# Lymphoma

## Disclaimer

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Red Flags

- Central nervous system (CNS) mass lesion or spinal cord compression
- Pericardial tamponade
- Superior or inferior vena cava obstruction
- Airway, intestinal or ureteric obstruction
- HIV

Background – About Lymphoma

- Lymphoma is a cancer of B or T lymphocytes, which develops in the lymph nodes, spleen, bone marrow, blood, or other organs, before eventually forming a tumour. It has many forms and diverse outcomes.
- It is the most common form of blood cancer in Australia and the sixth most common form of cancer overall.
- It may remain indolent (slow-growing low-grade cancers) and not require treatment for many years, or be rapidly progressive and may cause an acute emergency due to compression of vital internal structures.
- Is broadly divided into:
  - non-Hodgkin’s lymphoma (approximately 90%)
  - Hodgkin’s lymphoma (approximately 10%)
- Classification is complex and requires careful investigation and staging.
- Definitive diagnosis is made by adequate excisional lymph node biopsy and management is facilitated by appropriate imaging.
- If a non-Hodgkin’s lymphoma occurs in an HIV positive patient, it is an AIDS defining illness and some people may not be aware that they are at risk of HIV.
- Patients diagnosed with non-Hodgkin lymphoma should be screened for previously undiagnosed HIV.

Assessment

1. Suspect lymphoma if:
   - enlarged, usually painless lymph nodes (commonly in neck, axilla, or groin) for several weeks or more.
   - **B symptoms** are present.
     **B symptoms refer to systemic symptoms:**
     - Unexplained fever
     - Night sweats significant enough to need to change nightwear or sheets
     - Unintentional weight loss of more than 10% over 6 months
   - **other possible symptoms.**
     - Persistent fatigue
     - Flu-like illness
     - Abdominal pain, bloating or early satiety
     - Constipation or vomiting
     - Flank pain, haematuria
     - Recurrent infections
     - Bone pain
     - Back pain
2. Perform a physical **examination** with particular attention to lymph nodes, liver, and spleen.

**Examination**
- Check general features e.g. appearance of the patient, temperature, and other vital signs.
- Check for generalised lymphadenopathy by examining all accessible lymph node fields – suboccipital, supraclavicular, axillary, inguinal.
- Note the consistency of lymph nodes or lumps e.g. soft, firm, rubbery, or hard.
- If localised or regional lymphadenopathy, examine the lymphoid drainage area for a local lesion indicating infection or cancer.
- Check for oedema
- Perform cardiovascular and respiratory examination.
- Examine abdomen for hepatosplenomegaly, and ascites.
- Perform neurological examination if neurological symptoms are present. Back pain with upgoing plantars may indicate spinal cord compression.

3. Arrange **initial investigations**.
- FBE, ESR, electrolytes, urea, creatinine, liver function tests, lactate dehydrogenase (LDH), calcium, urate
- Serology for cytomegalovirus, Epstein-Barr virus, **hepatitis B, hepatitis C, HIV**
- Serum protein electrophoresis and immunoelectrophoresis, free light chains, and quantitative gamma globulins. (It is useful to arrange these as part of initial work-up as the turnaround time is approximately 2 weeks. They provide the haematologist with information about type of abnormality and the patient’s susceptibility to infection).
- CT chest, abdomen, and pelvis. In pregnant patients, or if concerned about radiation, consider chest X-ray and abdominal ultrasound scan.
- Chest X-ray for mediastinal imaging if CT is not an option
- Consider neck CT if obese patient (as palpation of lymph nodes may be difficult and may not be seen adequately on chest CT)

4. Consider a core biopsy of a lymph node via radiology if easily palpable.

5. Review **indicators for excision lymph node biopsy** (preferred for diagnosis).

**Indicators for excision lymph node biopsy**
- Significant constitutional symptoms
- Lymph nodes:
  - Greater than 2 cm diameter
  - Non-tender
  - Firm to hard texture
  - Swelling present several weeks
  - Multiple lymph nodes
- Abnormal chest X-ray or CT scan or abdominal ultrasound

**If indicators present:**
• contact by phone and refer urgently to a general surgeon,
• arrange coagulation screen, and peripheral blood flow cytometry (if lymphocytosis is present).

6. Arrange fine needle aspiration (FNA) biopsy via radiology for an abnormal cervical lymph node only in limited situations.

**Limited situations**
Moderate pre-test probability of a non-haematological malignancy e.g. older age, smoker, multiple skin cancers, or systems review suggests alternate diagnosis.

7. Consider differential diagnoses:
   • Infectious mononucleosis
   • Toxoplasmosis
   • Cytomegalovirus
   • HIV
   • Rubella
   • Hepatitis B, hepatitis C
   • Cat-scratch disease
   • Thymoma
   • Metastatic carcinoma
   • Tuberculosis (TB)
   • Sarcoidosis
   • Portal hypertension
   • Infiltrative disease of spleen
   • Extramedullary haematopoiesis
   • Myeloproliferative disease

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**Management**

**Initial management**

1. If signs of compression or obstruction present, arrange emergency department referral.

   **Signs of compression or obstruction**
   • Central nervous system (CNS) mass lesion or spinal cord compression
   • Pericardial tamponade
   • Superior or inferior vena cava obstruction
   • Airway, intestinal or ureteric obstruction

2. If HIV is detected, refer for HIV treatment and support services.

3. If excision biopsy is diagnostic or suggestive of lymphoma, refer for urgent or routine haematology referral within 2 weeks. If aggressive lymphoma (fast-growing high-grade cancer) with extensive lymphadenopathy, discuss with haematologist for consideration of urgent review or admission.

4. Use “watch and wait” management of indolent lymphomas (slow-growing low-grade cancers), if haematologist agrees that this is appropriate. See the patient every 1 or 2 months for the first 6 months, then every 3 to 6 months thereafter.
5. If FNA results indicate a non-haematological cancer, refer for urgent or routine oncology referral.

6. Consider travel and accommodation subsidies if the patient is travelling more than 100 km to receive care.

General practice follow-up of patients with indolent lymphoma

1. Review patient’s proposed shared-care follow-up plan which will be outlined in the patient’s multidisciplinary discharge summary from haematology oncology services.

   **Shared care for lymphoma**
   Patients with lymphoma are to be reviewed either by haematology or in general practice:
   - every 3 months for 3 years
   - every 6 months for a further 2 years (up to 5 years post treatment)
   - then annually

   **Multidisciplinary discharge summary**
   Most importantly should include what the patient has been told. This may include:
   - intentions.
   - goals and quantitative benefit of proposed treatment.
   - recurrence risk if known.

   Usually includes:
   - diagnostic tests performed and results.
   - tumour characteristics and other factors determining prognosis.
   - type and date of treatments and a treatment summary.
   - expectations of disease course, including expected discharge from oncology services.
   - interventions and treatment plans from other health professionals.
   - a process for rapid re-entry to specialist medical services for suspected recurrence.
   - a list of symptoms that might need prompt investigation.
   - a list of supportive care services provided and a plan for community care services, including what each service is to provide.
   - contact information for key care providers.

2. Refer for treatment if there is new development of obstructive or disfiguring lymphadenopathy, cytopenias, or the presence of B symptoms.

3. Recommend and review a symptom diary and encourage the patient to report any changes or concerns.

4. Check for enlarged lymph nodes and organomegaly.

5. Arrange regular investigations as advised by the haematologist:
   - FBE, ESR, electrolytes, urea, creatinine, LFT, lactate dehydrogenase, beta-2 microglobulin, and calcium, usually every 6 months or if symptomatic.
   - Quantitative immunoglobulins every 6 to 12 months. If low IgG is found, discuss with haematologist as these patients are at higher risk of infections.
• Imaging is not generally required unless specifically requested by treating specialist.

6. Ensure specific care for immunosuppression, as patients with low-grade lymphoma are relatively immune suppressed:
   • Offer immunisation and recall for:
     o annual flu vaccination.
     o pneumococcal vaccination.  
       Haematological malignancy is a Category A condition associated with the highest risk of invasive pneumococcal disease. Pneumococcal vaccination is recommended. The schedule is complex and depends on the patient’s age and previous pneumococcal vaccination history.  
       See the recommendations in Australian Immunisation Handbook – Pneumococcal Disease for further details.

   • Do not immunise with live vaccines (e.g. Zostavax). In immunocompromised patients there is a risk of adverse events or vaccine-related disease due to unchecked infection (replication) of the vaccine virus or bacteria.

   **Live vaccines include:**
   o Bacille Calmette–Guérin (BCG)
   o Japanese encephalitis
   o Measles, mumps, rubella (MMR)
   o Rotavirus
   o Oral typhoid
   o Varicella
   o Yellow fever
   o Zoster – Zostavax is contraindicated in haematologic malignancy:
     ▪ Zostavax contains live attenuated varicella-zoster virus, which contains 14 times more virus than childhood varicella vaccines.
     ▪ Administration to immunocompromised patients is associated with risk of disseminated disease from the vaccine virus. This includes haematological or generalised malignancies (including those not on treatment) e.g. lymphoma, acute or chronic leukaemia, Hodgkin’s disease.

7. Screen for other malignancies.  
   Lymphoma survivors have an increased risk of other malignancies:
   • Arrange annual skin checks.
   • Limit unprotected sun exposure.
   • Consider age-appropriate screening for cervical, breast, prostate, and bowel.

8. Treat intercurrent infections. Advise the patient to present early if any painful rash (possible shingles), or any sign of infection. Consider using appropriate antibiotics for bacterial infections in patients with low immunoglobulin levels.

9. Encourage healthy diet, exercise, relaxation, reduction of alcohol, and cessation of smoking.

10. Consider referral for appropriate support services:
    • Counselling
    • Allied health
    • Mental health
    • Leukaemia Foundation support services
Referral

- If **signs of compression or obstruction** present, arrange **emergency department referral**.
- If **HIV** is detected, refer for **HIV treatment and support services**.
- If excision biopsy is diagnostic or suggestive of lymphoma, refer for **urgent or routine haematology referral** within 2 weeks. If aggressive lymphoma (fast-growing high-grade cancer) with extensive lymphadenopathy, discuss with **haematologist** for consideration of urgent review or admission.
- If FNA results indicate a non-haematological cancer, refer for **urgent or routine oncology referral**.
- If low IgG is found on follow-up, discuss with **haematologist** as these patients are at higher risk of infections.
- Consider referral for appropriate support services:
  - **Counselling**
  - **Allied health**
  - **Mental health**
  - **Leukaemia Foundation support services**

Information

For health professionals

Further information
Cancer Council – [Optimal Cancer Care Pathway for People with Hodgkin and Diffuse Large B-cell Lymphomas](#)

For patients

- Leukaemia Foundation:
  - [Lymphoma Explained](#)
  - [Watch and Wait](#)
- Cancer Council:
  - [Lymphoma](#)
  - [Lymphoma Pathway](#)

Disclaimer

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