Pediatric Soft Tissue Tumors

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Case Presentation

• 2 year old girl presents with R thigh painless mass

• No developmental or physical delays
MRI
Case Presentation

- Radiography consistent with lipoblastoma
- Mass excised without complications
- Pathology showed lipoma
Pediatric Adipose Tumors

• Relatively uncommon

• Mostly benign

• Derived from white adipose tissue

Coffin CM. In *Pediatric soft tissue tumors, 2nd Ed.* 1998
Pediatric Adipose Tumors

- Lipoma 64%
  - Angiolipoma 28%
  - Fibrolipoma 8%
- Lipoblastoma 30%
- Hibernoma 2%
- Liposarcoma 4%

Lipoblastoma

• “Infantile lipoma“

• Rare, benign, mesenchymal tumor

• Predilection for extremities, trunk, head and neck
  • Less often in mediastinum, scrotum, retroperitoneum, buttock and perineum

• Anomalies of chromosome 8

Coffin CM. In Pediatric soft tissue tumors, 2nd Ed. 1998
Lipoblastoma

• 16 cases of lipoblastomas in Taiwan

• 4 recurrences: neck (n=2) and lower extremities (n=2)

• 3 year follow-up recommended

Lipoblastoma

www.histopathology-india.com/Lipoblastoma.htm
Lipoblastoma

www.histopathology-india.com/Lipoblastoma.htm
Hibernoma

- “Brown lipoma”
- Commonly found on back, chest, axilla, groin, thigh
- Wide spectrum of adipocyte maturation
- Endocrine secretory features

Coffin CM. In *Pediatric soft tissue tumors, 2nd Ed.* 1998
Infantile Myofibromatosis

- Most common fibrous tumor of infancy
- Solitary and multicentric forms
- Spindle cells, fibroblasts, bland necrosis with stippled calcification
- Diagnosis by biopsy
- Close observation appropriate

Pielop JA & Levy ML. UpToDate® 2006
Subcutaneous Fat Necrosis

- Rare condition in term or post-term newborns following perinatal complications

- Multiple firm nontender subcutaneous fat nodules that liquefy or calcify

- Granulomatous reaction in fat

- Expectant management

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Liposarcoma

- Relatively uncommon
- Common in thigh, upper extremity, chest, abdomen
- Predominantly myxoid liposarcoma
- Low malignant potential

Coffin CM. In *Pediatric soft tissue tumors, 2nd Ed.* 1998
Myxoid Liposarcoma

www.sarcoma.org/source/myxoid_liposarcoma2
Pleomorphic Liposarcoma

www.sarcoma.org/source/pleomorphic_liposarcoma2
Dermatofibrosarcoma Protuberans

- Uncommon soft tissue sarcoma with unique translocation in chromosomes 17 and 22
- Mostly low to intermediate-grade, rarely metastasize, but recur locally
- Complete resection with wide margins is optimal, adjuvant RT improves local control
- Mohs micrographic surgery preferred for large and recurrent tumors
- Imatinib can induce tumor regression in recurrent, unresectable, and metastatic disease

Mendenhall, WM, et al. UpToDate® 2006
Dermatofibrosarcoma Protuberans
Dermatofibrosarcoma Protuberans

www.bweems.com/dfsppolypg1
Dermatofibrosarcoma Protuberans

- Prospectively collected data in 159 pts at Memorial Sloan-Kettering Cancer Center from 1950-1998
- 134 pts (84%) had classic DFSP, FS variant in remaining 25 pts (16%)
- 75% 5-year local recurrence free survival rate (81% in classic and 28% in FS variant)
- 34 pts (21%) with recurrence, 2 deaths from metastatic disease
- Increased age, high mitotic index, and increased cellularity predict poor outcome, FS variant much more aggressive with metastatic potential

Infantile Fibrosarcoma

- Rapidly growing, poorly circumscribed spindle cell tumor
- Present at birth or during the 1st year of life
- Translocation results in fusion gene
Infantile Fibrosarcoma

- Locally infiltrative, rarely metastasize
- Good prognosis
- Amputation rate >50%
- Neoadjuvant chemotherapy may be helpful

Pielop JA & Levy ML. UpToDate® 2006
Infantile Fibrosarcoma

Summary & Recommendations

• Excise all large or rapidly growing tumors

• Communication with pathology

• Close follow up