Non bone forming tumours: Malignant

EWING SARCOMA

ging Large destructive lesion that involves the metaphysis & diaphysis. Maybe purely lytic or have variable amounts of reactive new bone formation. Periosteal reaction 'onion skin' & Codman's Δ. MRI large soft tissue component

ation Knee, femoral diaphysis, pelvis & prox humerus

es Children > 5yrs

Pain & fever, ↑ESR & WCC ∴ mimics infection

Ix t(11:22) chromosomal translocation EWS/FLI1 fusion gene
Bone marrow biopsy for staging. Involvement = poor prognosis

Tx Chemo & wide margin excision, Euro Ewing trial
RTx maybe used for spine & pelvis where resection is morbid VAC vincristine, dactinomycin & cyclophosphamide

Cx Mets lung, bone & bone marrow

Prog 5yr survival 2/3 Poorer spine & pelvis, >100 cm^3, poor response to chemo (<90% tumour cell necrosis)
↑ LOH & p53 mutn
Refer all cases to National Ewing's MDT

CHORDOMA

low signal T1, bright T2
Sacrum expanded

ging Not well visualised on AP pelvis due to overlying bowel & obliquity of sacrum. MRI lytic lesion with bony destruction ± calcification

ation Arises from remnants notochord ∴ occurs at ends of vertebral column: 50% sacral, 35% clivus of skull, 10% VB

es Pain, GI symptoms & nerve compression

to Vacuolated appearance called physaliferous cells

Tx Wide margin surgical resection + RTx if narrow margin
High rate local recurrence
To spare bladder function spare bilateral S2 or unilateral S2, 3 & 4