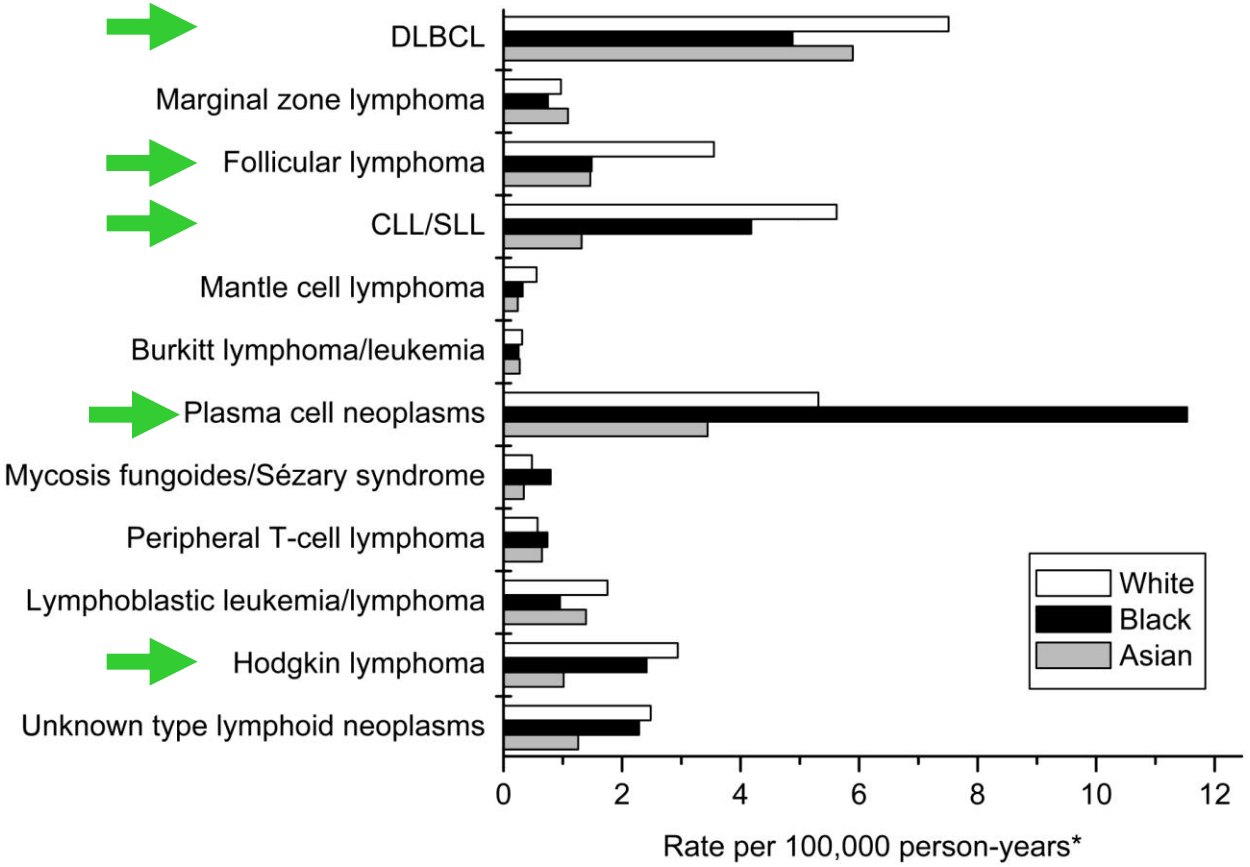
Quadro di presentazione generale dei linfomi

Linfoadenopatie in 2/3 dei casi	**1.Adenopatia persistente (> 1-2 cm per >4 settimane)** **2.Non dolente** **3.Andamento del volume delle adenopatie fluttuante nelle forme indolenti**	**Da differenziare** **1.Adenopatie infettive** **2.(Batteri, Mononucleosi CMV, HIV,Toxoplasmosi, Istoplasmosi)** **3.Sarcoidosi (mediastino)** **4.Adenopatie metastatiche**
Sintomi correlati a linfoadenopatie	**Mediastino** **Addome** **Extranodali**	**1.Tosse persistente** **2.Senso di fastidio retrosternale** **3.Sindrome mediastinica** **4.Senso di gonfiore, dolore,** **5.Splenomegalia** **6.Linfedema** **7.SNC, midollo spinale** **8.Gastrico** **9.Orbita** **10.ossa** **11.Testicolo** **12.Cute**
Sintomi sistemici	**20% circa dei casi**	**Soprattautto negli alti gradi e negli stadi avanzati**

blood
JOURNAL OF THE AMERICAN SOCIETY OF HEMATOLOGY

Incidence of lymphoid neoplasms by subtype and race, 12 SEER registries, 1992-2001



Morton, L. M. et al. Blood 2006;107:265-276

Causative factors

Virus	
EBV	Burkitt's + others
HTLV1	Adult T-cell leukemia/lymphoma
HCV	Indolent B-cell lymphoma
HHV-6	Angioimmunoblastic lymphadenopathy (variety of T-cell NHL / Hodgkin's disease (rare)
HHV-8	Body cavity Lymphoma (rare B-NHL)
Bacteria	
Helicobacter pylori	Mucosa associated lymphoid tissue (MALT) lymphoma (variety of marginal zone B-cell lymphoma)
Chlamydia psittaci	Orbit lymphoma

Types of non Hodgkin's lymphoma

- Clinically Indolent / clinically aggressive

(slow growth= low grade lymphoma

(rapid growth and invasiveness = high grade lymphoma)

- B-cell / T-cell
  (immunophenotype)

- Histopathologic types

Pattern of growth recalling primarily involved lymph node structure. (i.e. mantle zone, germinal centre, marginal zone)

Morphology and immunophenotype of the neoplastic cells; pattern of growth in the lymph node

Most frequent types of non Hodgkin's lymphoma

Clinically Indolent

- B-cell type

- Follicular lymphoma
- Marginal zone B-cell lymphoma → *extranodal (gastric)*, *Splenic*, *nodal*
- Small lymphocytic lymphoma/CLL
- Lymphoplasmacytic lymphoma (Waldenstrom macroglobulinemia)

- T-cell type

- Peripheral T-cell lymphoma (some)
- Mycosis fungoides
- LGL expansion (T or NK)

Most frequent types of non Hodgkin's lymphoma

Clinically aggressive

- B-cell type

- **Diffuse large B-cell lymphoma**
- **Mantle cell lymphoma**
- **Anaplastic large cell lymphoma**
- **Burkitt's lymphoma**
- **Lymphoblastic lymphoma**

- T-cell type

- **Anaplastic large cell lymphoma (ALK+)**
- **Peripheral T-cell lymphoma (some)**
- **Sezary's syndrome**
- **T- Prolymphocytic leukemia**
- **Lymphoblastic lymphoma**
- **Angioimmunoblastic lymphoma**

Presentation picture and diagnosis

- Sistemic symptoms

B symptoms in Ann Arbor staging system: unexplained fever > 38°C; weight loss >10% body weight over 6 months, night sweats = pruritus

other sistemic: pruritus

- Tumor-related symptoms

- Superficial adenopathy > 1cm for more than 4 weeks (wax and wane in low-grade lymphomas)
- Thorax (cough, discomfort, superior vena cava syndrome)
- Abdomen (chronic pain, early satiety, left quadrant discomfort, jaundice, intestinal symptoms)
- Lymphedema
- Extra-nodal (depending on tissue involved)

Diagnosis

- Biopsy of any lymph node enlargement > 1-2 cm for > 4 weeks without an obvious explanation
- Imaging techniques according to symptoms
- No blood test is specific for NHL

Essentials for Diagnosis and Staging

- Histopathology: histologic type allows for the identification of distinct clinical behaviour: low grade lymphoma vs high grade or indolent vs aggressive lymphoma
  Each entity deserve different treatment
- Visit with documentation of systemic (B) symptoms
- CT scan (thorax and abdomen)
- CNS study in special subtype (i.e. Burkitt's lymphoma) or in symptomatic patients
- Bone biopsy (BM involvement) + Complete blood count (possible leukemic involvement)
- Liver +renal function, uric acid, LDH, calcium beta-2-microglobulin, electrophoresis

Table 77-1 Ann Arbor Staging System

I—involving a single lymph node region (stage I) or a single extralymphatic organ or site (stage IE)

II—two or more involved lymph node regions on the same side of the diaphragm (stage II) or localized involvement of an extralymphatic organ or site (stage IIE)

III—lymph node involvement on both sides of the diaphragm (stage III), or localized involvement of an extralymphatic organ or site (stage IIIE), or spleen (stage IIIS), or both (stage IIIES)

IV—refers to the presence of diffuse or disseminated involvement of one or more extralymphatic organs (e.g., liver, bone marrow, lung), with or without associated lymph node involvement

The presence or absence of systemic symptoms should be noted with each stage designation; A for asymptomatic; B for presence of fever, sweats, or weight loss >10% of body weight.

Entità clinicopatologica	Presentazione	Evoluzione istologica e clinica e/o commenti
Linfomi indolenti		
a) Linfoma linfocitico	Frequentissimo coinvolgimento ematico (LLC)	Linfoma ad alto grado (s Richter)
b) linfoma marginale	- Variante extranodale (MALT) Linfomi gastrici, bronchiali, gh. salivari - Variante linfonodale - Variante splenica (linfoma marginale splenici con o senza linfocicti villosi circolanti)	Linfoma malt con componente ad alto grado (grandi cellule) Linfoma marginale ad alto grado
c) linfoma centrofollicolare	Malattia frequentemente disseminata	Linfoma alto grado a grandi cellule (p53, p16) (avviene nel 5-10% dei casi per anno)
Linfomi aggressivi		
a) linfoma del mantello	Frequente iniziale coinvolgimento BM e PB Malattia spesso disseminata (sedi linfonodali ed extranodali, gastrointestinali), con splenomegalia e leucemizzazione	Trasformazione in linfoma mantellare blastoide
b) linfoma B diffuso a grandi cellule	Crescita rapida ed invasiva (compressione vasi sanguigni, nervi, bronchi, ossa)	Tenere presente possibile estensione tardiva al SNC
c) linfoma a grandi cellule con sclerosi del mediastino	Localmente invasivo (mediastino)	Se buona risposta alla terapia (CTx + RT) remissioni durature
d) linfomi T periferici	Svariate entità di malattia	
e) linfoma anaplastico CD30+	primitivo interessamento cutaneo malattia disseminata	Buona risposta alla terapia