EXTRAMEDULLARY TUMORS
OF THE PEDIATRIC SPINE

Eugene Wang
1/20/12
Dent Neurologic Institute

Introduction

- 2/3 of all intraspinal tumors of childhood are extramedullary
- 50% Extradural
- 10-15% Intradural
- Back pain, radicular pain are most frequent presenting symptom for spinal tumors (25-30%)
- Torticollis, UE weakness, scoliosis, urinary incontinence
- Histologic type of tumor cannot be predicted by clinical presentation (Barkovich, Pediatric Neuroimaging)

Classification for Extramedullary Tumors

- Classified according to site of origin
- CSF Dissemination of Intracranial Neoplasms
- Spinal column tumors
  - Aneurysmal bone cysts, LCH, giant cell tumors, Ewing sarcoma, osteoid osteoma, osteoblastoma, osteosarcomas
- Intraspinal Tumors
  - Meningeal (meningiomas)
  - Nerve roots/nerve root sheaths (neurofibromas, schwannomas)
  - Lymphomas, Leukemias, PNETs
- Extraspinal tumors that invade epidural space
  (neuroblastoma-ganglioneuroblastoma-ganglioneuroma spectrum)
CSF Dissemination of Intracranial Neoplasms

- Dissemination of tumor from intracranial neoplasms via CSF occurs more frequently in pediatric age group than in adults.
- 1/3 of patients with medulloblastoma will eventually disseminate tumor through the CSF, most often to the spine.
- Ependymomas, anaplastic gliomas, germinomas, choroid plexus tumors, pineal tumors also frequently disseminate through CSF and deposit in the spinal canal.
- Contrast enhanced MR is imaging study of choice to search for CSF spread of tumor.
Spinal Column Tumor - Aneurysmal Bone Cyst

- Most common in children and young adults
- Not true neoplasms but expansile vascular lesions of unknown cause
- On MR, show variable signal intensities, usually high T2 signal, high or medium T1 signal
- Fluid-filled levels if present are very helpful for diagnosis.
- Can displace cord
Spinal Column Tumor-Osteoid Osteoma

- Benign osteoblastic lesion of unknown etiology, often presents as painful scoliosis of T and L-spine
- Nidus of osteoid matrix and stroma of loose vascular connective tissue, often with calcification
- Tc bone scintigraphy most accurate modality with uptake of bone tracer, targeted CT also helpful
- MRI: nidus appears isointense on T1W and hypointense on T2 weighted FSE sequences
- Gadolinium enhancement can improve conspicuity of osteomas.

Spinal Column Tumor-LCH

- Group of rare disorders characterized by abnormal proliferation of histiocytes in variety of organs: skin, bone, internal organs
- Eosinophilic Granuloma is most frequent and benign—manifest as unifocal or multifocal osseous lesion
- Hand-Schulter-Christian presents in children or young adults: exophthalmos, osteolytic skull lesions, DI
- Disseminated LCH (Letterer-Sieue Disease): multiorgan involvement, lethal outcome, in <3 yo
- Imaging: Vertebral plana: Often show vertebral body collapse or lytic vertebral lesion at time of presentation. Two intervertebral discs in apposition without intervening vertebral body (classic appearance)
- Soft tissue extending into spinal canal with homogenous enhancement may be present as well.
Spinal Column Tumor-LCH

Develop primarily in adults, only few anecdotal pediatric cases seen.

Usually present in 2nd decade with localized pain or mass.

Variable gross pathologic appearance (soft and friable to firm and calcified). Because of this, imaging is variable as well.

MRI usually shows the tumor as hypointense T1, hyperintense T2. Tumors that are heavily ossified may be dark on T2 or T2*.

Spinal Column Tumor-Osteogenic Sarcoma

Extremely rare in spine, though 20% of all sarcomas.

Develop primarily in adults, only few anecdotal pediatric cases seen.

Usually present in 2nd decade with localized pain or mass.

Variable gross pathologic appearance (soft and friable to firm and calcified). Because of this, imaging is variable as well.

MRI usually shows the tumor as hypointense T1, hyperintense T2. Tumors that are heavily ossified may be dark on T2 or T2*.
Spinal Column Tumor-Osteogenic Sarcoma

- 4% of primary benign bone tumors
- Peak incidence in 3rd decade, sometimes teens affected
- Destructive, lytic lesions with poorly defined, nonsclerotic margins.
- Vertebral body selectively involved in most cases. Involved bone is expanded.
- MR: Low to medium signal on T1W sequences, and high signal intensity with low signal rim on T2W. Minimal enhancement

Spinal Column Tumor-Giant Cell Tumor

- 4% of primary benign bone tumors
- Peak incidence in 3rd decade, sometimes teens affected
- Destructive, lytic lesions with poorly defined, nonsclerotic margins.
- Vertebral body selectively involved in most cases. Involved bone is expanded.
- MR: Low to medium signal on T1W sequences, and high signal intensity with low signal rim on T2W. Minimal enhancement
Spinal Column Tumor-Chordomas

- Extremely rare in children
- Arise from rests of notochord cells and primarily involve the clivus and sacrum.
- Only 15% of chordomas involve the vertebrae.
- Chordomas grow slowly by erosion and compression of adjacent structures
- Imaging: Lytic lesions cause destruction of one or more adjacent vertebrae. Sclerosis is commonly seen in bone adjacent to tumor.
- MRI: Similar appearance to other primary bone tumors. Sharply defined, destructive lesions with prolonged T1 and T2 relaxation times.
- Absence of contrast enhancement favors diagnosis of chordoma, as most other tumors of pediatric spinal column show significant enhancement.

Spinal Column Tumor-Ewing Sarcoma

- Second most common primary bone tumor in childhood.
- Vertebral body or sacrum
- Most common age presentation is 2nd decade, M:F 2:1
- CT or plain radiograph of Ewing sarcoma reveals lytic lesions with margination.
- MRI: Hypointense compared to normal bone on T1 and variable signal on T2.
- Uniform enhancement on post GAD sequence
- Primary osseous or extra-osseous spinal tumor (peripheral PNET)
Spinal Column Tumor - Ewing Sarcoma

- Less than 1% of primary bone tumors
- Most common in males during 2nd and 3rd decades of life
- Tumors usu 2 cm or greater, therefore pain 2/2 compression of nerve roots or spinal cord sometimes part of presentation
- Well-defined solitary masses
- MR: well-defined masses showing slight hypointensity on T1W images, slight hyperintensity on T2W images compared with normal bone
- Commonly occur in articular facets, pedicles, lamina.
- Variable contrast enhancement, usu mild

Spinal Column Tumor - Osteoblastoma

- Less than 1% of primary bone tumors
- Most common in males during 2nd and 3rd decades of life
- Tumors usu 2 cm or greater, therefore pain 2/2 compression of nerve roots or spinal cord sometimes part of presentation
- Well-defined solitary masses
- MR: well-defined masses showing slight hypointensity on T1W images, slight hyperintensity on T2W images compared with normal bone
- Commonly occur in articular facets, pedicles, lamina.
- Variable contrast enhancement, usu mild

Intradural, Extramedullary Tumors - Meningioma

- Uncommon in children, incidence of 2-3% of pediatric intraspinal tumors, but histological, clinical, therapeutic features similar to meningiomas occurring later in life
- Homogeneous extramedullary lesions isointense with spinal cord and surrounded by CSF.
- Dural based and sharply margined.
- 90% are intradural, 80% arise in Thoracic spine.
- Isointense to hypointense compared with gray matter on T1 and T2. Homogenously enhances on post-GAD
Intradural, Extramedullary Tumors—Neurofibromas or Schwannomas

- Pathological distinction of neurofibroma from schwannoma from neurofibroma is somewhat hazy; separate solitary encapsulated lesions generally defined as schwannoma, while nerve inseparable from nerve sheath tumor are defined as neurofibromas.

- Schwannomas are very uncommon in children, except when assoc with NF2.
  - 70% are intradural, 30% are extradural.
Intradural, Extramedullary Tumors - Neurofibromas or Schwannomas

Neurofibromatosis
Neurofibromatosis

Extraspinal tumors that invade epidural space: Neuroblastoma

- Tumors originating in paraspinous soft tissues sometimes enter spinal canal through intervertebral foramen.
- Neuroblastomas are the 4th most common tumors of childhood.
- Most often originate in adrenal medulla (40%), but also arise in sympathetic nervous system.
- Often metastasize to bone, including spine, sometimes resulting in spinal cord compression.
- MRI: optimally shows paraspinous or bony mass and extension of tumor into spinal canal, without need for intrathecal contrast. Mass enhances heterogeneously. On noncontrast images, mass appears homogenous and isointense to nervous tissue.
Extraspinal tumors that invade epidural space—Neuroblastoma

Extraspinal tumors that invade epidural space—Ganglioneuroma

- Ganglioneuromas represent tumors of SNS that are at the benign end of neuroblastoma—ganglioneuroblastoma—ganglioneuroma spectrum.
- Most frequent site of origin for ganglioneuromas is posterior mediastinum.
- No metastasis.
- When spinal canal is involved, it is result of dumbbell mass extending from paraspinous region through neural foramen into epidural space.
- Calcification freq present within the tumor.

Extraspinal tumors that invade epidural space—Ganglioneuroblastoma

- Intermediate between neuroblastoma and ganglioneuroma.
- Least common, tends to present in young children.
- Can metastasize to bone or extend directly into spinal epidural space through neural foramina.
- MRI shows full extent of paraspinous and epidural tumor can be defined noninvasively and without use of ionizing radiation.
Extraspinal tumors that invade epidural space—Leukemia

- Leukemic involvement of spinal epidural space is rarely symptomatic.
- Majority of patients with symptomatic spinal epidural leukemic deposits are in pediatric age groups—usually present with signs of meningeal irritation.
- MR: Epidural mass is isointense to neural tissue on noncontrast studies; homogenous enhancement occurs after contrast. May be identical to PNET and lymphoma.

Extraspinal tumors that invade epidural space—Leukemia and Lymphoma

- Relatively rare in children.
- In children with either leukemia or lymphoma, normal high signal intensity of bone marrow on noncontrast T1 images often replaced by lower signal intensity.
- Not clear whether this abnormal signal results from actual leukemic infiltration of vertebreal bodies, increased activity of bone marrow, or result of chemotherapy.
- On MR, epidural lesions in Leukemia or Lymphoma are isointense to hyperintense on T1W images and isointense to hypointense on T2W images, may demonstrate mod to marked contrast enhancement.
- Often respond rapidly to chemo and radiation.

Extraspinal tumors that invade epidural space—Lymphoma
Extraspinal tumors that invade epidural space—Leukemia

- When children develop peripheral PNETs in the spine or paraspinal soft tissues, typically present with increasing paraparesis or quadriplegia, secondary to spinal cord compression.
- T1W: hypointense to isointense; iso to hyperintense on T2W images. Variable enhancement postcontrast.
- Masses are heterogeneous, with cystic or necrotic areas and solid areas.
- When lesions are of paraspinal origin, they commonly invade spinal canal via neural foramina, resulting in foraminal enlargement and sometimes spinal cord compression. Cannot differentiate from neuroblastoma-ganglioneuroblastoma-ganglioneuroma continuum.
Extraspinal tumors that invade epidural space-PNET