Dedifferentiated Chondrosarcoma

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Chondrosarcoma

- Malignant tumour of bone
  - Produces cartilage matrix
  - Primary or secondary
  - Intramedullary or surface (usu from osteochondromas)
  - 5\textsuperscript{th} - 6\textsuperscript{th} decades
  - M:F 1.5:1
  - Ollier’s & Maffucci’s syndromes
    - 3\textsuperscript{rd} - 4\textsuperscript{th} decades
Chondrosarcoma

- 3 additional types
  - Clear cell chondrosarcoma
    - Low grade ?adult equivalent of chondroblastoma
    - Femoral & humeral epiphyses – good prognosis
  - Mesenchymal chondrosarcoma
    - Rare. Spine, ribs & jaw. Females
    - 3rd decade, can be massive & metastasise
  - Dedifferentiated chondrosarcoma
Dedifferentiated chondrosarcoma

- Rare form
  - 11% of all chondrosarcomas
- Mixed low grade chondrosarcoma with high grade spindle cells
  - Spindles not identifiable as cartilage origin
- May have features of osteosarc, MFH, hystiocytyoma
- Biphasic on x-ray
  - Endosteal scalloping/cortical thickening
  - Cortical destruction & soft-tissue invasion
Dedifferentiated chondrosarcoma

- Older patients >50yrs
- Presentation
  - Pain
  - Swelling
  - Nerve compression
  - Rare pathological #
- Site
  - Pelvis>prox femur>prox humerus>distal femur>rib
Anatomic location

Dedifferentiated Peripheral Chondrosarcoma - 18 cases
Age, gender and anatomic locations

Staals et al 2007
Dedifferentiated chondrosarcoma

• Classification
  – Surface
  – Central
    • I – like conventional central but cortical penetration, soft-tissue mass, pathological #, ossification & rapid progression
    • II – benign/low-grade cartilaginous tumour with aggressive malignancy developed
    • III – no x-ray evidence of cartilage component
Radiographic features

- Biphasic
  - Differential aggressiveness
- Large areas of bone destruction
  - Little/no reactive changes
  - Large unmineralised soft tissue mass (72% cases)
Dedifferentiated chondrosarcoma
Dedifferentiated chondrosarcoma
Management

- Establish diagnosis
  - History + exam
    - Rapidity of onset
    - Local pressure effect
  - Imaging
    - X-ray
    - MRI
    - Bone scan

- Blood tests
- Tissue biopsy
  - Specialist centre
- Staging
  - Chest x-ray & CT

- Chemotherapy
  - Controversial
  - No role for neoadjuvant
Operative treatment

- May not be possible
- Amputation as effective as excision & EPR
- Debulking/intralesional excision may be considered on location & symptoms where cure not feasible
Outcome

- Universally poor
- Median survival
  - < 6/12  
    Dahlin et al. 1971
  - 7.5/12  
    Dickey et al. 2004
- 5 yr survival
  - 13%  
    Mercuri et al. 1995
  - 7.1%  
    Dickey et al. 2004
- Surgery + chemo 36% 5 yrs
- Surgery alone 0% at 1 yr  
  Mitchell et al. 2000
Outcome

• European-wide review
  – 337 pts from 9 centres
  – Median age 59
    • 53% M
  – Commonest sites femur & pelvis
  – 21% mets at presentation
    • Median survival 5/12
    • 10% 2 yr survival

• 266 pts with local disease only
  – 254 surgery
    • 79% limb salvage

• 10 yr survival 28%

• Poor prognosis
  – Pathological #
  – Pelvic location
  – ↑Age

• Survival rel to excision margin
Summary

• Dedifferentiated chondrosarcoma
  – Mixed grade tumour
  – Biphasic on x-ray
  – > 50 yrs
  – Femur & pelvis
  – Surgery is best treatment but may not be feasible
  – Chemo controversial
  – 5 yr survivorship poor