

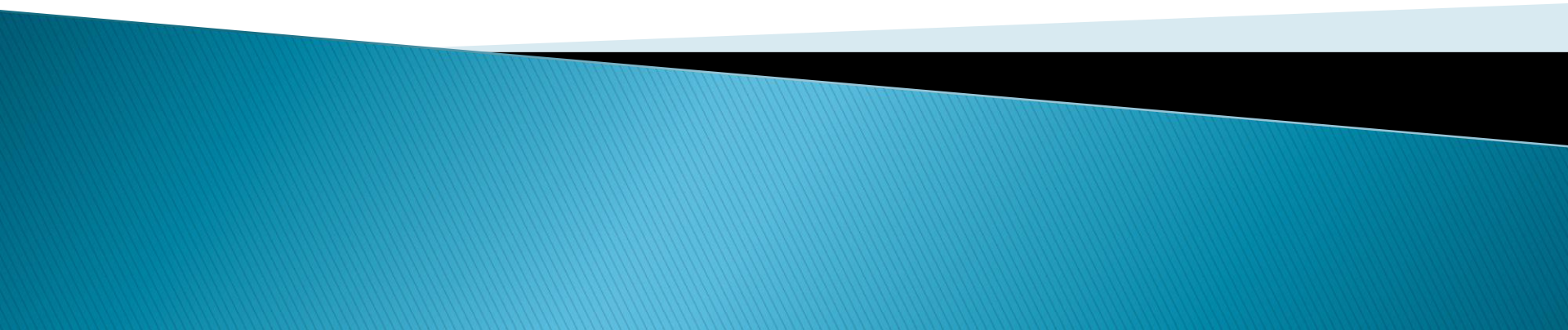
Ovarian lymphoma

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Burkitt lymphoma



Definition

- Highly aggressive lymphoid neoplasm but curable
 - often presentation in extra nodal sites or as an acute leukemia
 - Usually with a demonstrable MYC gene translocation to an IG locus
 - A combination of several diagnostic techniques is necessary for diagnosis
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Epidemiology

- ▶ **Three variants:**
 - ▶ Endemic
 - ▶ Sporadic
 - ▶ Immunodeficiency-associated

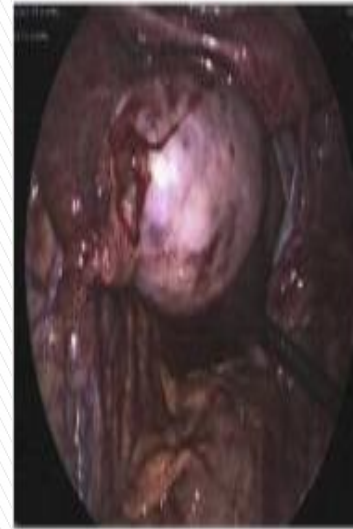
Endemic

- Equatorial Africa & Papua New Guinea with distribution that overlap with regions endemic for malaria
- The most common childhood malignancy
- peak 4–7 Y.O
- M to F ratio is 2:1
- EBV in more than 95%
- Jaws & other facial bones are most common
- Distal ileum , caecum , omentum , gonads , kidney , breast

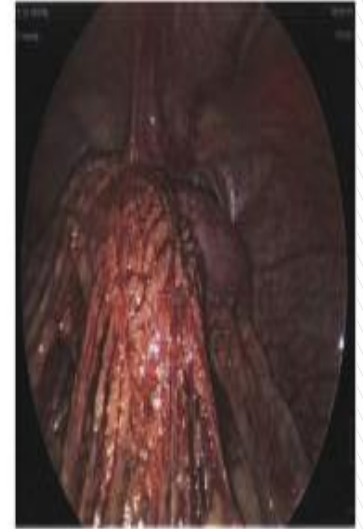


Sporadic

- Throughout the world
- Mainly children & young adult
- M to F ratio is 2-3 :1
- EBV in 20-30%
- Abdominal masses are most common(ileocecal),Ovaries ,kidney &breast
- Facial structures in particular jaws are very rare
- lymph node presentation is unusual but more common in adult than in children



(A)

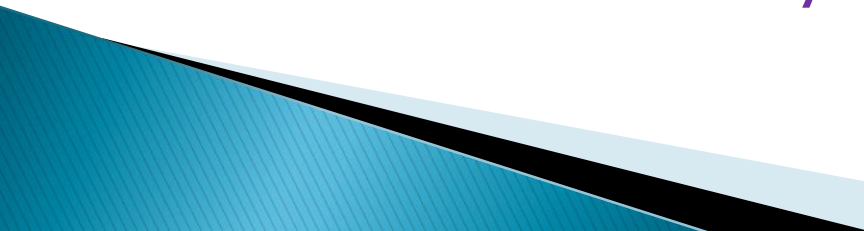


(B)

Immunodeficiency associated

- ▶ More common in HIV
- ▶ Often occurs as the initial manifestation of the disease
- ▶ EBV in 25–40%
- ▶ Nodal localization & bone marrow involvement are frequent

Burkitt lymphoma

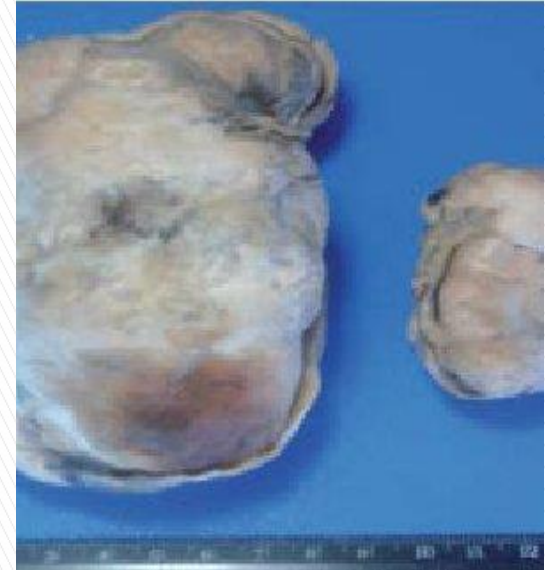
- ▶ In all 3 variants patients are at risk for **CNS involvement**
 - ▶ Patients often present with **bulky disease**
 - ▶ Specific manifestation at presentation may vary in variants
 - ▶ Involvement of BM or presentation as acute leukemia is uncommon in endemic BL
 - ▶ Nodal involvement is rare in endemic & sporadic BL , but more frequent in immunodeficiency-associated BL
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Burkitt leukemia variant

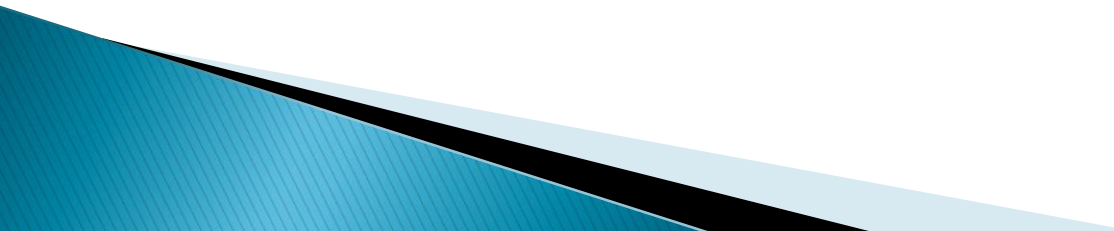
- ▶ A leukemic phase can be observed in patients with bulky disease ,but only rare cases ,typically in males ,present purely as leukemia with peripheral blood & bone marrow involvement

Macroscopy:

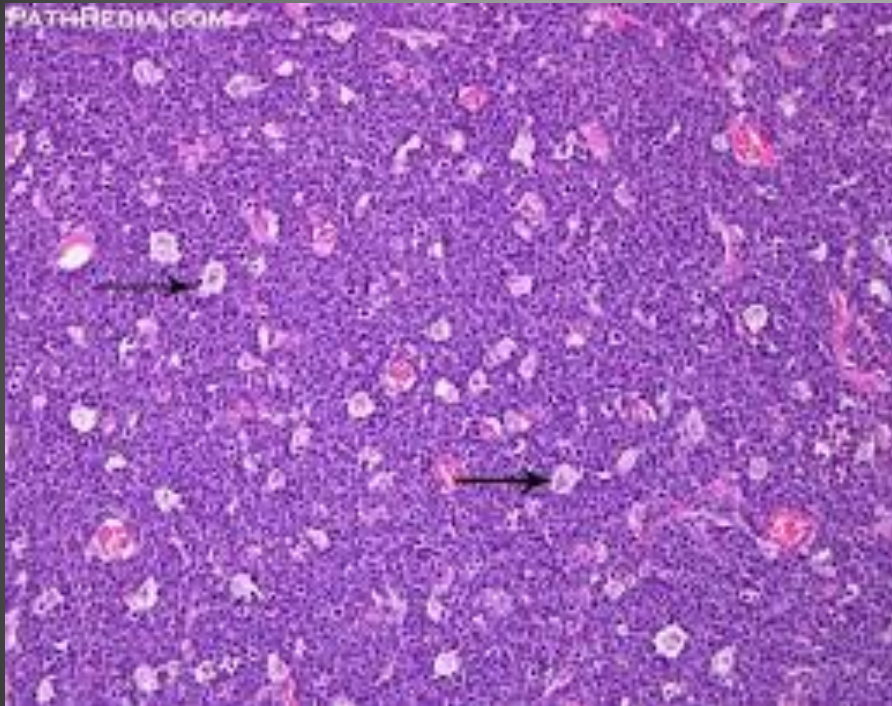
- Involved organs are replaced by masses with a fish-flesh appearance often associated with haemorrhage & necrosis
- Adjacent organs or tissue are compressed and/or infiltrated



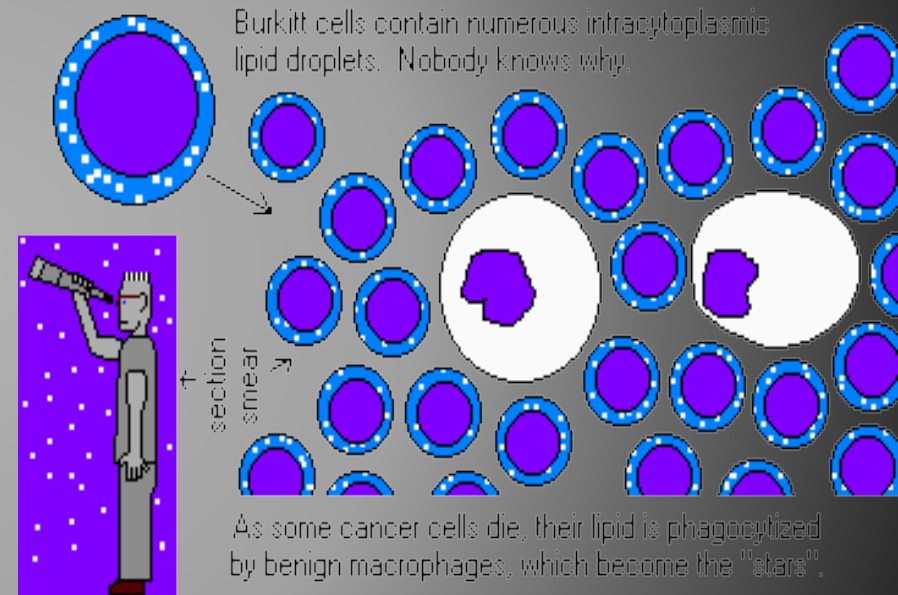
Microscopy:

- ▶ Tumor cells are medium sized with diffuse monotonous growth pattern
 - ▶ Nuclei :Round , finely clumped chromatin
 - ▶ Nucleoli :Multiple basophilic medium – sized
 - ▶ Cytoplasm : Deeply basophilic ,usually lipid vacuole
 - ▶ Extremely high proliferation rate with mitotic figures & apoptotic cells
 - ▶ Usually Starry sky (tingible body macrophages)
 - ▶ Florid granulomatous reaction in some cases (limited stage & especially good prognosis)
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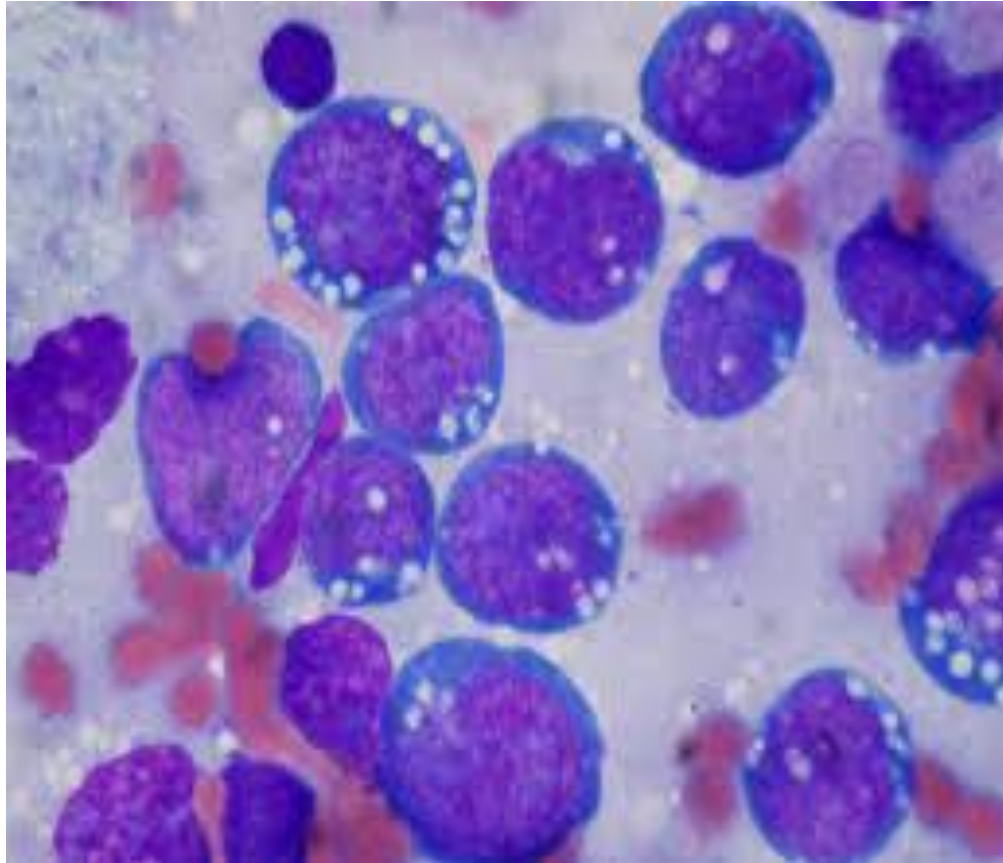
Starry sky



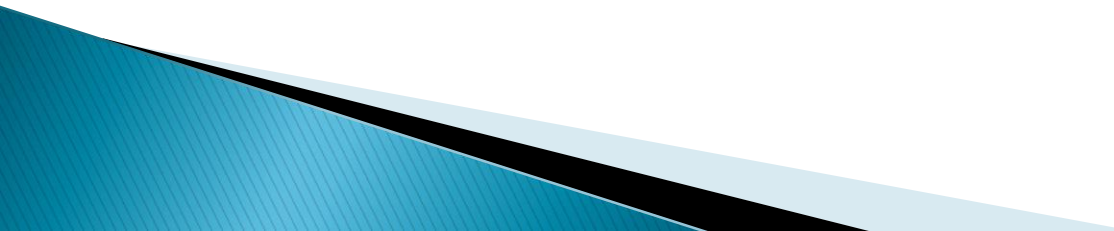
Burkitt's Lymphoma: Starry Sky



Lipid vacuoles



Immunophenotype

- ▶ Mod to strong membrane IgM with light chain restriction
 - ▶ B cell antigens (CD19 ,CD 20 ,CD 22 ,CD 79a & PAX5)
 - ▶ Germinal center markers (CD 10 & BCL 6)
 - ▶ CD38 ,CD77 & CD43 are frequently positive
 - ▶ Almost all BLs have strong **MYC** in most cells
 - ▶ **Proliferation rate is very high , with nearly 100% of the cells positive for Ki67**
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Immunophenotype

- ▶ Very few infiltrating T cells
- ▶ Neoplastic cells are usually negative for CD5 ,CD23 ,CD138 ,BCL2 & TdT
- ▶ The immunophenotype maybe more variable in sporadic BLs in older patients.High BCL2 expression should suggest the presence of an additional BCL2 breakpoint consistent with high grade B cell lymphoma with MYC and BCL2 and/or BCL6 rearrangements

Cytogenetic abnormalities

1. t(8,14)

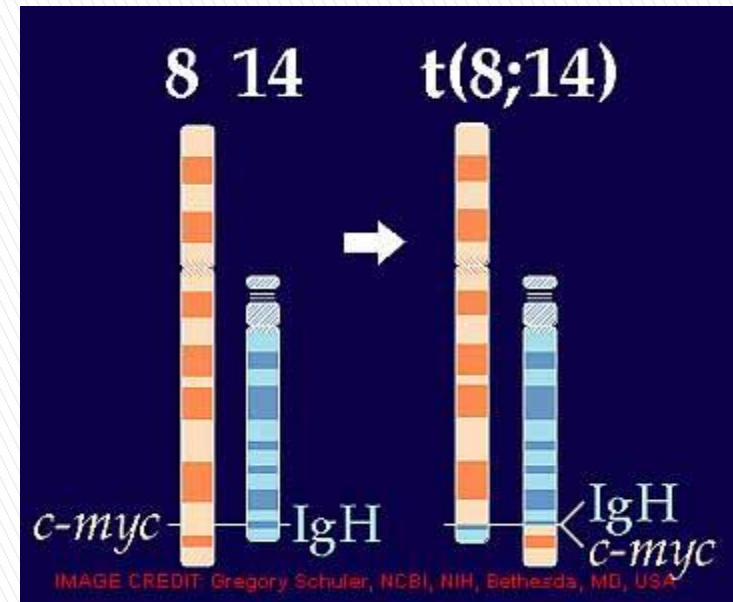
2. t(2,8)

3. t(8,22)

–MYC translocations are not specific for BL ,& may occur in other types of lymphoma(Transformed follicular lymphoma ,rare cases of DLBCL , Double hit lymphoma)

–Other chromosomal abnormalities may also occur in BL

–10%of classic BL ,lack an identifiable MYC rearrangement



Prognosis

- ▶ Highly aggressive but potentially curable
- ▶ With intensive chemotherapy :long-term survival in 70–90% cases
- ▶ Adverse prognostic factors:
 - ▶ 1–advanced stage
 - ▶ 2–BM & CNS involvement
 - ▶ 3–unresected tumor >10cm
 - ▶ 4–high LDH
- ▶ Relapse :usually within first year after diagnosis

Burkitt-like lymphoma with 11q aberration

- ▶ A subset of lymphoma that resemble BL morphologically, to a large extent phenotypically but the **lack MYC rearrangement**. Instead, they have a chromosome 11q alteration
 - ▶ They have more pleomorphism, occasionally a follicular pattern, & frequently a nodal presentation
 - ▶ Similar cases reported in post-transplant setting
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