SH Workshop Chicago 2017

An Aggressive Burkitt-like Lymphoma

SH2017-0137

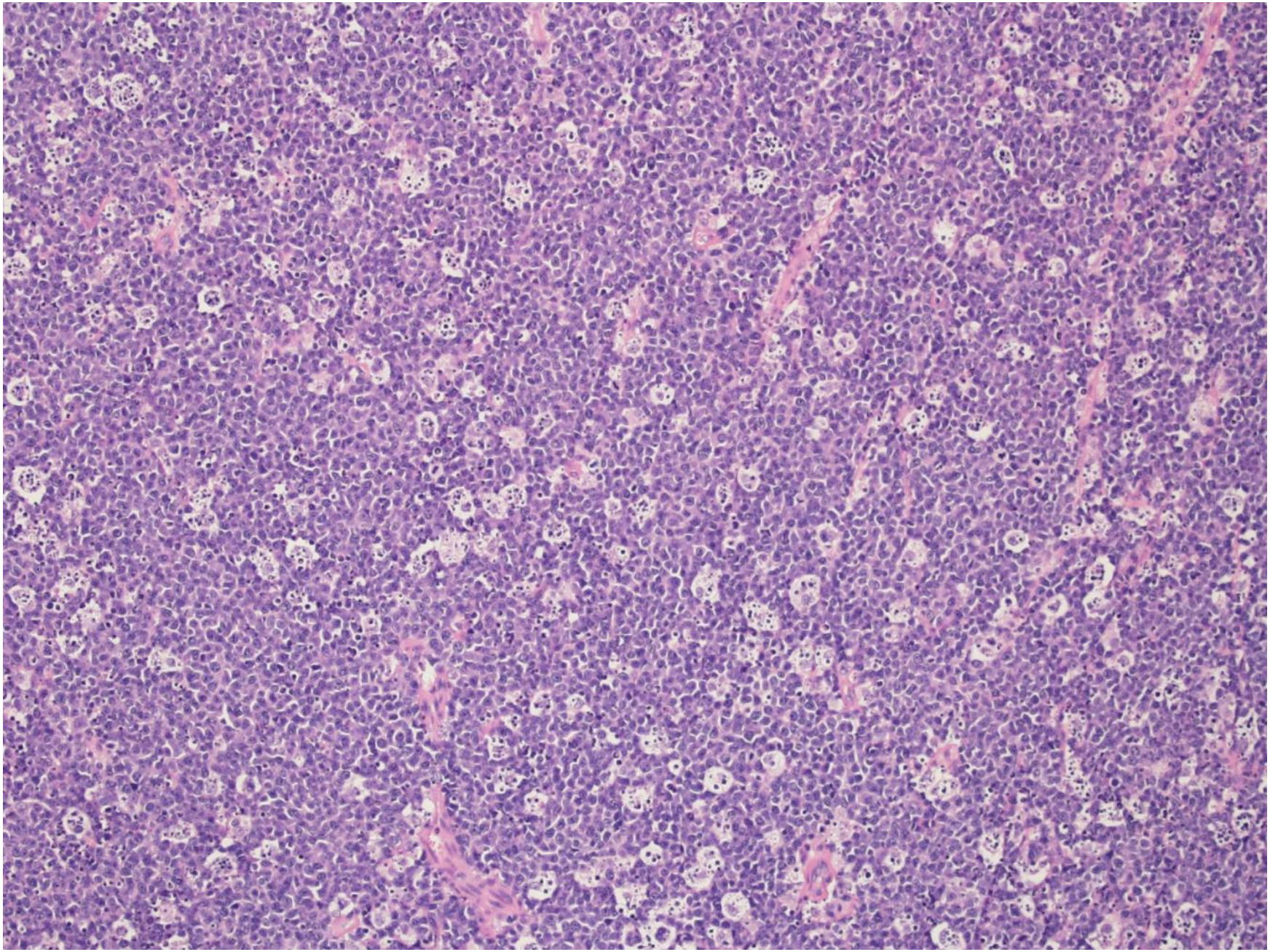
Katrin Hüttl, MD
German Ott, MD

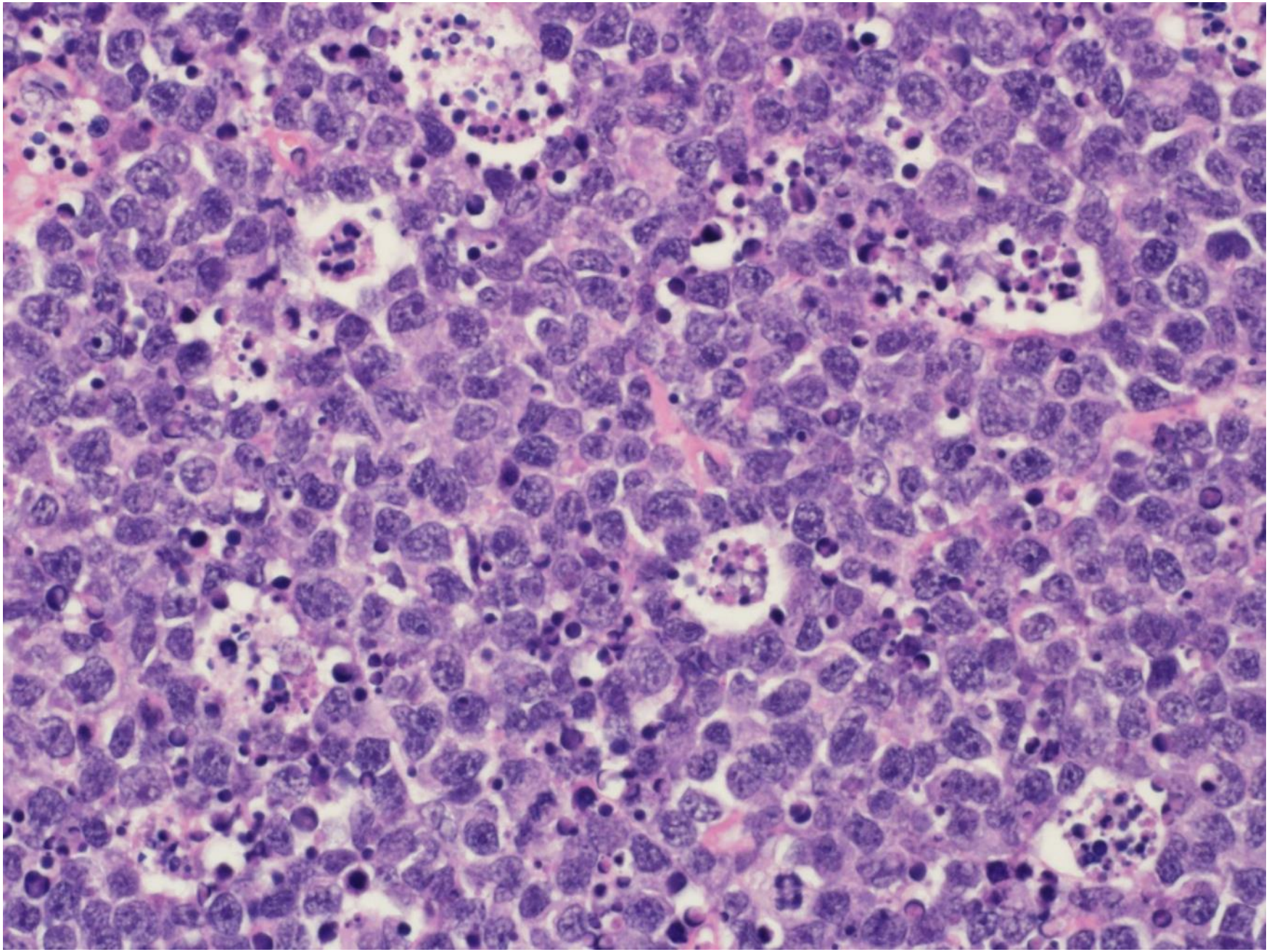
Robert-Bosch-Krankenhaus Stuttgart, Germany

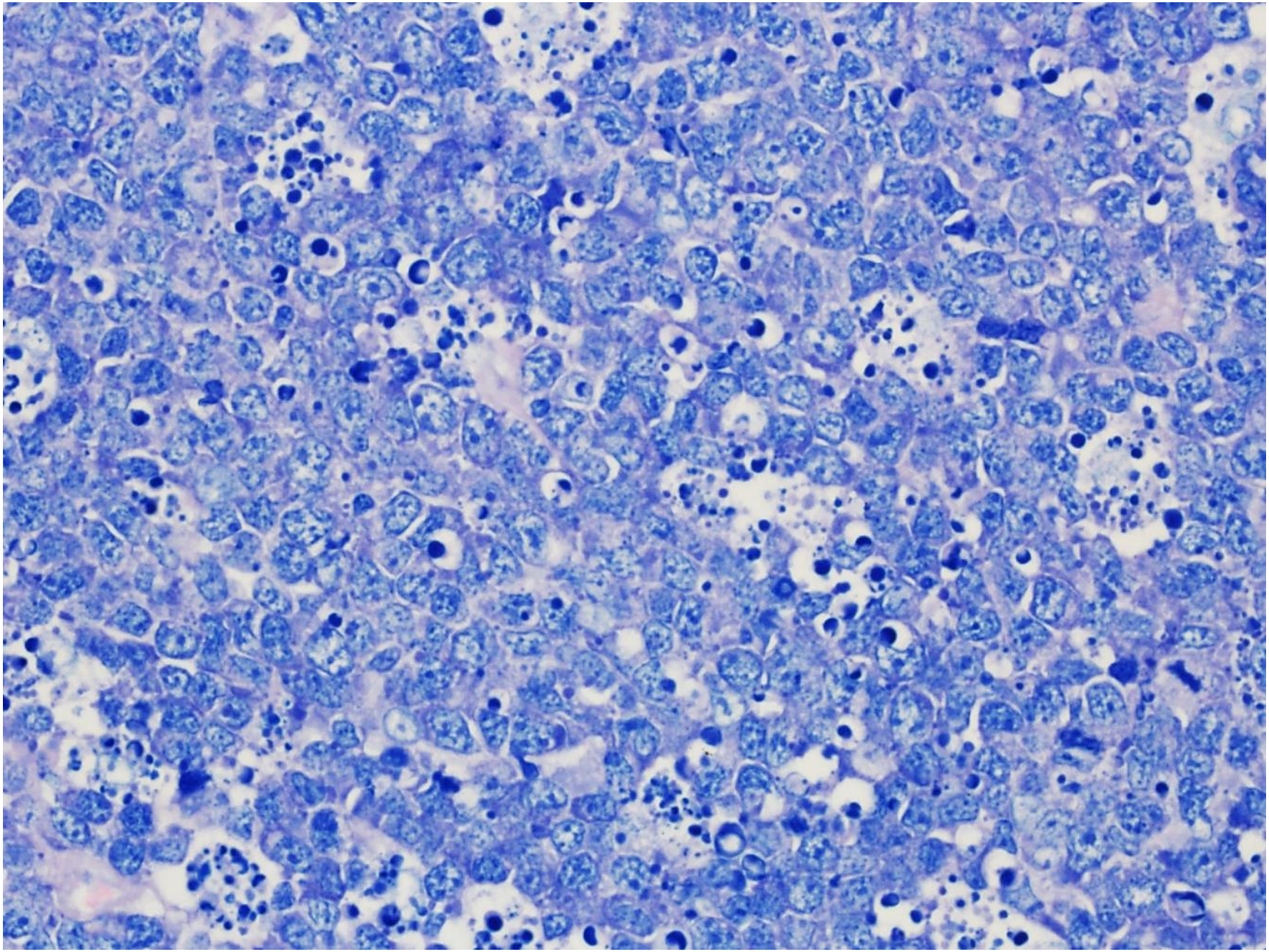
Clinical Data

- **A 21-year old male patient**
- **<u>10/2015</u>**
  Presents with acute abdominal symptoms. Laboratory tests unremarkable with exception of slight anemia (Hb 9,3 g/dl). Diagnosis of a tumor in the appendiceal region. Clinical staging reveals Stage IVA disease with involvement of the omentum.
- **<u>11/2015</u>**
  Opstipation, abdominal discomfort and pain. After diagnosis of an ileus, extended surgery with omentectomy and resection of parts of terminal ileum and colon. Repeated staging reveals no enlarged lymph nodes cervical, mediastinal, abdominal. No bone marrow involvement, no CNS involvement.

Histology reveals aggressive lymphoma.







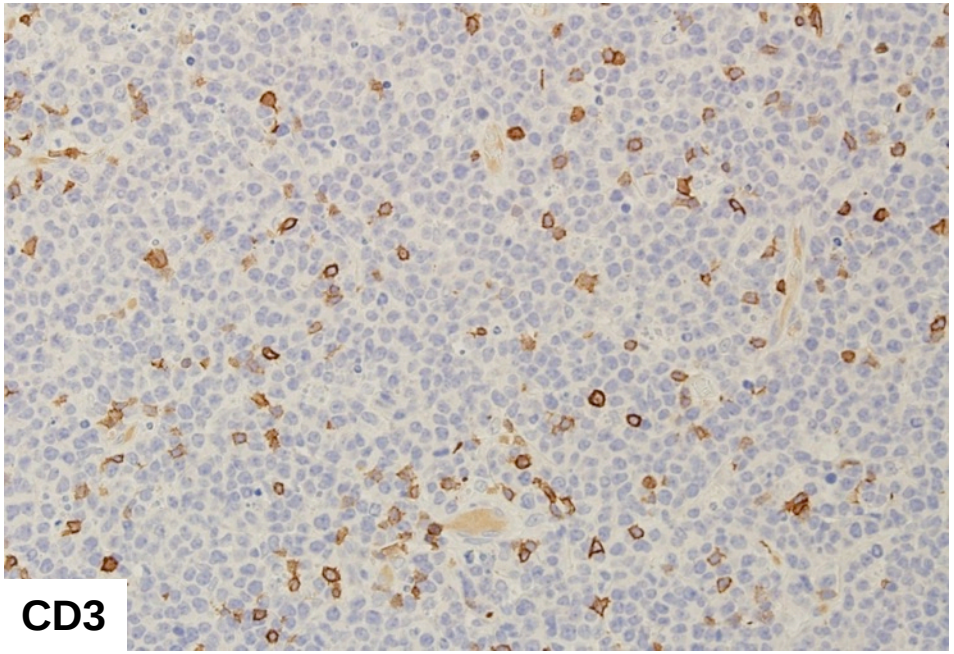
Summary of Features

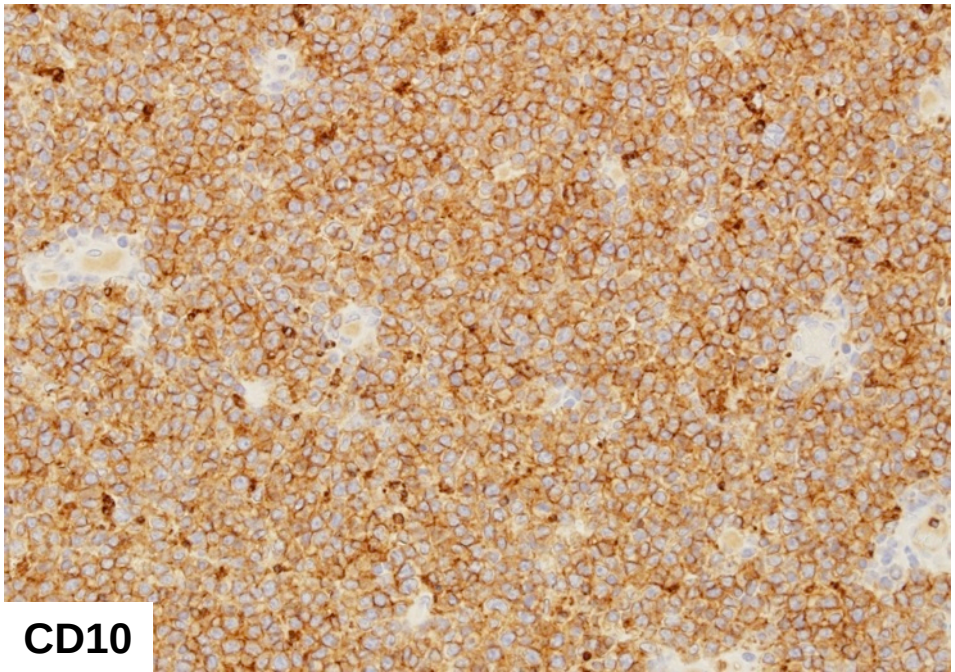
- Large tumor mass in the area of the terminal ileum, cecum and appendix
- Aggressive lymphoma with a „starry sky“ pattern
- Medium-sized to large cohesive blasts

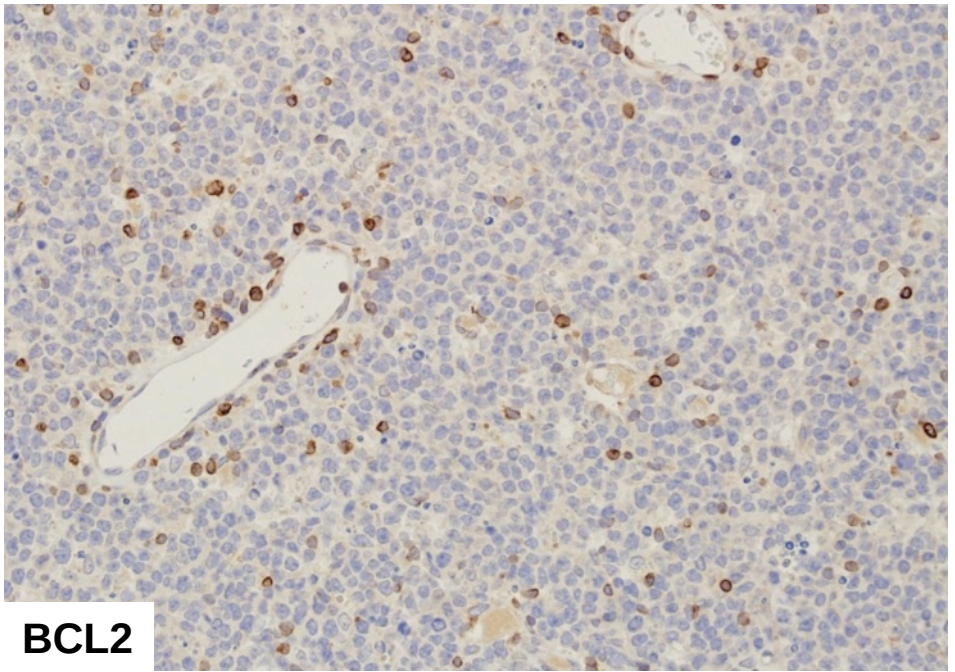
Burkitt-Lymphoma?

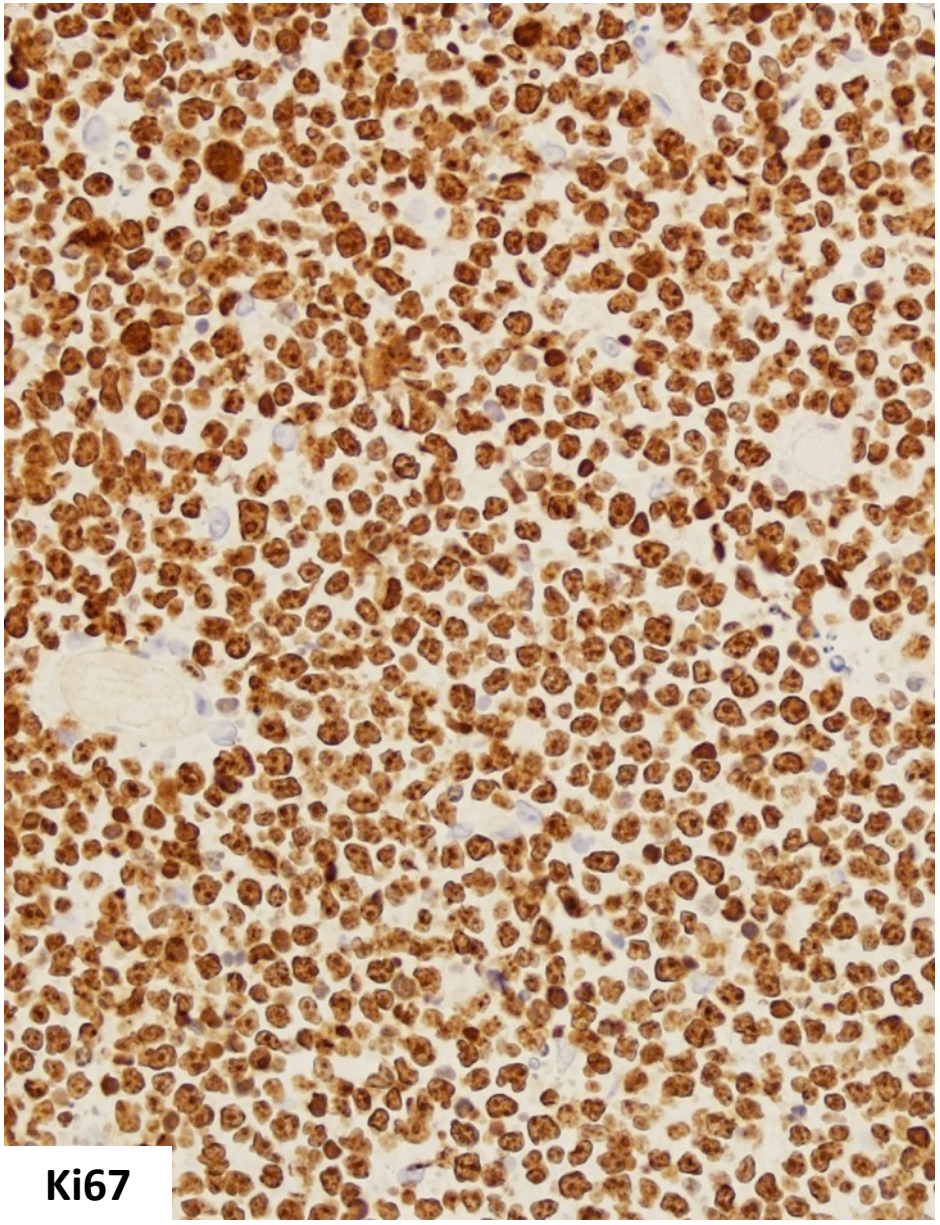
DLBCL with „Burkitt-like“ features?


CD20

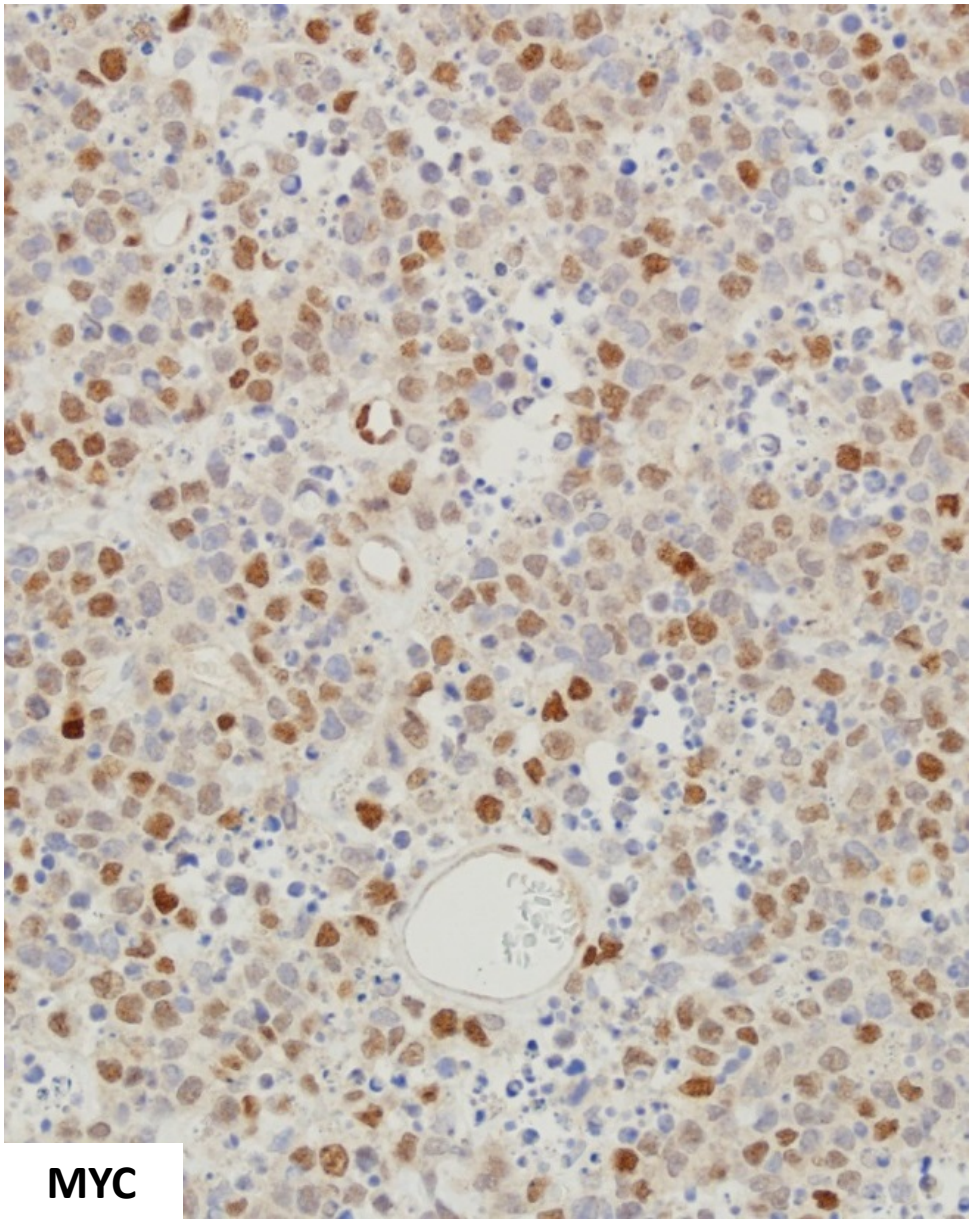

CD3


CD10


BCL2



Ki67



MYC

Summary of Immunohistochemistry

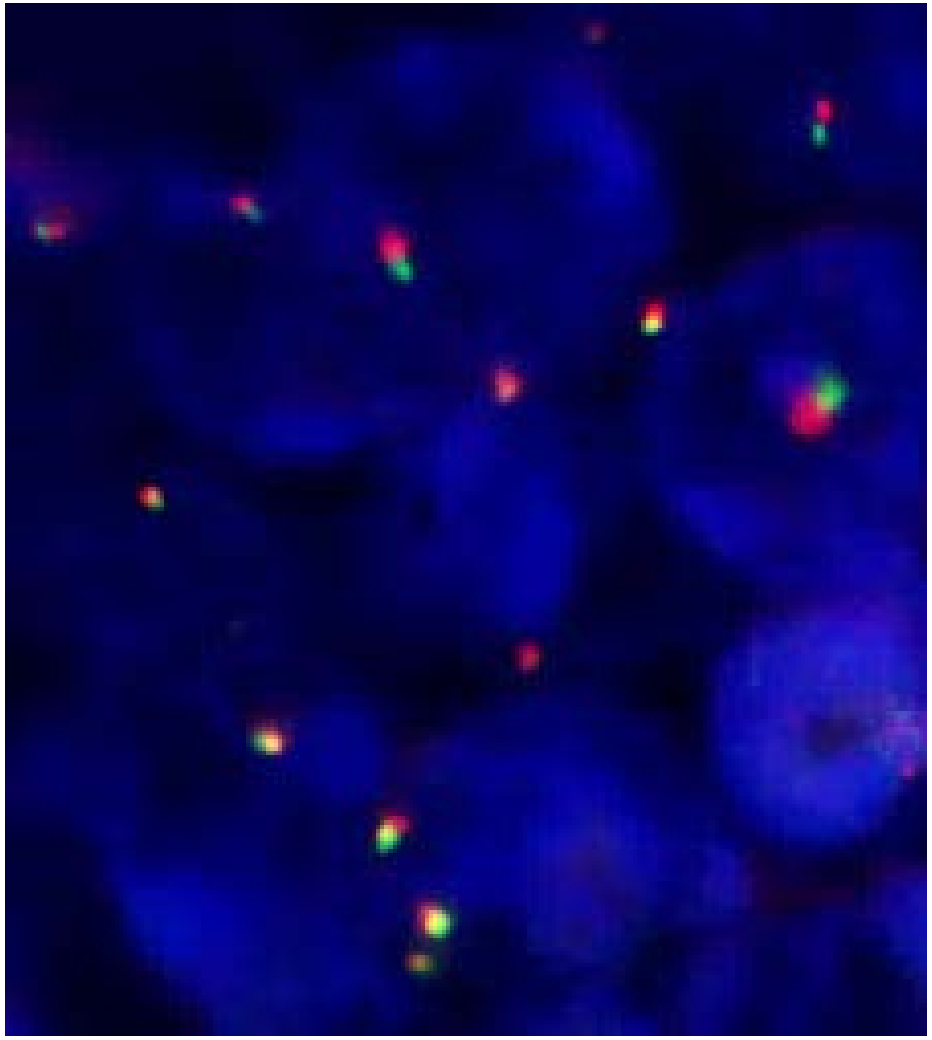
CD20+, CD10+, BCL2-, BCL6+,
IRF4/MUM1-, Ki67 100%

→ **„Classical“ Burkitt phenotype**

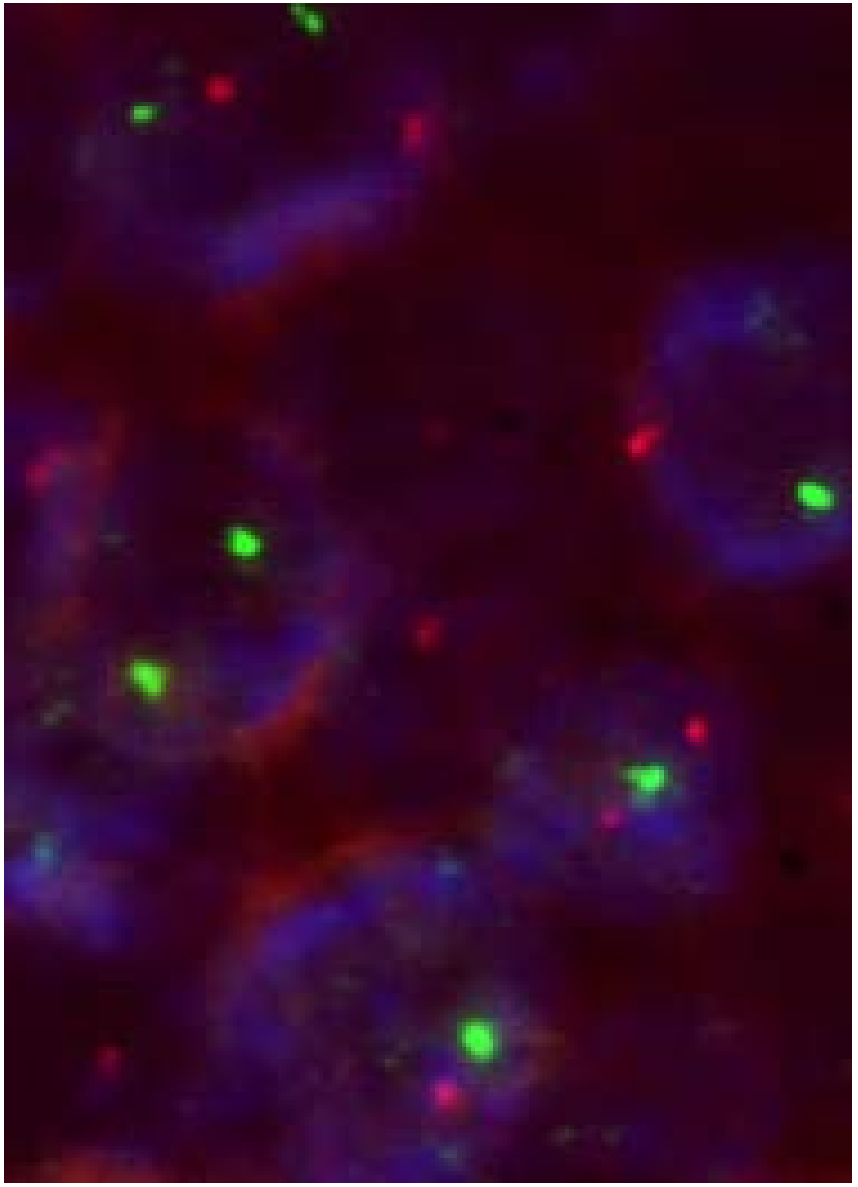
But: No „significant“
over-expression of MYC

FISH:

MYC-BAP



MYC t(8;14)



Burkitt-Lymphoma without MYC Rearrangement?

A recurrent 11q aberration pattern characterizes a subset of MYC-negative high-grade B-cell lymphomas resembling Burkitt lymphoma

Salaverria et al. BLOOD, 20 FEBRUARY 2014 • VOLUME 123, NUMBER 8

Key Points

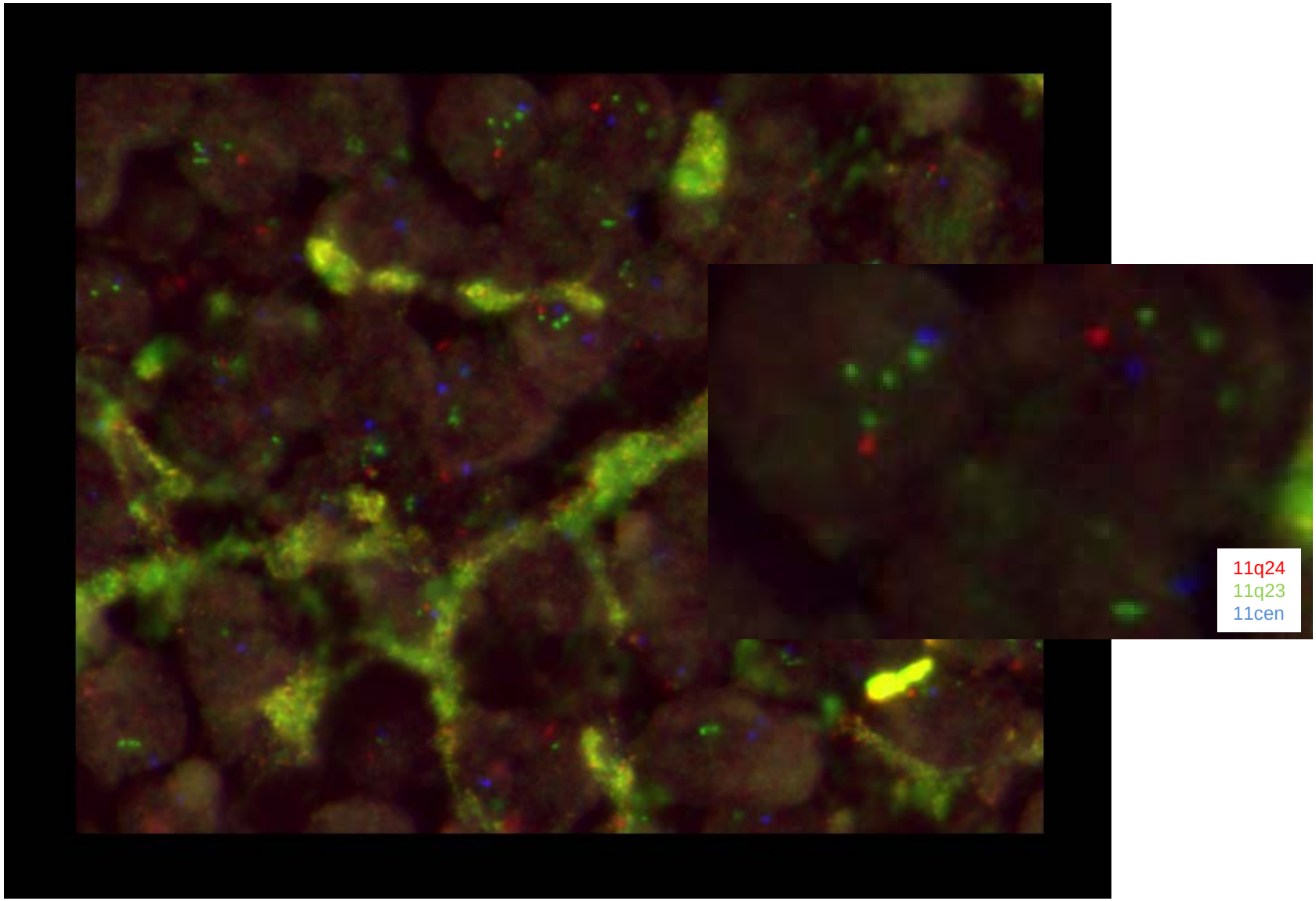
- A subset of lymphomas with gene expression and pathological characteristics of Burkitt lymphomas but absence of *MYC* translocation does exist.
- These lymphomas carry chr 11q proximal gains and telomeric losses, suggesting co-deregulation of oncogenes and tumor suppressor genes.

These findings indicate the existence of a molecular distinct subset of B-cell lymphomas reminiscent of BL which is characterized by deregulation of genes in 11q (*Blood*. 2014;123(8):1187-1198).

Pienkowska-Grela Med.Oncol. 2011
Ferreiro Haematologica 2015
Zajdel Tumour Biol 2015

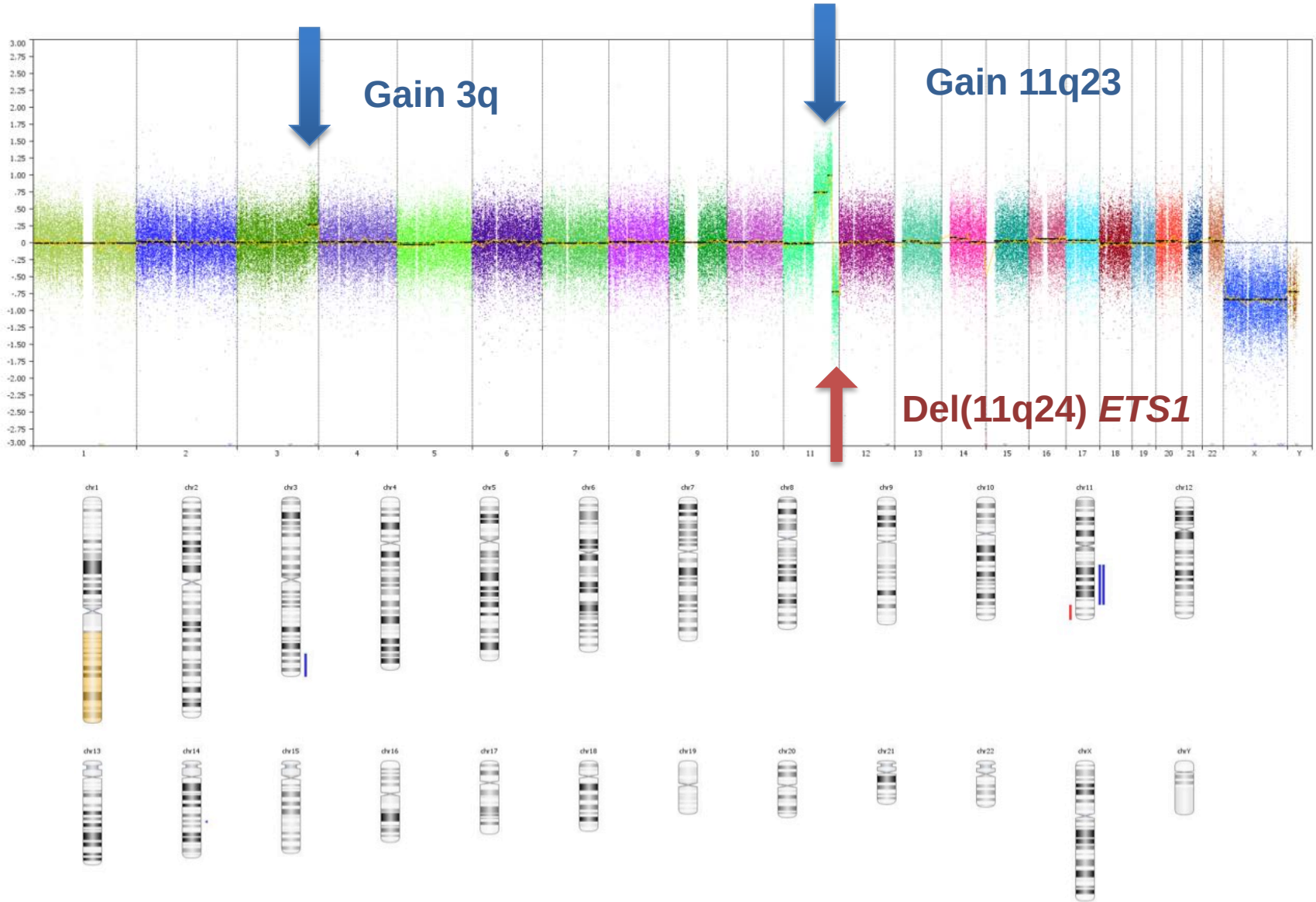
FISH Analysis of Chromosome 11

Courtesy R. Siebert, Kiel



OncoScan Analysis of the Lymphoma

Courtesy R. Siebert, Kiel



Diagnosis:
(WHO-Update 2016)

Burkitt-like Lymphoma with 11q Aberration (Provisional)

Swerdlow et al. Blood 2016

Diagnostic Strategy

- The diagnosis can be suspected basing on the slightly different cytomorphology and *MYC* breakpoint negativity
- FISH with probes directed at the hotspots in chromosome 11 helpful for screening
- Advantage of FISH: easy to perform, allows for targeted analysis of tumor cells if identifiable
- When FISH shows a clearly detectable gain or deletion, Oncoscan is not necessary
- Difficult interpretation of the FISH or – at the moment – negativity → Oncoscan is needed

Follow-up

- **11/2015 – 03/2016:**

  Therapy with 8 cycles of chemotherapy according to the B-ALL protocol. Atypical pneumonia after the fourth cycle.

- **Last staging 08/2017: Complete Remission.**

Acknowledgements

Prof. Reiner Siebert, MD and Rabea Wagener PhD, University of Ulm, Germany

Matthias Vöhringer, MD, Department of Hematooncology, Robert-Bosch-Krankenhaus, Stuttgart, Germany

Annette Staiger PhD, and Heike Horn PhD, Molecular Diagnostics Unit, Robert-Bosch-Krankenhaus and Dr. Margarete-Fischer-Bosch Institute of Clinical Pathology, Stuttgart, Germany.

Panel Diagnosis:

Burkitt-like Lymphoma with 11q Aberration