Lymphoma

Hodgkin’s Disease

Definition
Usually a cancer of a germinal centre derived B cell, often with crippling mutations of rearranged Ig genes. (Occasionally germinal derived T cell with mutations of TCR genes.)

Epidemiology
<25% of all lymphoma. Young adults; second peak in middle age. Higher socio-economic groups with smaller family sizes.

Pathology
Usually a B cell lymphoma as above. Several distinct histological subtypes.
Nodular Sclerosing – classically mediastinal mass in young adults.

Develops in single lymph node group, commonly neck, mediastinum or groin and usually spreads “contiguously” untreated to others, on same and then other side of diaphragm, and spleen. Late spread is to liver, lung and bone marrow.

“B symptoms”: weight loss (>10%), night sweats, itching, alcohol induced pain.

Staging: Ann Arbor –
I One lymph node group
II Two lymph node groups on same side of diaphragm
III Nodes on both sides of diaphragm
IV Extra nodal spread

A/B Absence / Presence of B symptoms.

Treatment: Combination Chemotherapy / Involved Field Radiotherapy

Prognosis: 75% cure overall. Better in earlier stage. Some relapses curable (some with peripheral blood stem cell transplant).

Non- Hodgkin’s Lymphoma

“Low Grade”

**Definition**
Neoplasms of lymphocytes pursuing indolent courses, usually considered incurable under most circumstances.

**Incidence**
Increases with age. Male>Female.

**Pathology**
Several – majority are follicular lymphoma (B cell – CD20+)

Others: **B cell**:
- MALT lymphomas (especially gut associated – *H. pylori*) – good prognosis.
- Mantle cell lymphoma – “intermediate grade” – combines worst of low and high grade – most dead in 3 years.
- Small lymphocytic lymphoma – equivalent to CLL.

**T cell**:
Cutaneous lymphomas.

**Follicular**:
Often many lymph node groups at presentation. Bone marrow involvement also common. “B symptoms” less common.

Pathology: recapitulation of normal lymphoid follicle architecture with small lymphoid cells and variable numbers of larger, more blastic cells.

**Staging**: Ann Arbor will do. Also includes extranodal disease.

**Treatment**: Stage I: involved field radiotherapy will cure 30%
- Stage II: Watch and Wait; simple chemotherapy with Rituximab (MoAb against CD20) often repeated years later. Radiotherapy to bulk for symptom control. Palliation. Some stem cell transplants in younger patients.

**Prognosis**

“Incurable” – except some Stage I and some young patients with BMT. Median survival 10 years with inexorable course. However, frequently disease of elderly who “outlive” the disease.

May transform to high grade lymphoma (approx 3% per year) (clonal evolution of part of disease burden) – treat as high grade but do badly.
“High Grade”

**Definition**
Aggressive neoplasms of lymphocytes, some are curable.

**Incidence**
Increases with age. However, many occur in younger patients.

Incidence overall steadily increasing (particularly extranodal eg CNS), partly due to HIV, immunosuppression following solid organ transplantation.

Many present at an extranodal site eg gastric, skin (T>B), pulmonary.

**Pathology**
Majority B cell: Diffuse large B cell lymphoma (DLBCL) is 40% of all lymphomas. (CD20+). Also T cell and anaplastic large cell.

Often Stages I and II at presentation eg confined to chest or abdomen or neck. Bone marrow involvement in Stage IV commonest, also liver and lung. “B symptoms”.

**Pathology**: effacement of lymph node architecture with increased numbers of blast cells. Varied.

**Staging**: Ann Arbor will do. Also includes extranodal disease.

**Treatment**: Combination chemotherapy / radiotherapy
Chemotherapy with involved field radiotherapy for Stage I and to bulk in Stage II. Chemotherapy alone to Stages III and IV.
Cure 50% overall, better in earlier Stage and younger patients.

Addition of AntiCD20 monoclonal antibody to chemotherapy: “Rituximab (Mabthera)” to Stages II-IV for all patients, exact improvement in cure rates unclear – 10-15%?

Some patients under 60 cured at relapse by peripheral blood stem cell transplant.

**Prognosis**
Cure 50% overall. Those that fail to respond to salvage die quickly of a rapidly advancing disease.