VII CORSO del GRUPPO ITALIANO DI EMATOPATOLOGIA

La **DIAGNOSTICA EMATOPATOLOGICA** nell'**ERA** della **MEDICINA** di **PRECISIONE**

Unexpected onset in CLL

Ludovica Borgia

U.O.C. Anatomia Patologica
Ospedale Civile Spirito Santo – Pescara

asl pescara

Bologna, Aula Prodi | 11-12 maggio 2026

- 65-year-old
- Male
- Clinical History: arterial hypertension and skin tumors

April 2025
The patient presents with a right submandibular swelling.

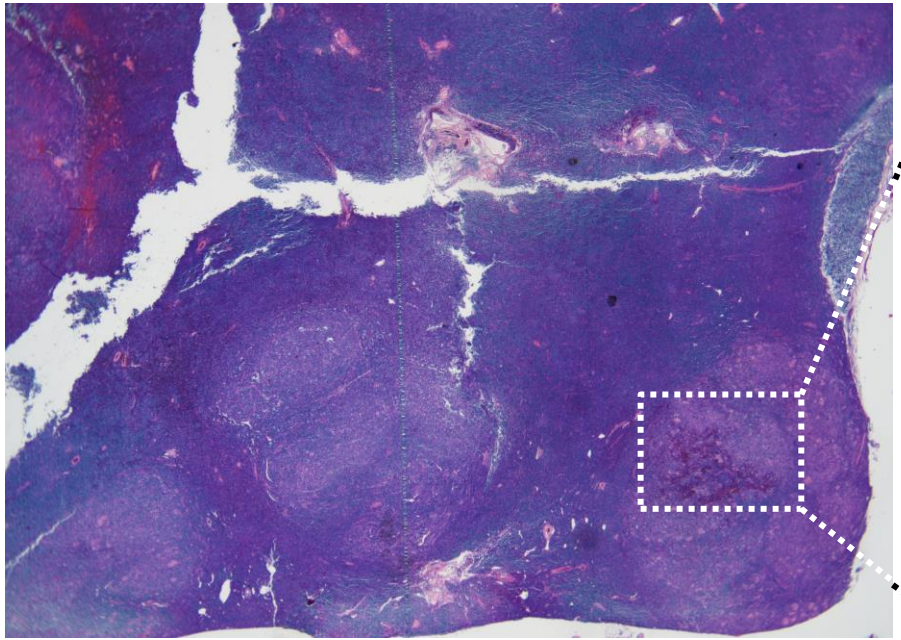
Blood Cells Count
WBC 10,000 (Ly 6,500), Hb 15, PLT 180.000

No B symptoms

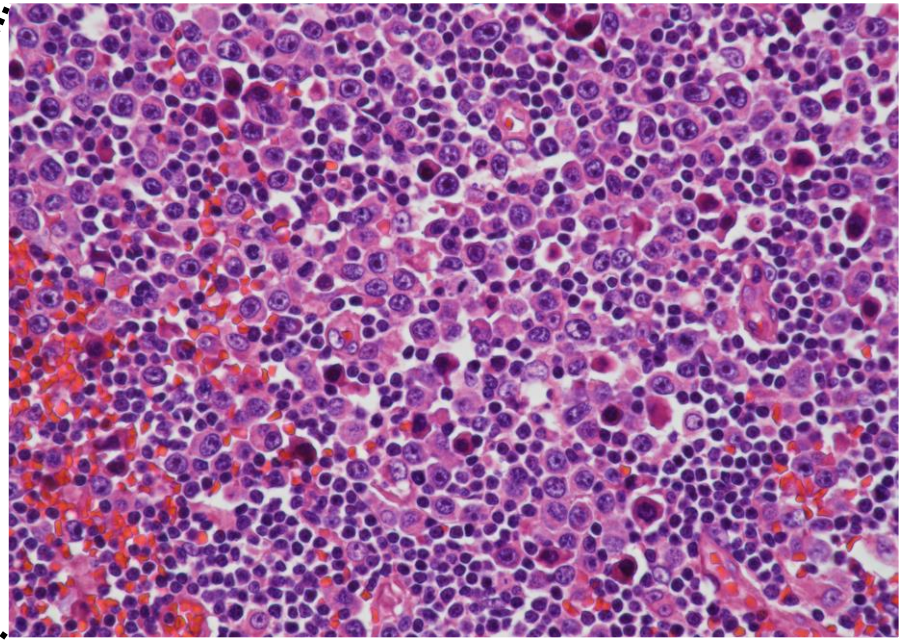
Neck CT scan
Diffuse, bilateral lymphadenopathy, with a 4.5x3.5 cm laterocervical lymph node cluster.

July 2025

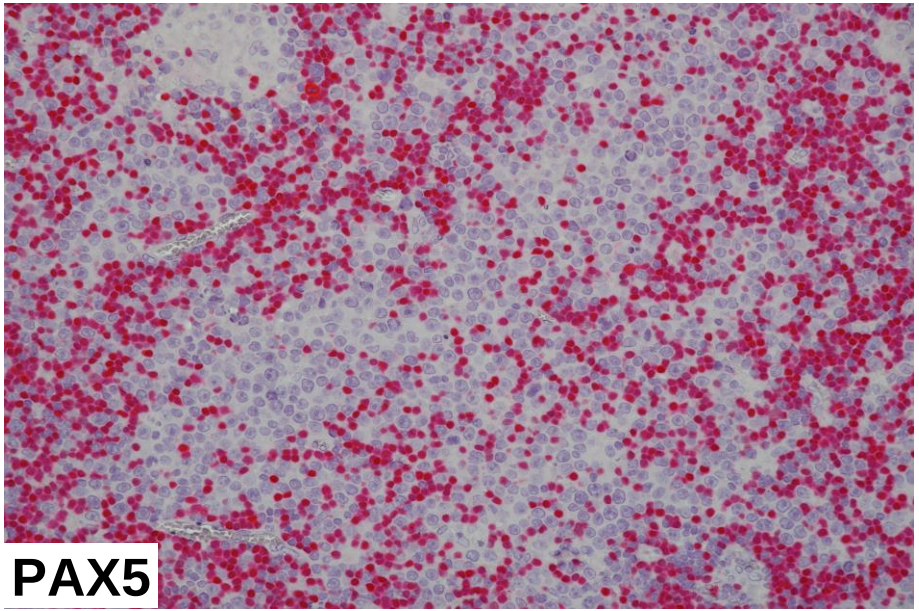
Submandibular lymph node excisional biopsy (3 cm)



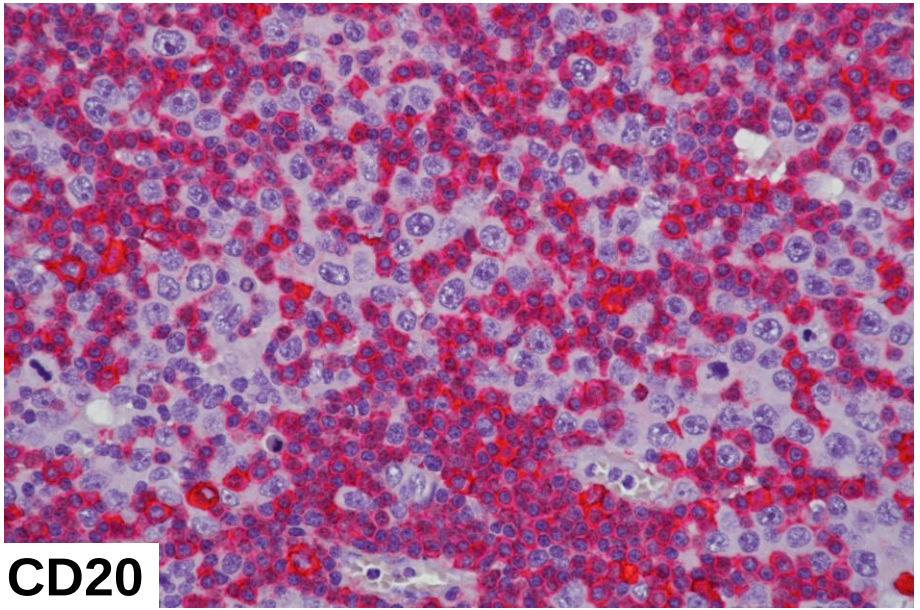
4x



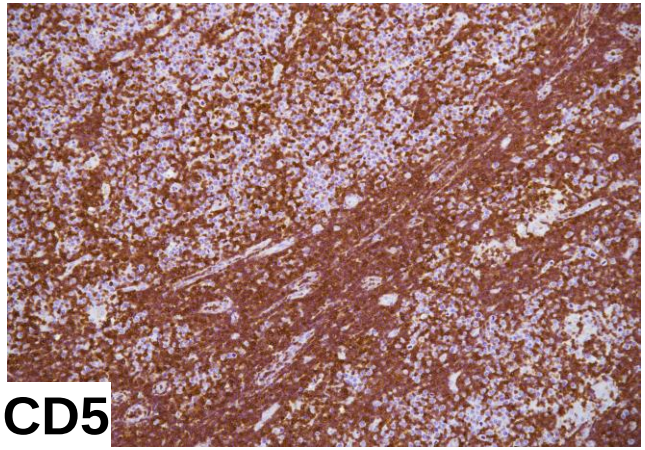
40x



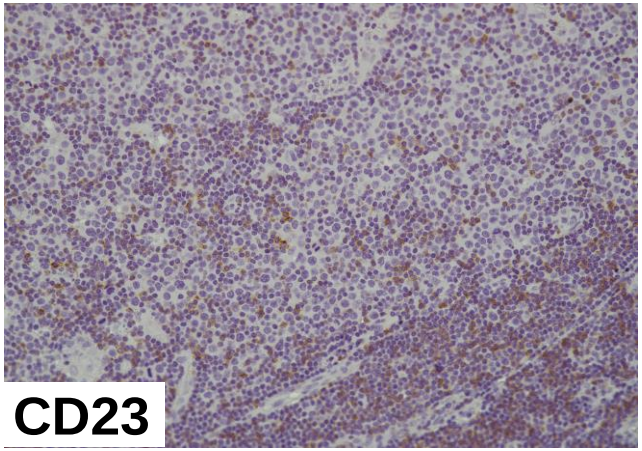
PAX5



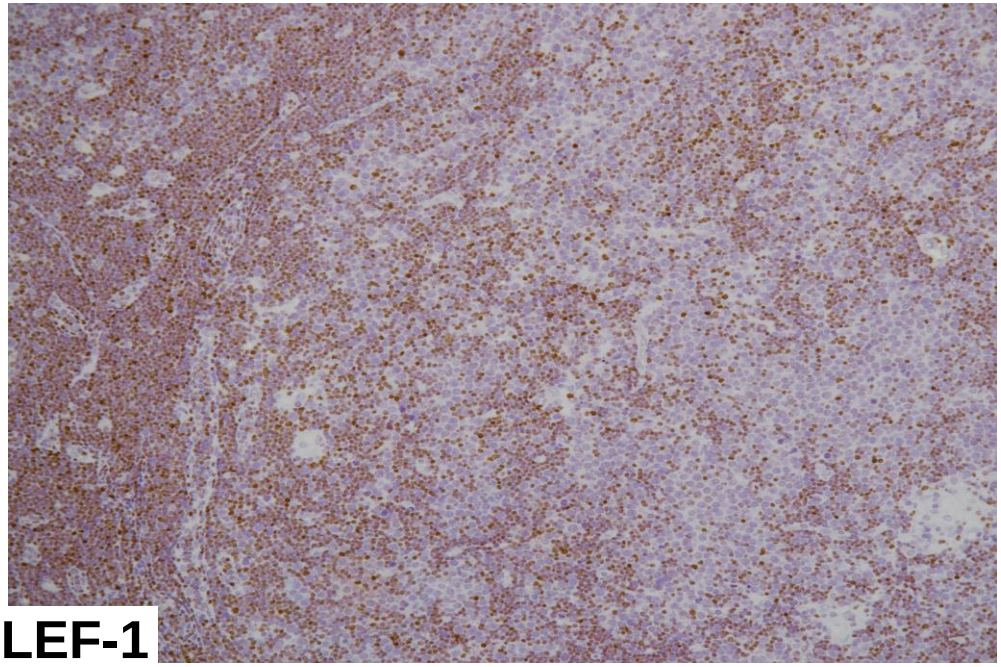
CD20



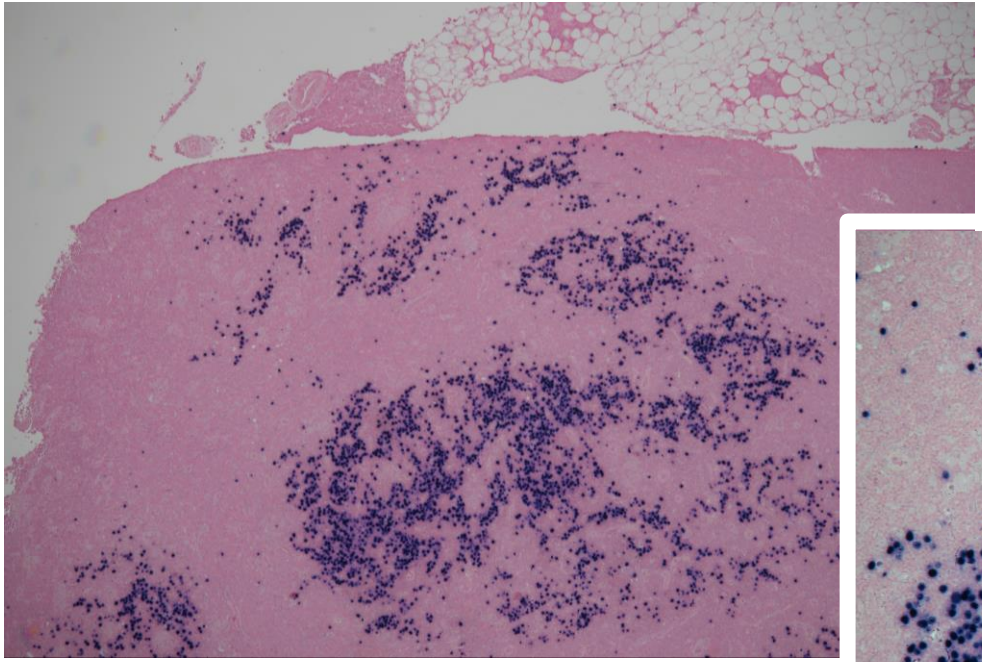
CD5

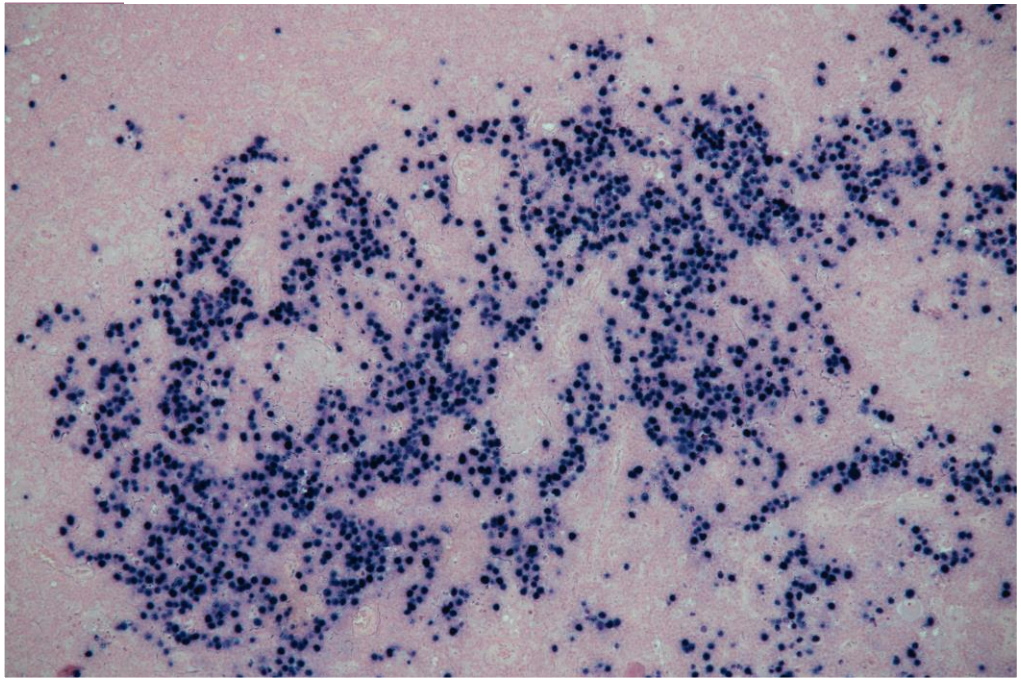


CD23

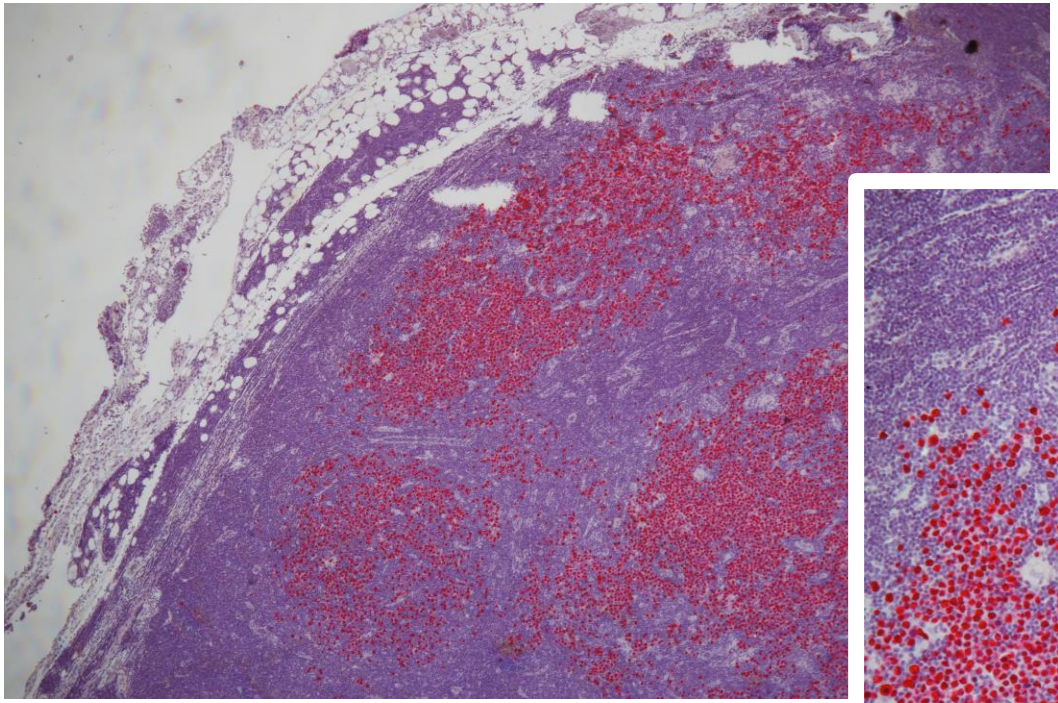


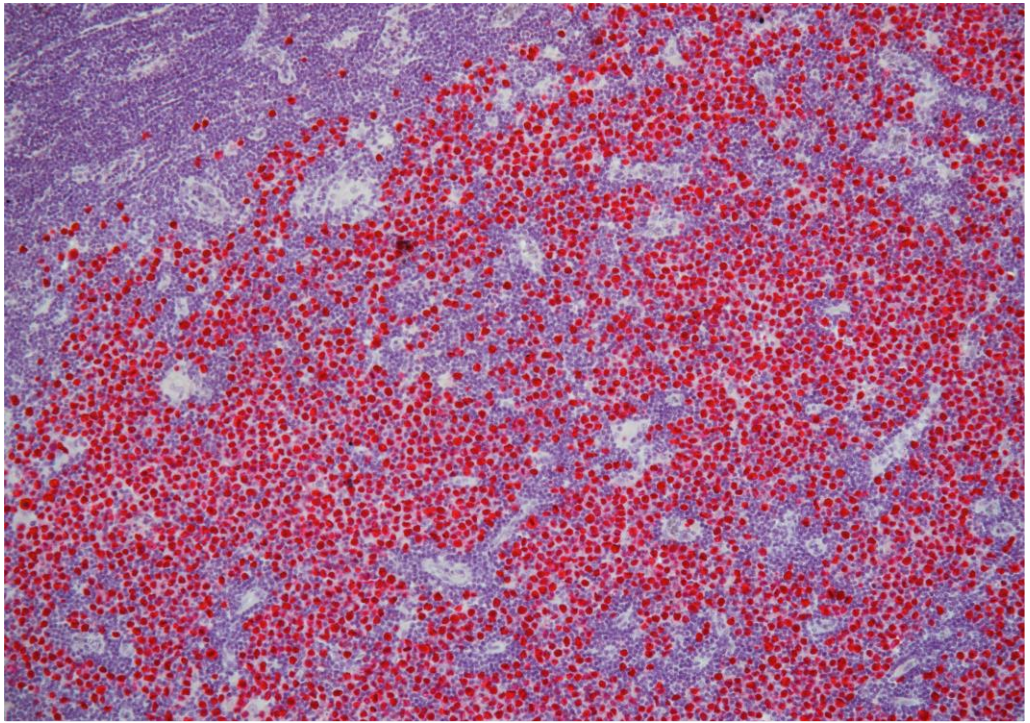
LEF-1



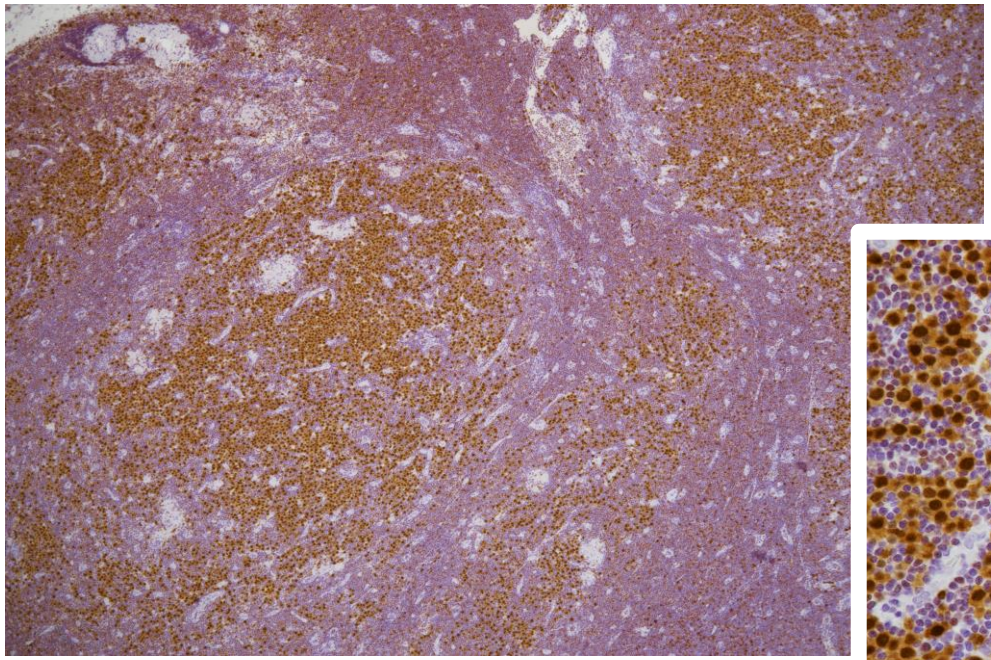


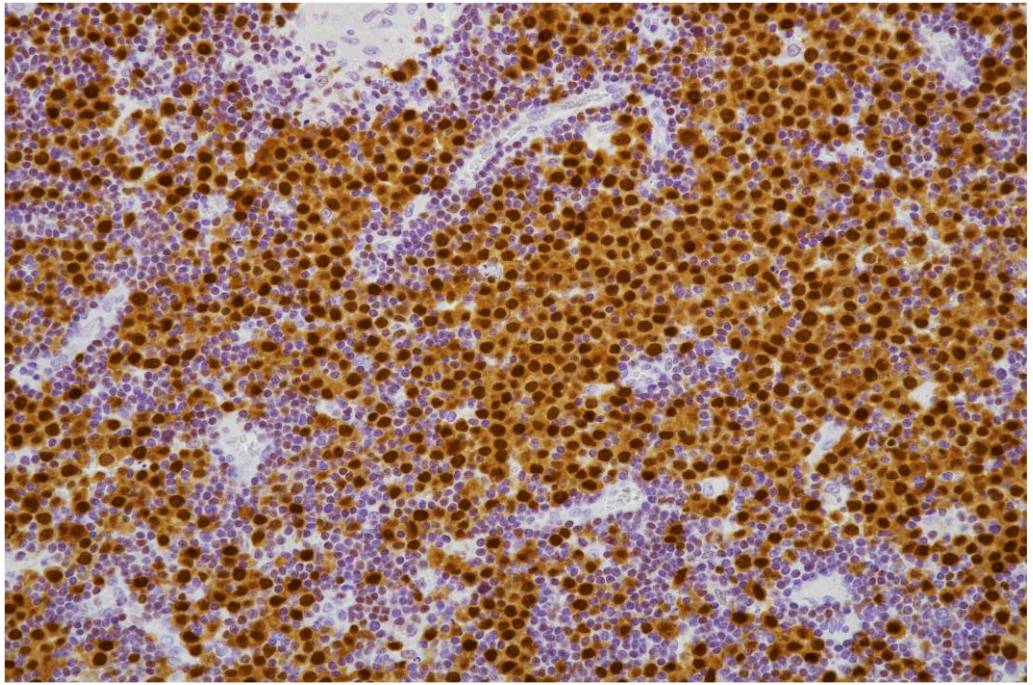
EBER ISH



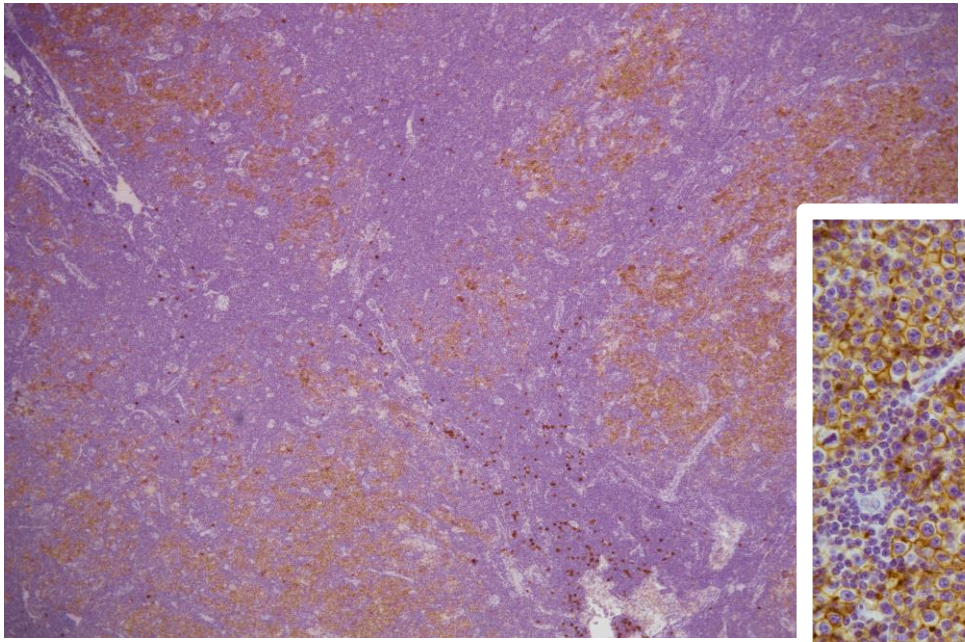


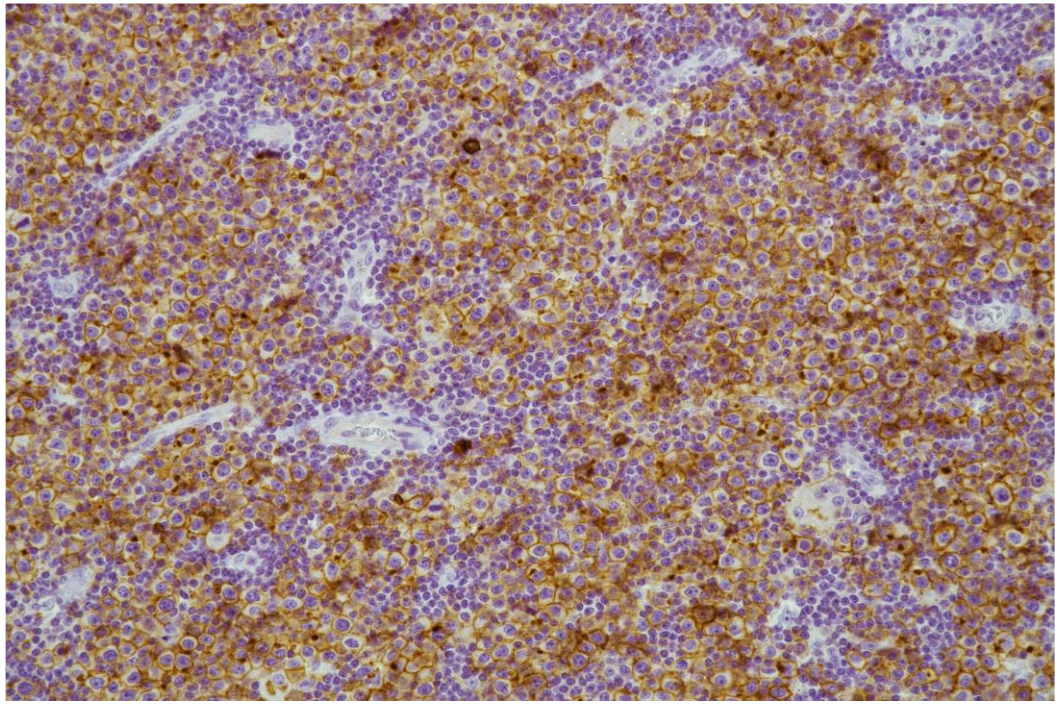
HHV8



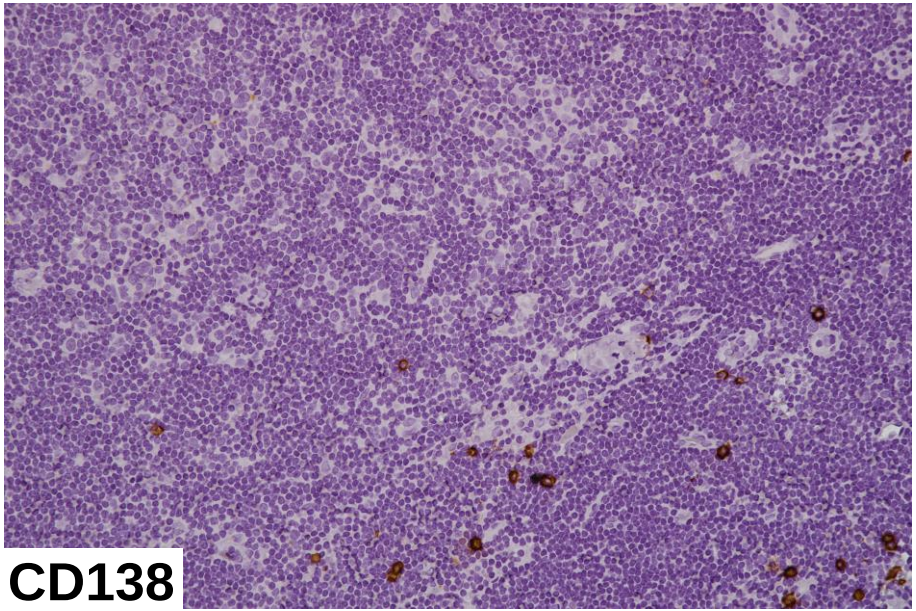


MUM1

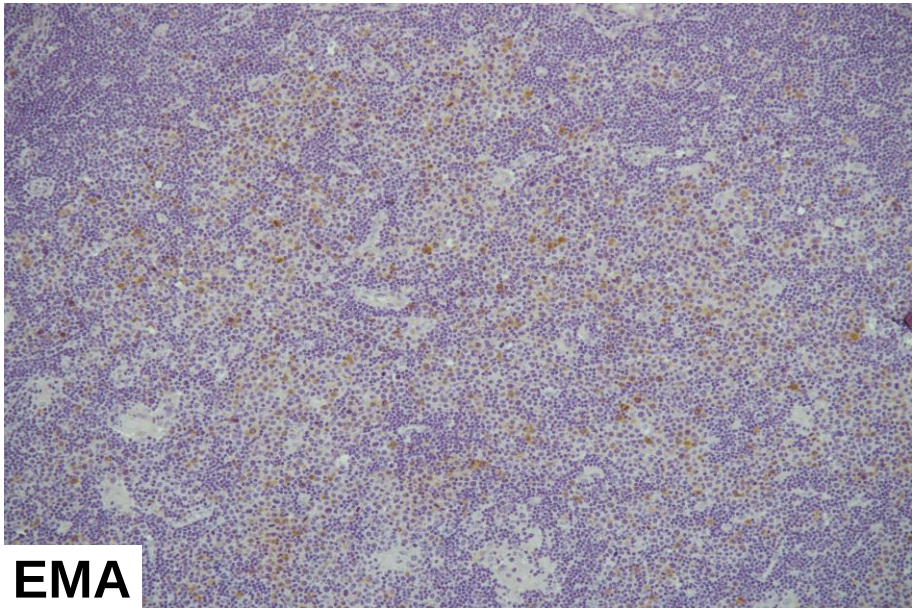




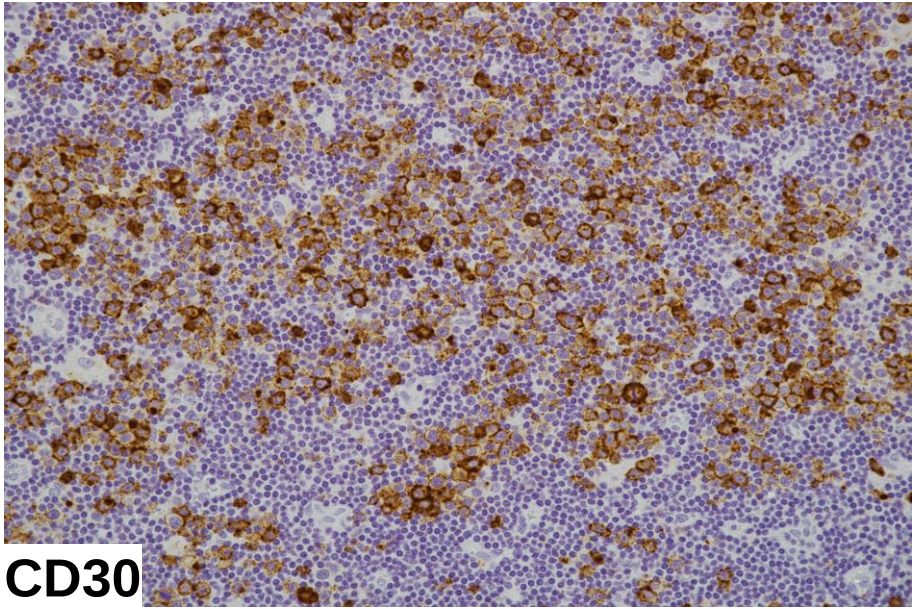
CD38



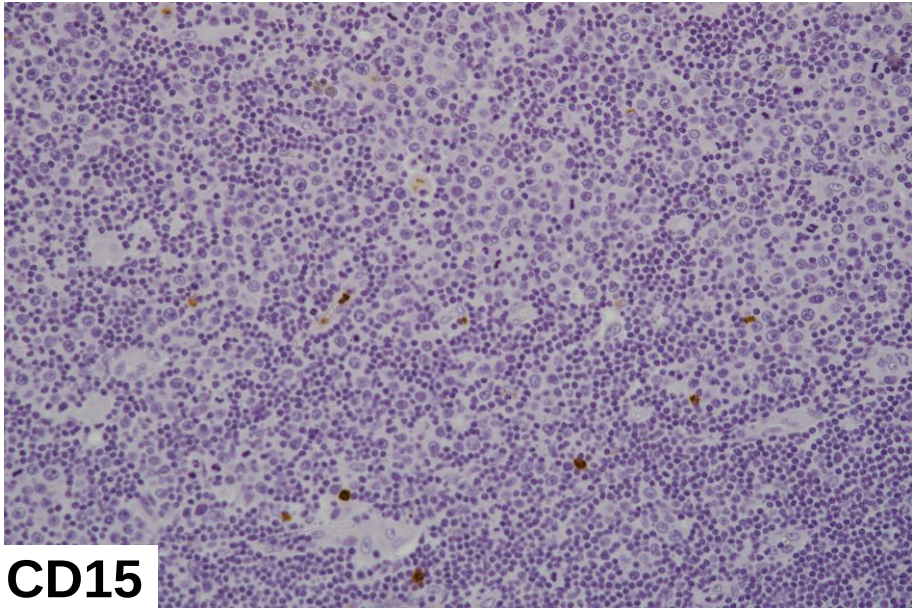
CD138



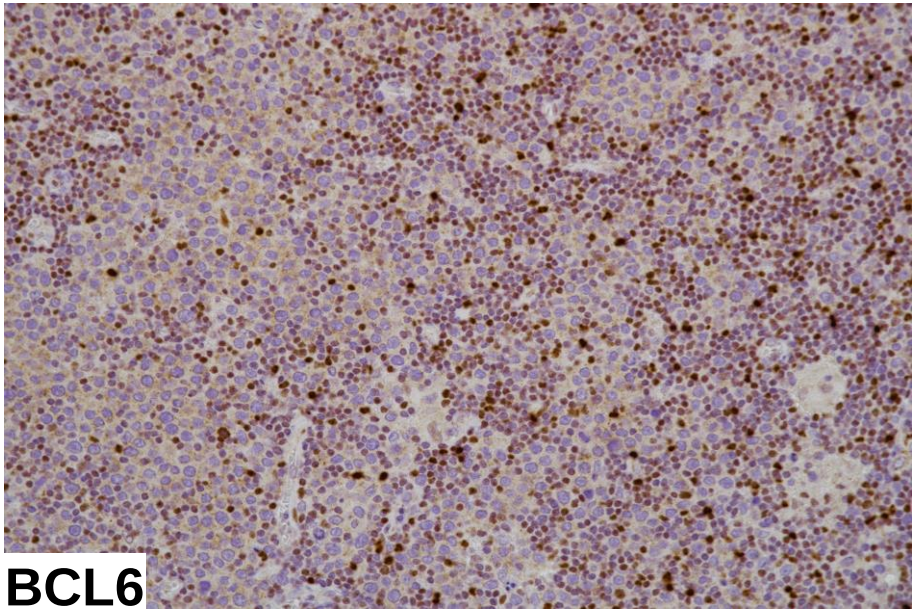
EMA



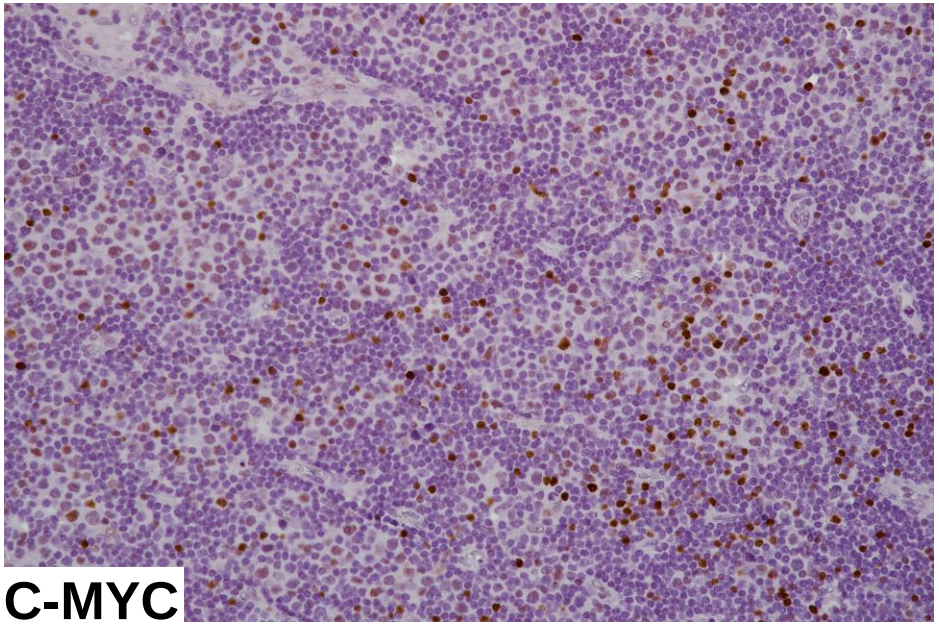
CD30



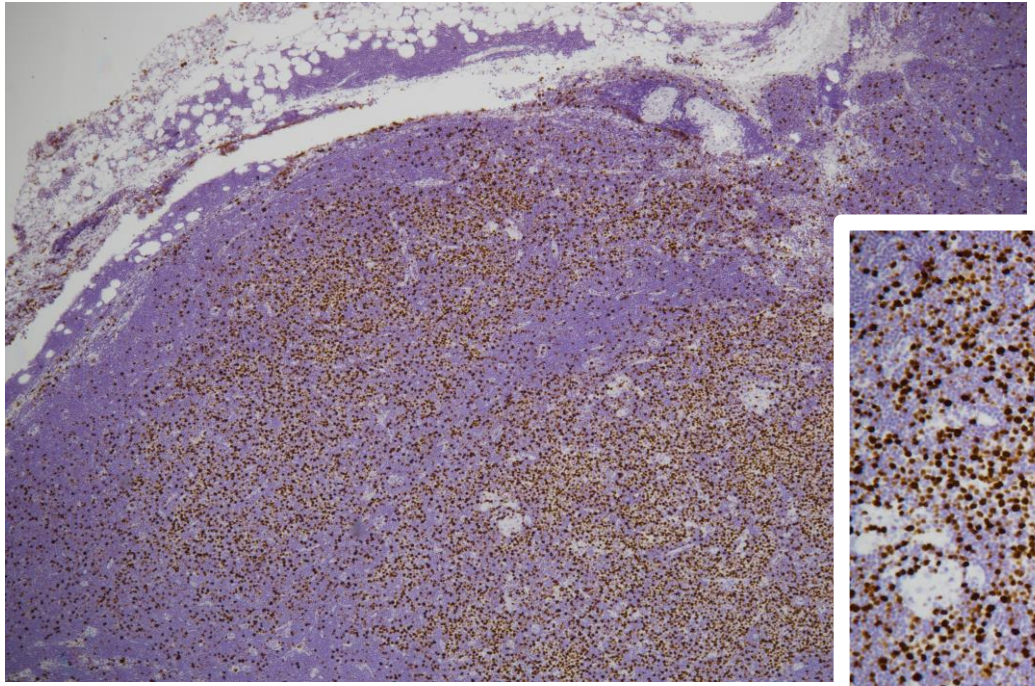
CD15

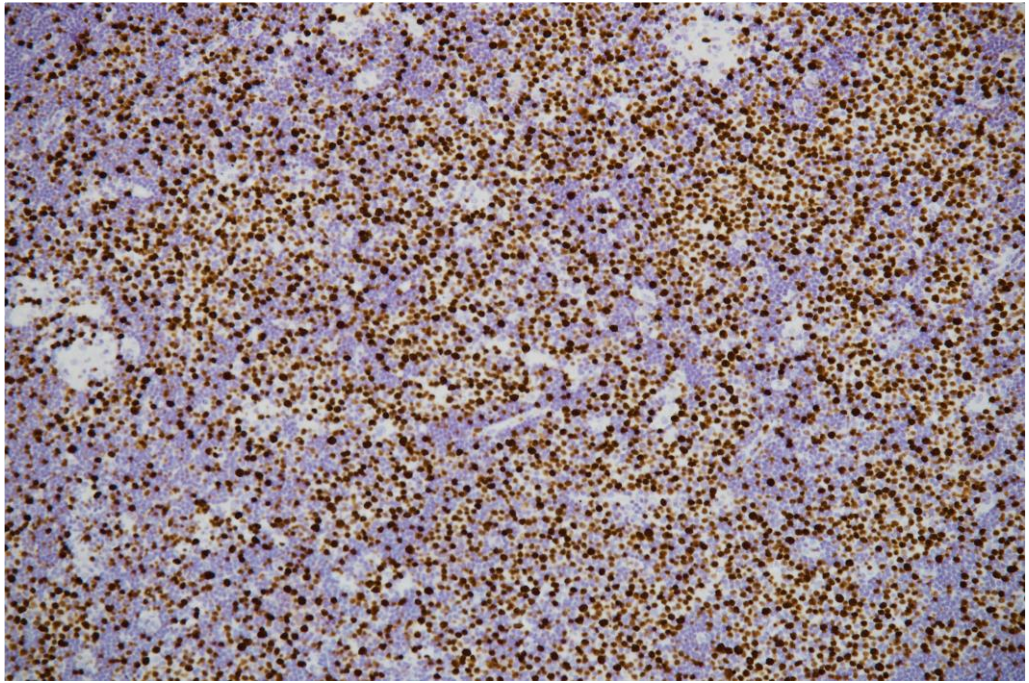


BCL6



C-MYC





Ki67

Small cells component

CD20+
CD5+
CD23+
LEF1+
Low Ki67

Large plasmoblastic cells nodules

PAX5-
CD20-
CD79a+(weak)/-
CD19-
CD138-
CD38+
MUM1+
BCL6-
CD10-
Lambda+
CD30+/-
C-MYC+/-
HHV8+
EBV/EBER ISH+
High ki67

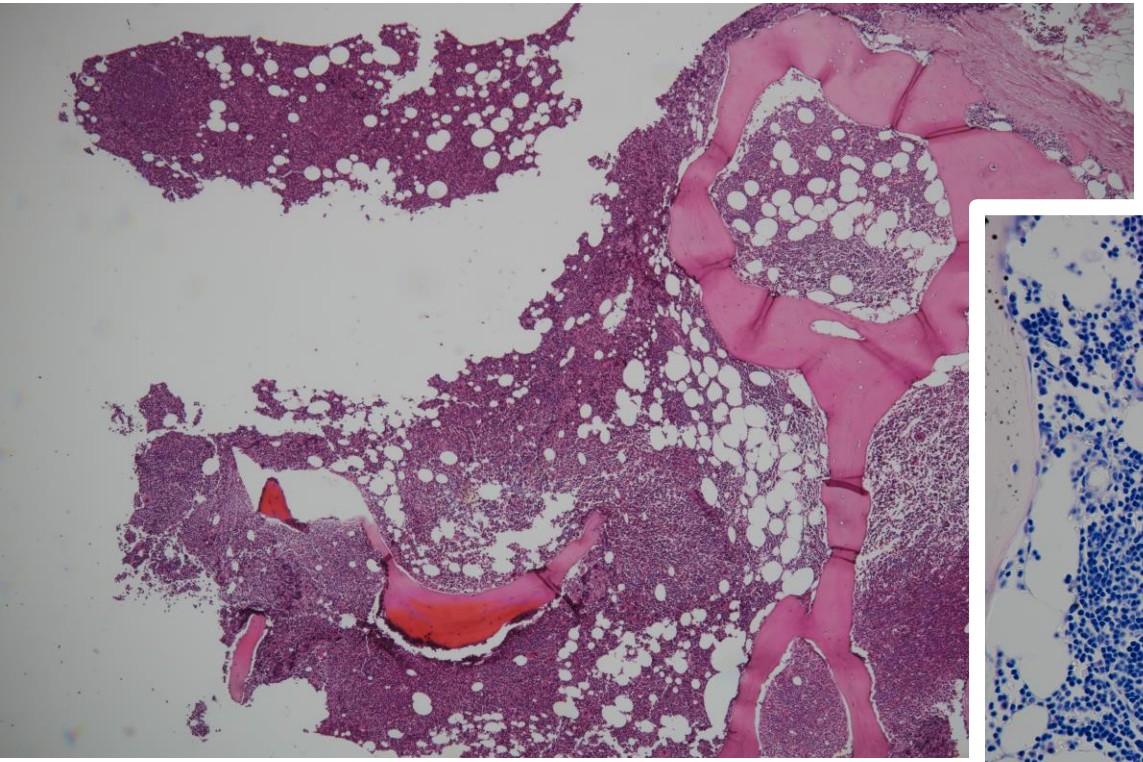
Diagnosis

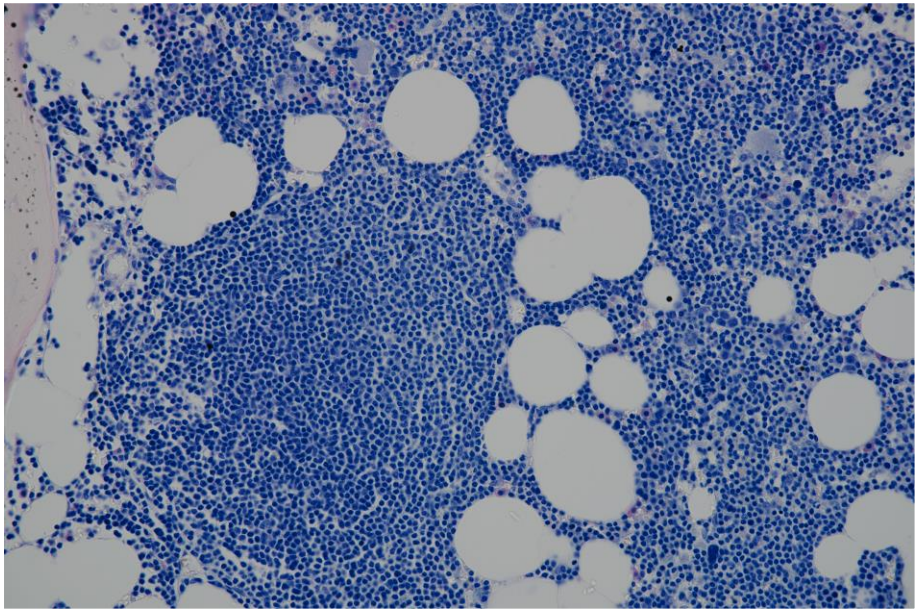
Foci of large plasmablastic cell lymphoma with a phenotype consistent with "serous lymphoma, extracavitary" (HHV8+/EBV+), in adenopathy with chronic lymphocytic leukemia.

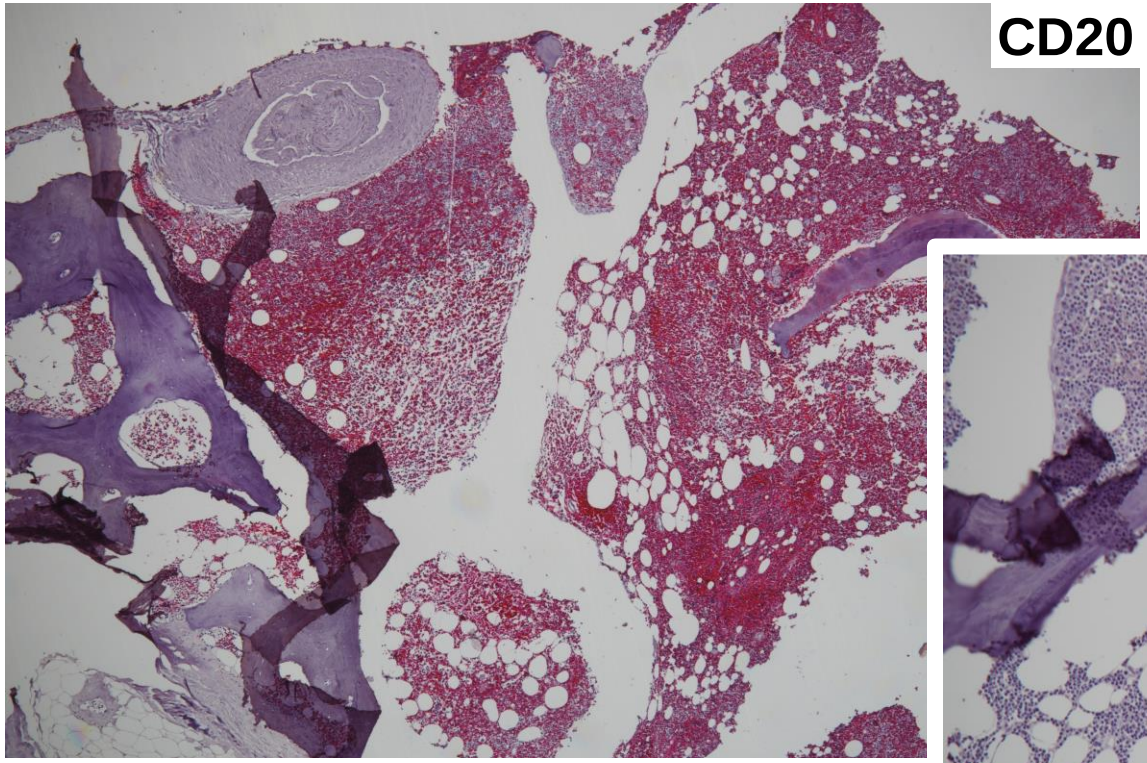
The coexistence of these two conditions suggests Richter's syndrome.

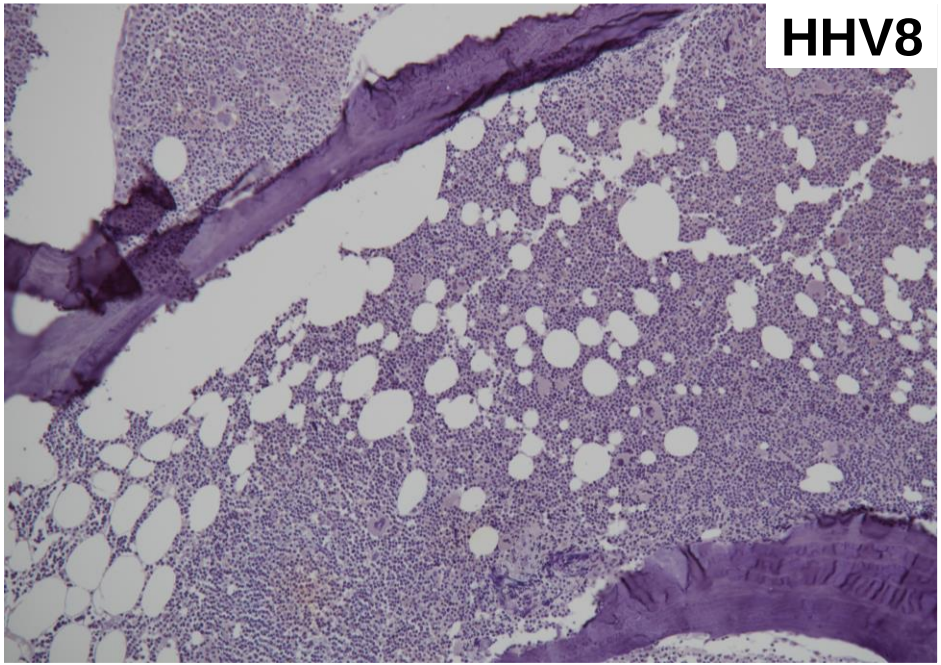
Verify acquired immunosuppression.

The patient undergoes bone marrow biopsy






CD20


HHV8

DIFFERENTIAL DIAGNOSIS

- Classic Hodgkin Lymphoma (CHL-RT):

  RS cells CD20-, PAX5+ (weak), CD30+, EBV+ or -, HHV8-; inflammatory background.

- Diffuse Large B Cells Lymphoma (DLBCL-RT):

  CD20/CD19/PAX5+, HHV8-.

- Plasmoblastic Lymphoma (PBL-RT):

  CD20/CD19/PAX5-, MUM1/CD38/CD138+, EBV+, HHV8-; plasmoblastic morphology.

RICHTER PLASMABLASTIC TRANSFORMATION

Exceptionally rare, aggressive form of Richter transformation (RT), where CLL/SLL evolves into a highly aggressive neoplasm with plasmablastic differentiation (immunoblastic morphology and terminal B-cell differentiation).
Often associated with TP53 mutations or del17p.
Rapidly progressing lymphadenopathy, B-symptoms (fever, weight loss, night sweats), high LDH, and often extranodal involvement (skin, gastrointestinal tract, bone marrow).

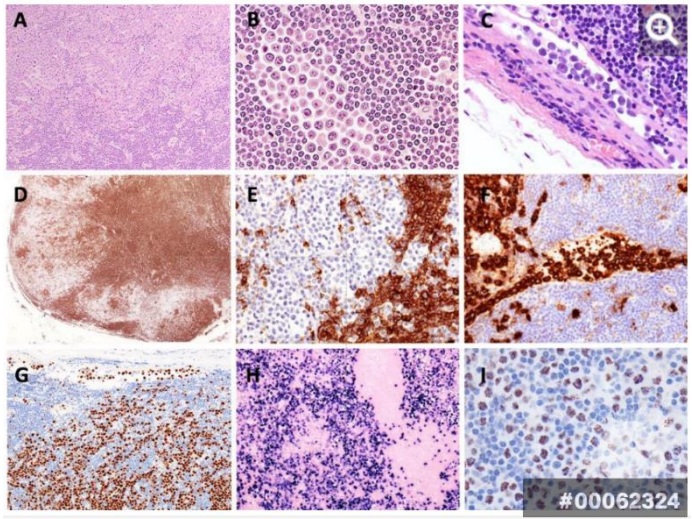
Recognition and incorporation in new classifications of plasmablastic RT as a distinct entity is critical, as its biology and resistance profile differ from classical RT.

Ramsey MC. et al., eJHaem 2023, 4:1203-1207
Filipovic J. et al., Diagnostics 2026, 16:702
Hayes C. et al., J. Hemopathlogy 2014, 7:189-193
Castonguay M. et al., Current Oncology 2025, 32:550
Chan KL. et al., Br. J Haematology 2017, 177:319
Kittai AS. et al., Blood 2025, 146: 291

PRIMARY EFFUSION LYMPHOMA (PEL)

EXTRACAVITARY PEL (EC PEL)

- PEL comprises ~4% of human immunodeficiency (HIV) related lymphomas and < 1% of non-HIV associated lymphomas.
- Present with lymphomatous effusions involving serous cavities.
- Large cells with immunoblastic, plasmablastic or anaplastic features.
- Characteristic phenotype: positive for CD38, CD138; negative for B cell associated markers (CD19, PAX5, CD20).
- Positive for KSHV/HHV8 and EBV-encoded small RNAs (EBER).



https://imagebank.hematology.org/image/62324/nodal-extracavitary-variant-of-primary-effusion-lymphoma-as-complication-of-chronic-lymphocytic-leuk

No confirmed immunodeficiency

August 2025

First BFM cycle (reduced dose, due to his age).

PET CT
Diffuse lymphadenopathy, with a high SUV corresponding to a retroperitoneal lymph node cluster near the left common iliac (SUV of 18).

Retroperitoneal para-aortic lymph node biopsy
Histology similar to the initial one.

Two additional BFM cycles

PET CT

Low SUV.

April 2026

Axillary lymph node biopsy
Histology characterized by CLL only, with no blasts.

Patient is currently in clinical remission.

ACKNOWLEDGMENTS

Elena Sabattini
IRCCS Azienda Ospedaliero-Universitaria di Bologna
Dipartimento di Oncologia e di Ematologia
S.S.D. di Emolinfopatologia

SERVIZIO SANITARIO REGIONALE
EMILIA-ROMAGNA
Azienda Ospedaliero - Universitaria di Bologna

Policlinico S. Orsola-Malpighi

ALMA MATER STUDIORUM
UNIVERSITÀ DI BOLOGNA