Haematological Malignancy in Orthopaedics

**Hodgkins Lymphoma**

- Peak incidence in early adulthood and elderly

- **Sx:**
  - Contiguous painless lymphadenopathy – often cervical
  - Hepatosplenomegaly and extra-lymphatic involvement (CNS, lung, skin, bone)
  - Pruritis, fever, night sweats, weight loss (25%)
  - Anaemia, neutrophilia, eosinophilia – predisposition to Herpes Zoster infection

- **Ix:**
  - ↑ ESR and LDH
  - Multinucleated Reed-Sternberg cell
  - bone marrow aspirate and trephine biopsy allows staging, along with CT/MRI

- **Staging:**
  - Stage 1 = single lymph node region or structure
  - Stage 2 = 2+ lymph node regions, on the same side of the diaphragm
  - Stage 3 = lymph node regions on either side of the diaphragm
  - Stage 4 = dissemination to other organs (liver, bone marrow, CNS)
  - A = no systemic sx ; B = systemic sx ; E = extralymphoid disease (lung, skin)

- **Mx:**
  - Radiotherapy alone for Stage 1A and 2A disease
  - Advanced disease requires combination chemotherapy (e.g. 6 cycles of ABVD)
  - Subsequent deep X-ray therapy is given for bulky disease

- **Lymphocyte predominant** → good prognosis
- **Lymphocyte depleted or systemic signs** → poor prognosis

- **Prognosis**
  - If achieve complete remission, for 1 year → good prognosis (possible cure)
  - Else use high dose therapy with stem cell rescue.
  - Stage 1 and 2 has a <90% cure
  - Stage 4 has a 50% 5 year survival

**Non Hodgkin’s Lymphoma**

- In bone, frequently called a reticulum cell sarcoma which resembles Ewing sarcoma
  - Distinguished on reticulin or silver staining by the presence of a fine mesh of matrix fibres
  - Lesions are often permeative which blend into normal bone architecture
  - A small cell tumour

- **Distinguish from Hodgkin’s disease by:**
  - Higher prevalence
  - Older mean age at dx
  - Non-contiguous, multi-centric spread
  - Extranodal involvement is more common
  - Associated with EBV (Burkitt’s lymphoma), HTLV-1 and HIV

- **Sx** = painless lymphadenopathy, and systemic symptoms (e.g. night sweats)
  - Pancytopaenia, hypogammaglobulinaemia. Paraprotein?
  - Bone pain and pathological fracture

- **Low grade** = indolent, treatable and becomes more aggressive with time
- **High grade NHL** = higher early mortality, but more amenable to Rx.

- **Variants:**
  - CLL
  - Burkitt’s lymphoma – jaw tumour in African children, associated with EBV
  - HTLV-1 related lymphoma – hypercalcaemia, skin involvement (Japan & Carribean)
  - Myeloma-like – bony disease with paraprotein, lymphadenopathy and splenomegaly. e.g. lymphoplasmacytoid lymphomas ( = Waldenström’s macroglobulinaemia )
  - GI lymphomas:
    - Mucosa associated lymphoid tissue (MALT) lymphomas
    - T-cell NHL complicating Coeliac’s disease

- **Mx:**
  - Deep X-ray therapy with 6 cycles of CHOP (± rituximab, R-CHOP)
  - Stem cell rescue
  - > 50% will relapse
Multiple Myeloma

- Monoclonal malignant proliferation of plasma (B) cells with effects on the bone through cell proliferation and expansion within the medullary canal, as well as osteoclast stimulation resulting in generalised osteopaenia and lytic lesions.

- Can affect any age, but predominantly elderly

- Plasma cells secrete:
  - IgG (± free light chain); 50%
  - IgA; 20%
  - Free light chain only; 20%

- Sx:
  - Bone destruction by humoral activation of osteoclasts, without normal osteoblasts
  - Cord compression or root compression from vertebral collapse
  - Diffuse osteopaenia seen on X-ray, or solitary punched-out lesions (= plasmacytomas)
  - Hypercalcaemia – bones, stones, abdominal groans and psychic moans, dehydration
  - Hyperviscosity:
    - paraprotein alters RBC surface causing aggregation
    - lethargy, confusion, ↓consciousness, thrombo-embolic disease
    - seen particularly in Waldenström's macroglobulinaemia (IgM secreting, r, with CHOP)
  - Renal failure 2° to:
    - nephrotoxic paraprotein
    - hypercalcaemia
    - hyperuricaemia (urate nephropathy)
    - infection
    - amyloid deposition
  - ↑ Susceptibility to infection, anaemia or pancytopenia
  - Bone pain; pathological fractures in spine/ribs
  - Weight loss

- I:
  - ↑ESR
  - Paraprotein band on immunoelectrophoresis
  - ↓ normal Ig
  - Osteopaenia or local bone destruction (particularly in phalanges)
  - ↑ Plasma cells in bone marrow aspirate
  - Bence-Jones proteinuria replaced by Serum PCR for Light Chains

- Mx:
  - IV fluids
  - Treat any infections
  - Analgesia for bone pain
  - Steroids or bisphosphonates
  - Internal fixation for fractures, with packing of cavities with methylmethacrylate cement
  - Plasmaphoresis to remove light chains and control viscosity
  - Oral melphalan (an alkylating chemotherapy agent)
  - Radiotherapy to control bone pain

- Poor prognosis with median survival only 2-3 years.

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1 A malignant mono-clonal gammopathy, with the clinical presentation similar to that of multiple myeloma except that (1) organomegaly is common in Waldenström macroglobulinemia and uncommon in myeloma, and (2) lytic bone lesions and renal disease are uncommon in Waldenström macroglobulinemia but common in myeloma.