INTRODUCTION

- Primary malignant bone tumor of embryonic notochordal remnants
- 1-4% of primary bone tumors, <0.1 per 100,000
- Location: Sacral (50%), Skull base (35%), Vertebral column (15%)
- Classic, chondroid (5-15%), and dedifferentiated (5%) variants
- Most commonly in late middle age (50s to 60s)
- Low-grade, slow growing tumor
- But locally aggressive, high rate of local recurrence (20% in 1st year)
- Local recurrence is most important predictor of mortality
- Metastasis only occurs very late in disease
- Median survival of about 6 years, less than 12 months with mets
- 5 year survival of 70%, 10 year survival of 40%
- Primary therapy- aggressive surgical resection (if possible)
- New targeted therapies currently under investigation

Patient Presentation

32 year old with non-specific low back pain and radiculopathy
No other clinical findings

Common Findings
- Often clinically silent until late in disease
- Insidious onset of pain is most common initial symptom
- May exist for 14-24 months prior to diagnosis
- Neurologic deficits can occur based on location

Radiological Imaging

Pathology and Immunohistochemistry

Gross Appearance
- Purple to Gray
- Cystic and gelatinous

CT-guided Needle Core Biopsy
- Weak S-100 Positive Staining
- Strong CK19 Positive Staining

Immunohistochemical Markers
- Strong positive staining for cytokeratin 19 (epithelial marker)
- Weakly positive staining for S-100 protein
- Positive staining for epithelial membrane antigen (EMA)
- Positive staining for Brachury (transcription factor)
- Negative staining for D2-40, GFAF, CD10, CD20, CD138

Histopathologic Variants

Chondroid Chordoma- hyaline or myxoid cartilage, better prognosis
Dedifferentiated Chordoma- high-grade spindle cells, poor prognosis

Histopathologic Differential Diagnosis

Chondrosarcoma- cartilage, negative CK19,EMA, Brachury staining
Multiple myeloma, myxopapillary ependymoma, chordoid meningioma

REFERENCES

Future Directions

Proton or Heavy Particle Radiation Therapy
Deliver higher dose radiation with minimal injury to surrounding tissues
No exit dose and potential radiobiologic effects
Evidence suggests improved local control and lower recurrence
Significant construction and operational expense of hadrons

Molecular Targeted Therapy
Overexpress receptor tyrosine kinases PDGF, EGFR, KIT, c-MET
Investigational anti-tumor treatment with TK inhibitors
Clinical response from Imatinib treatment in advanced disease
Stable disease with symptomatic improvement

Erlotinib, Gefitinib, Cetuximab (EGFRi) and Sunitinib (PDGFi, VEGFi)