyxoma
Aggressive angiomyxoma
Superficial Angiomyxoma

• Cutaneous and subcutaneous mass
• Subset in children and adolescents
• Angiomyxoid nodules, mucin pools, sparse inflammation, small blood vessels, epithelial component
• May recur locally
Superficial angiomyxoma
Myxoma

- Superficial or deep soft tissue, juxtaarticular soft tissue, or muscle
- Slow growth, completely benign
- Multiple myxomas associated with fibrous dysplasia of bone
- Stellate and spindle cells with fibroblastic-myofibroblastic features in an abundant myxoid background, no atypia or mitoses, no prominent vascular component
- $GNAS1$ mutation (intramuscular myxoma)
Benign Soft Tissue Masses with Myxoid Features

- Neurofibroma
- Dermal nerve sheath myxoma
- Plexiform fibromyxoma
- Ossifying fibromyxoid tumor
- Ganglion cyst
- Myxoid pattern variation in many others
Neurofibroma
Dermal nerve sheath myxoma

S100

GFAP
Plexiform fibromyxoma
Ossifying fibromyxoid tumor
Ganglion cyst
Intermediate Myxoid Soft Tissue Tumors

- Primitive myxoid mesenchymal tumor of infancy (PMMTI)
- Solitary fibrous tumor (SFT)
- Giant cell fibroblastoma and dermatofibrosarcoma protuberans
- Myxoid pattern variation in others
Vimentin

PMMTI
Solitary fibrous tumor
Solitary fibrous tumor
Solitary fibrous tumor
Giant cell fibroblastoma
Myxoid DFSP, CD34
Myoepithelioma
Sarcomas with Myxoid Features

- Low grade fibromyxoid sarcoma
- Myxoinflammatory fibroblastic sarcoma
- Myxofibrosarcoma
- Undifferentiated sarcoma with myxoid features
- Extraskeletal myxoid chondrosarcoma
- And many others!
Low grade fibromyxoid sarcoma
Low grade fibromyxoid sarcoma
Myxoinflammatory fibroblastic sarcoma
Undifferentiated sarcoma
Myxoid chondrosarcoma (extraskeletal)
Myxoid chondrosarcoma
Adipose and Myxoid Tumors: Summary

• A diagnostic challenge in some cases

• Distinguishing features
  • Clinical aspects
  • Superficial or deep
  • Architectural pattern
  • Cellularity, vascularity, atypia

• Diagnostic adjuncts
  • Immunohistochemistry
  • Cytogenetics, molecular genetics

• Pitfalls with nonneoplastic conditions and non-mesenchymal neoplasms