



Case 2

Presented by
Dr Dina Osman
Haematology Registrar
University Hospital Birmingham

Conflicts of Interest

- There are no conflicts of interest to disclose.

Case 2

A 55-year- old previously fit and well gentleman presented with a few month history of:

- Bilateral inguinal lymphadenopathy
- Drenching night sweats

Blood Results

- FBC:

Hb 125g/L

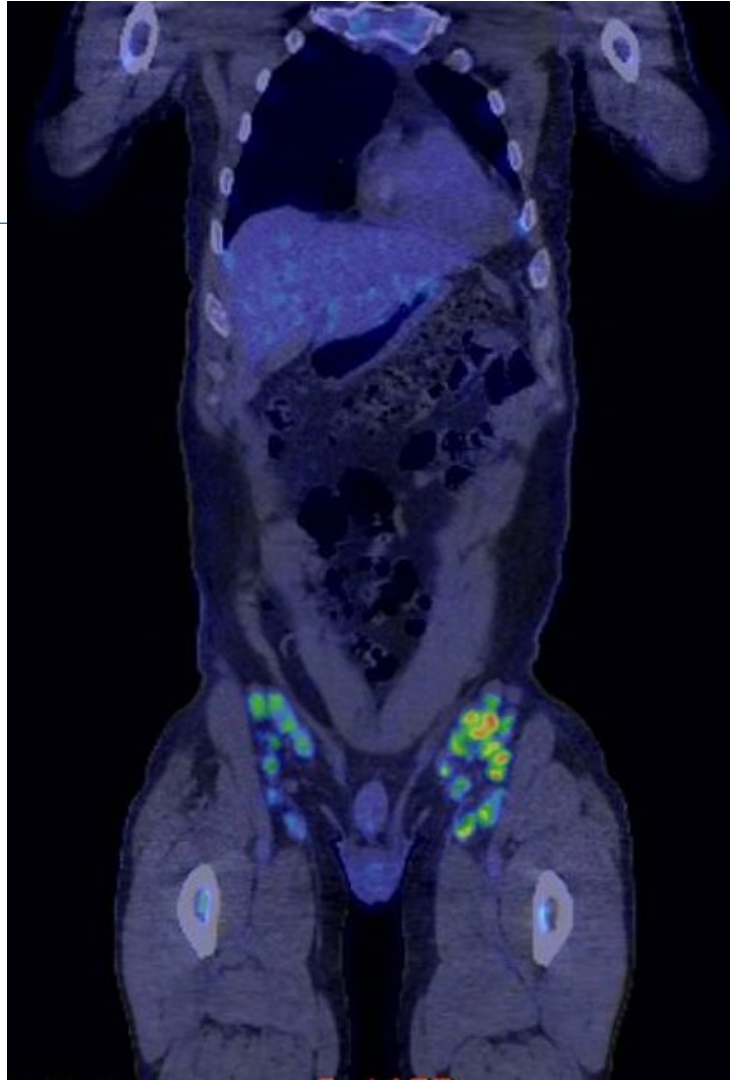
WBC $20.07 \times 10^9/L$

Platelets $408 \times 10^9/L$

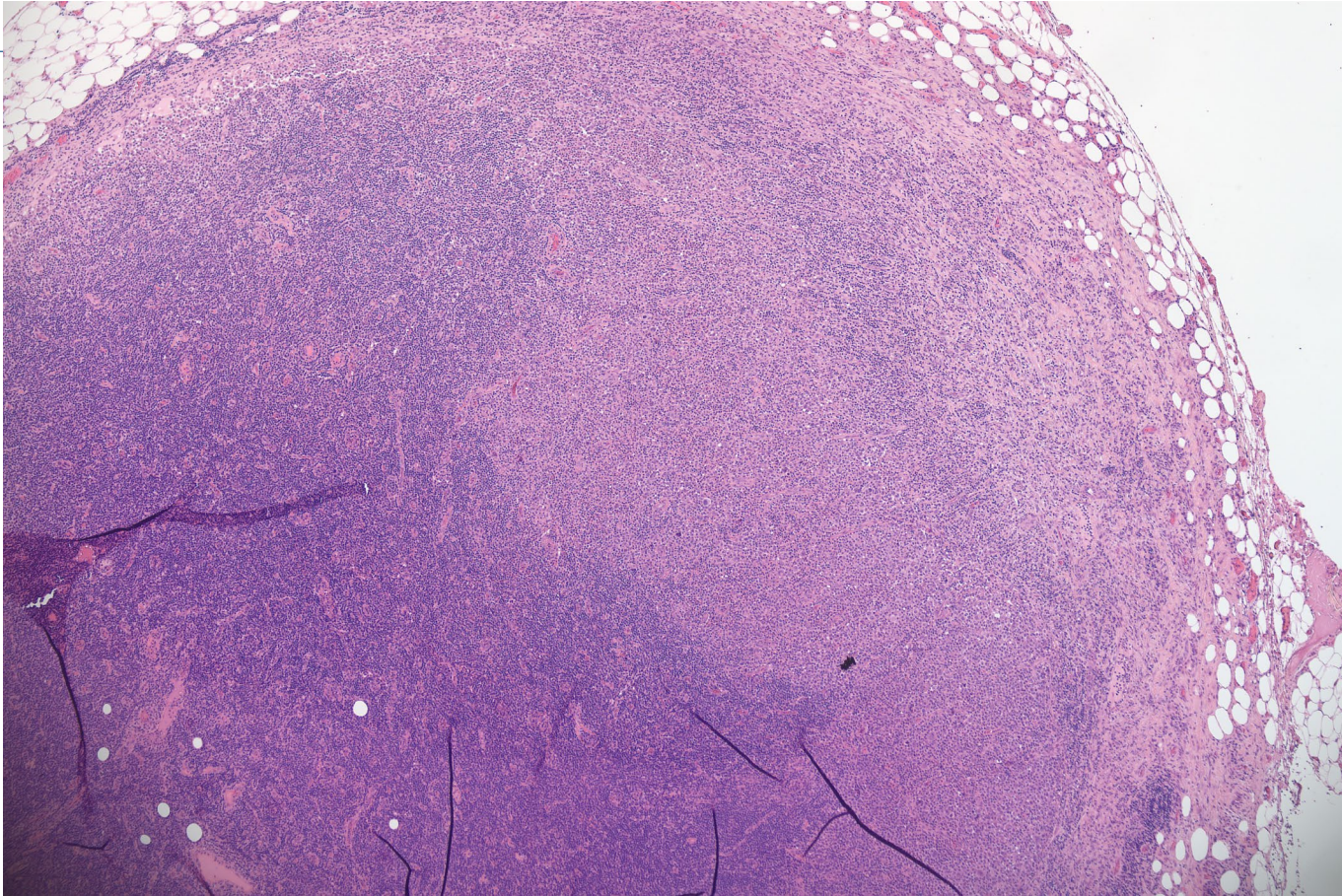
Neutrophils $16.64 \times 10^9/L$

Lymphocytes $2.7 \times 10^9/L$

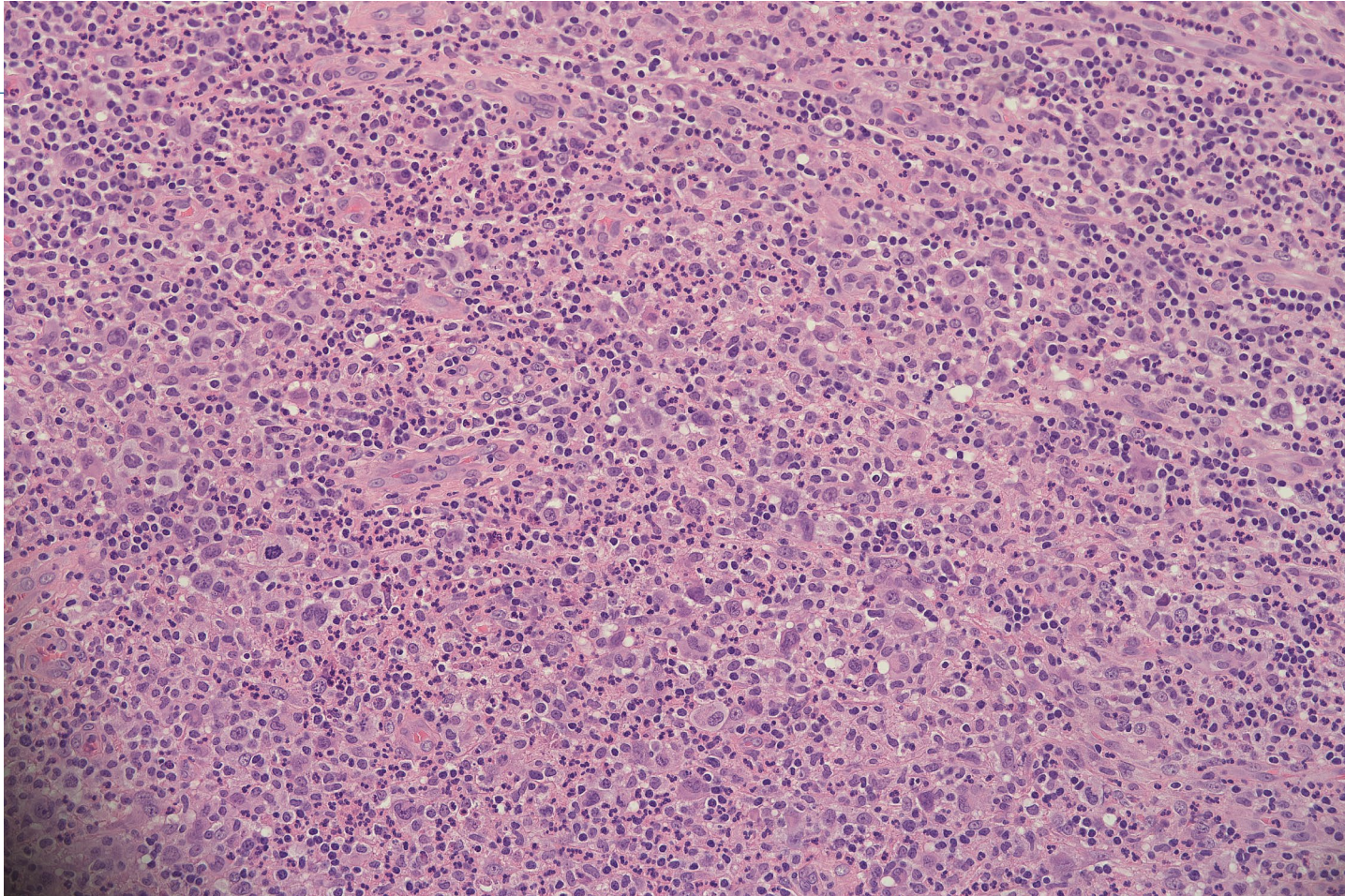
- LDH 390 U/L (NR 125-220U/L)



Inguinal Core Lymph Node Biopsy x4



Inguinal Core Lymph Node Biopsy x10



Immunohistochemistry of the LN

- CD10 +
- CD20+
- BCL2+

- BCL6-
- CD5-
- CD30-
- IRF4 -

What is Your Differential Diagnosis?

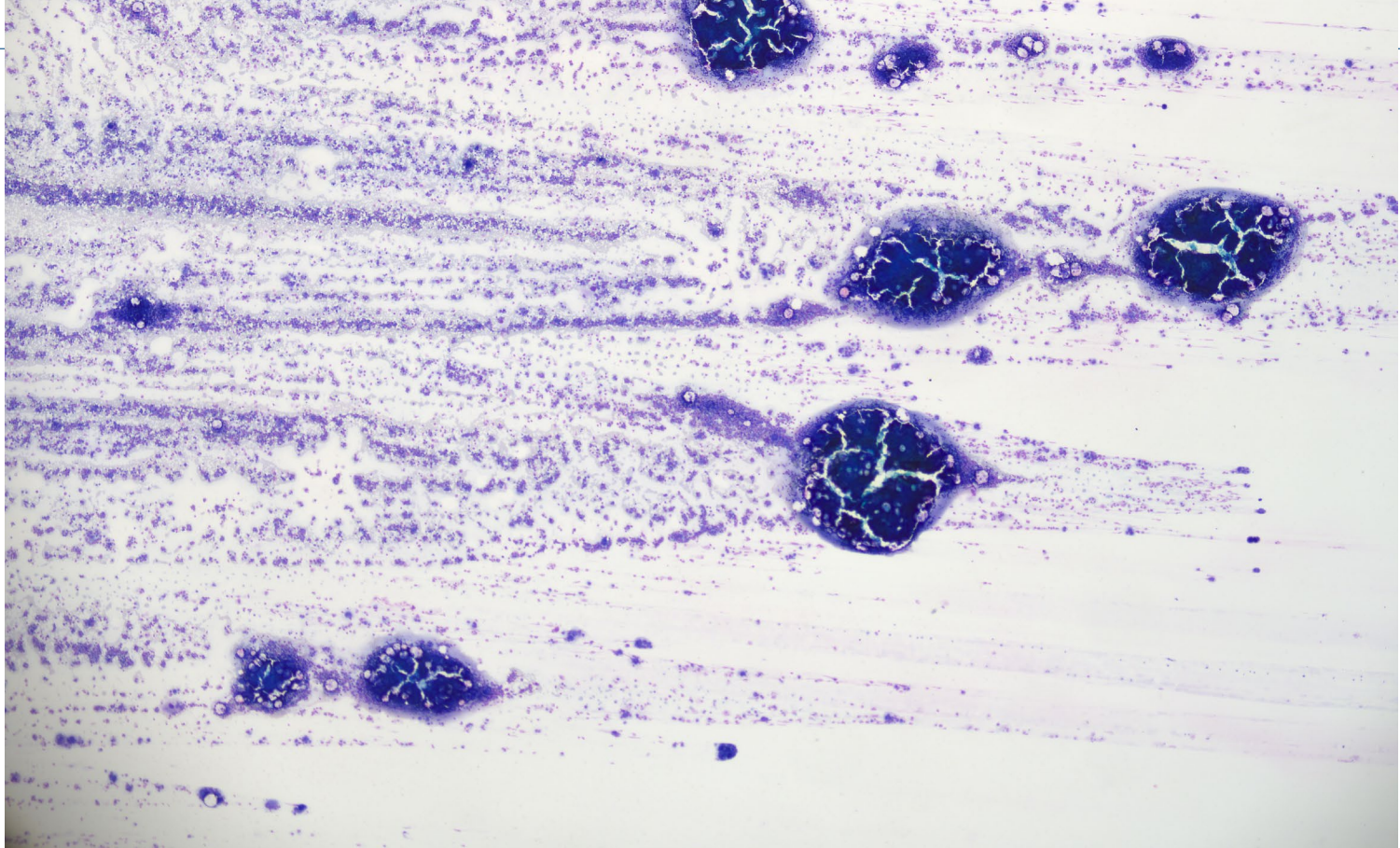
- A) Low grade B-cell lymphoma
- B) Low grade B- cell lymphoma with high grade transformation
- C) High grade B-cell lymphoma
- D) T-cell lymphoma



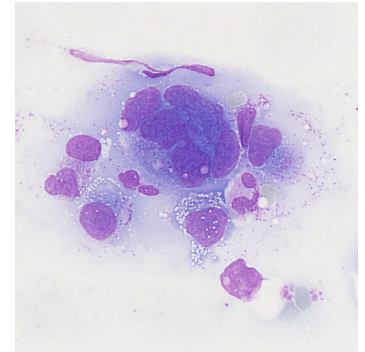
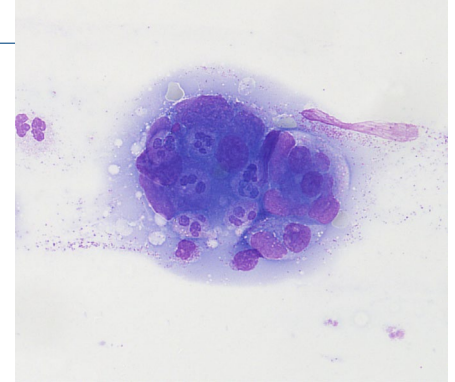
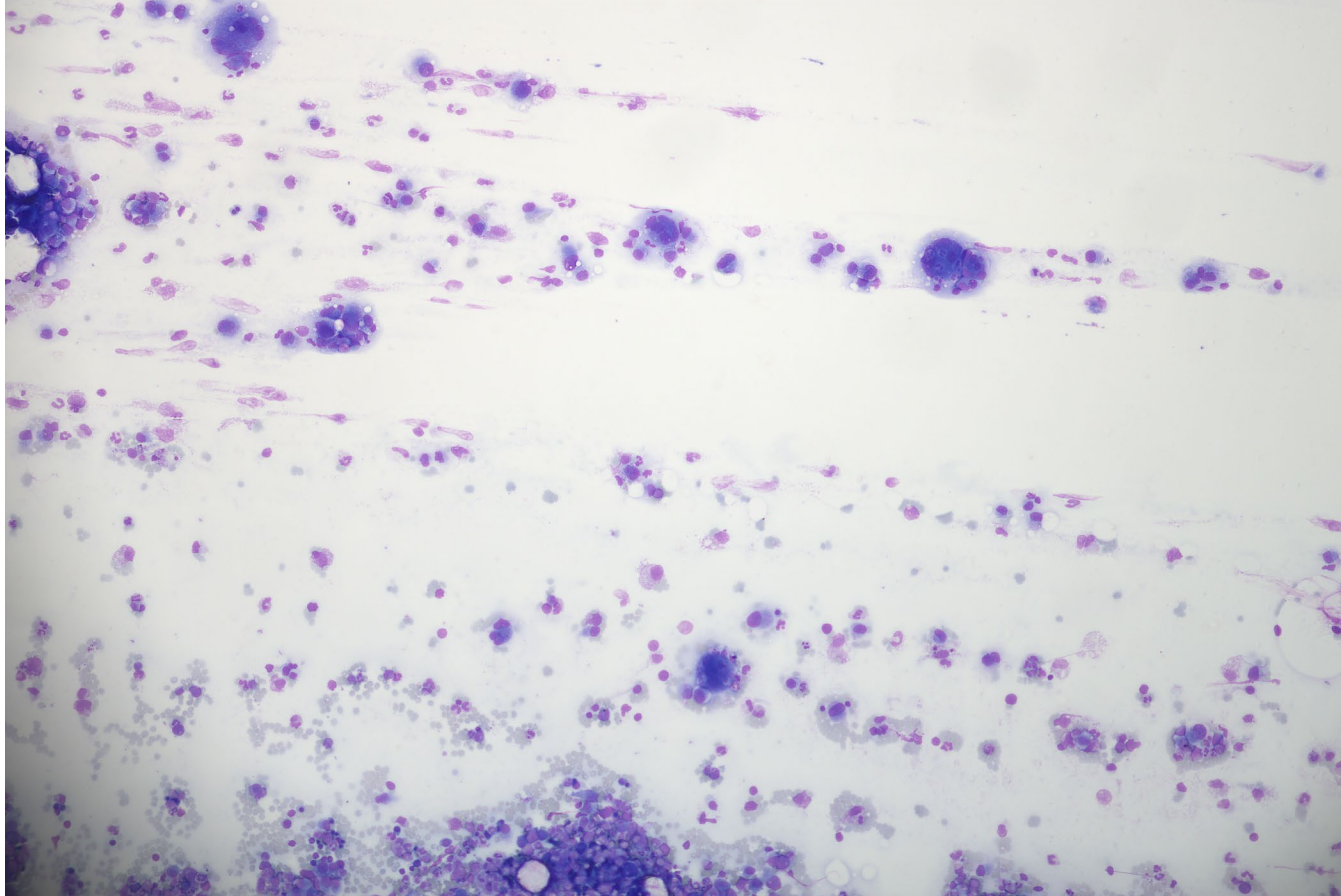
Diagnosis....follicular lymphoma

- The patient was diagnosed with follicular lymphoma
- Ki67 was 10%
- t(14;18) was positive by FISH at 12%
- A bone marrow biopsy was done for staging purposes

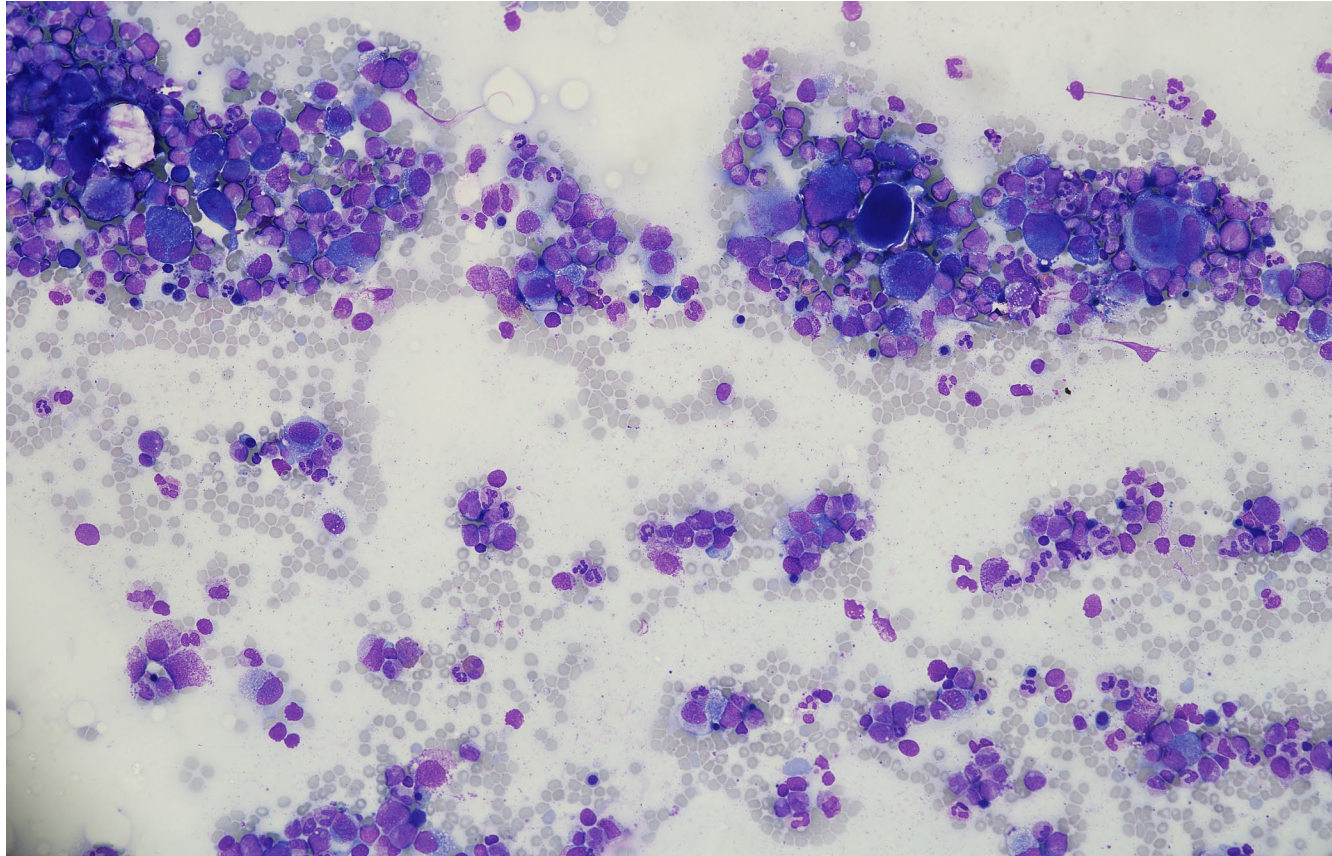
Bone Marrow Aspirate x 4



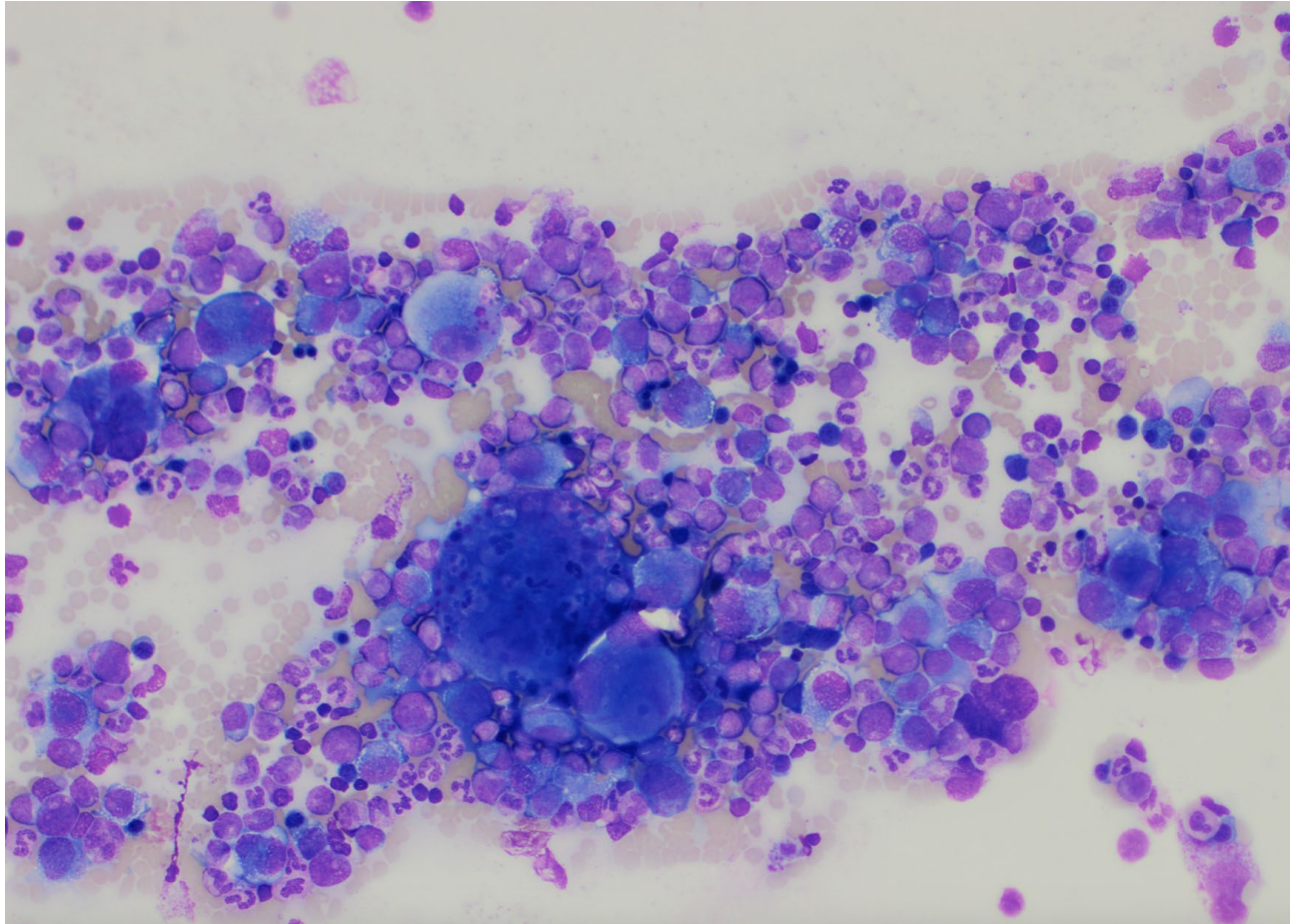
Bone Marrow Aspirate x10



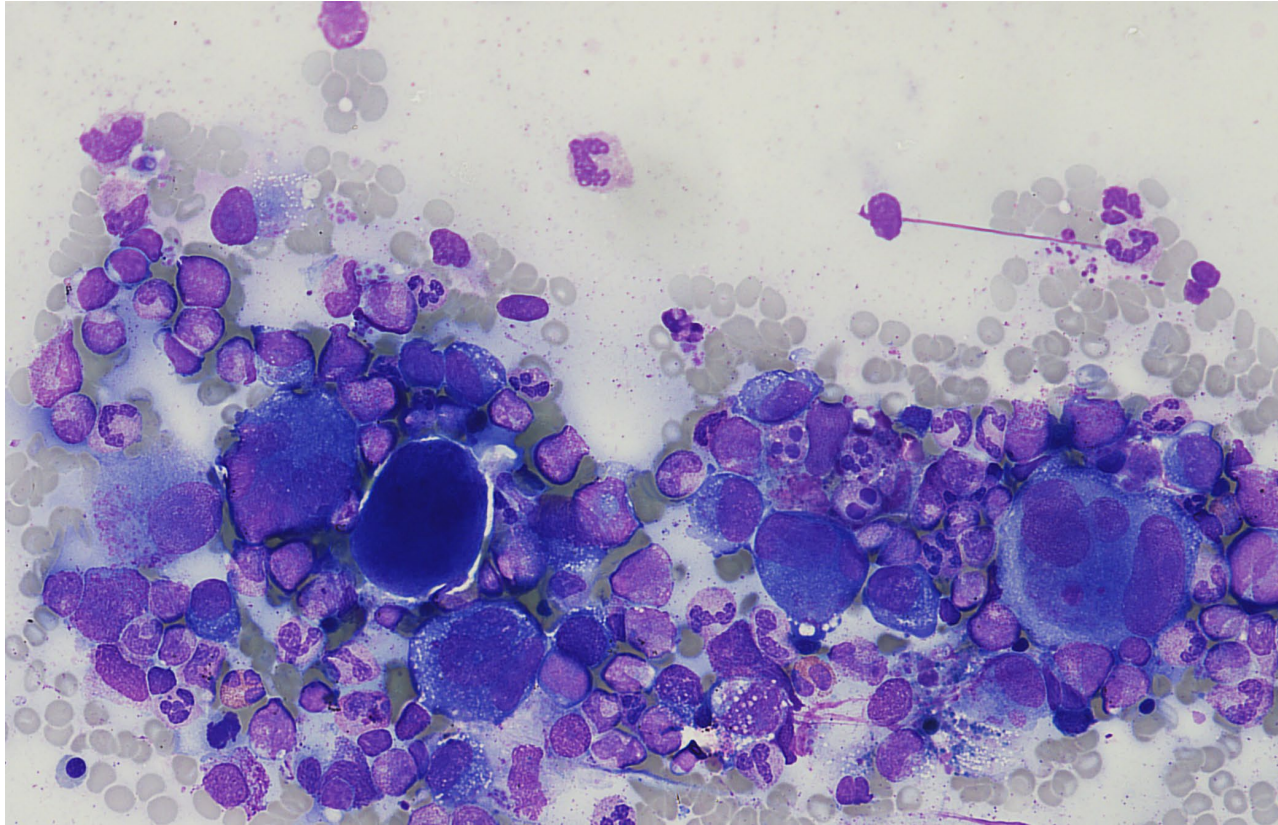
Bone Marrow Aspirate x 20

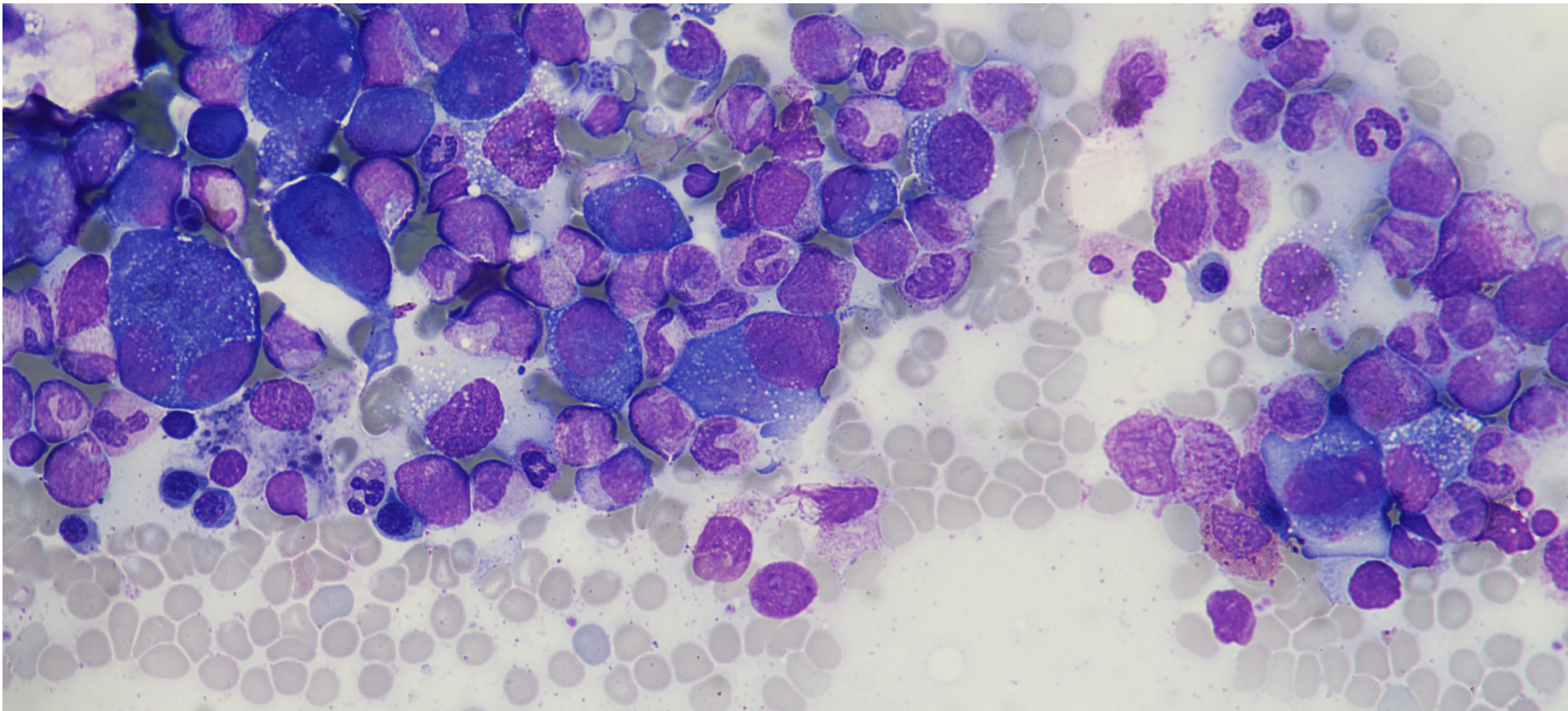


Bone Marrow Aspirate x20

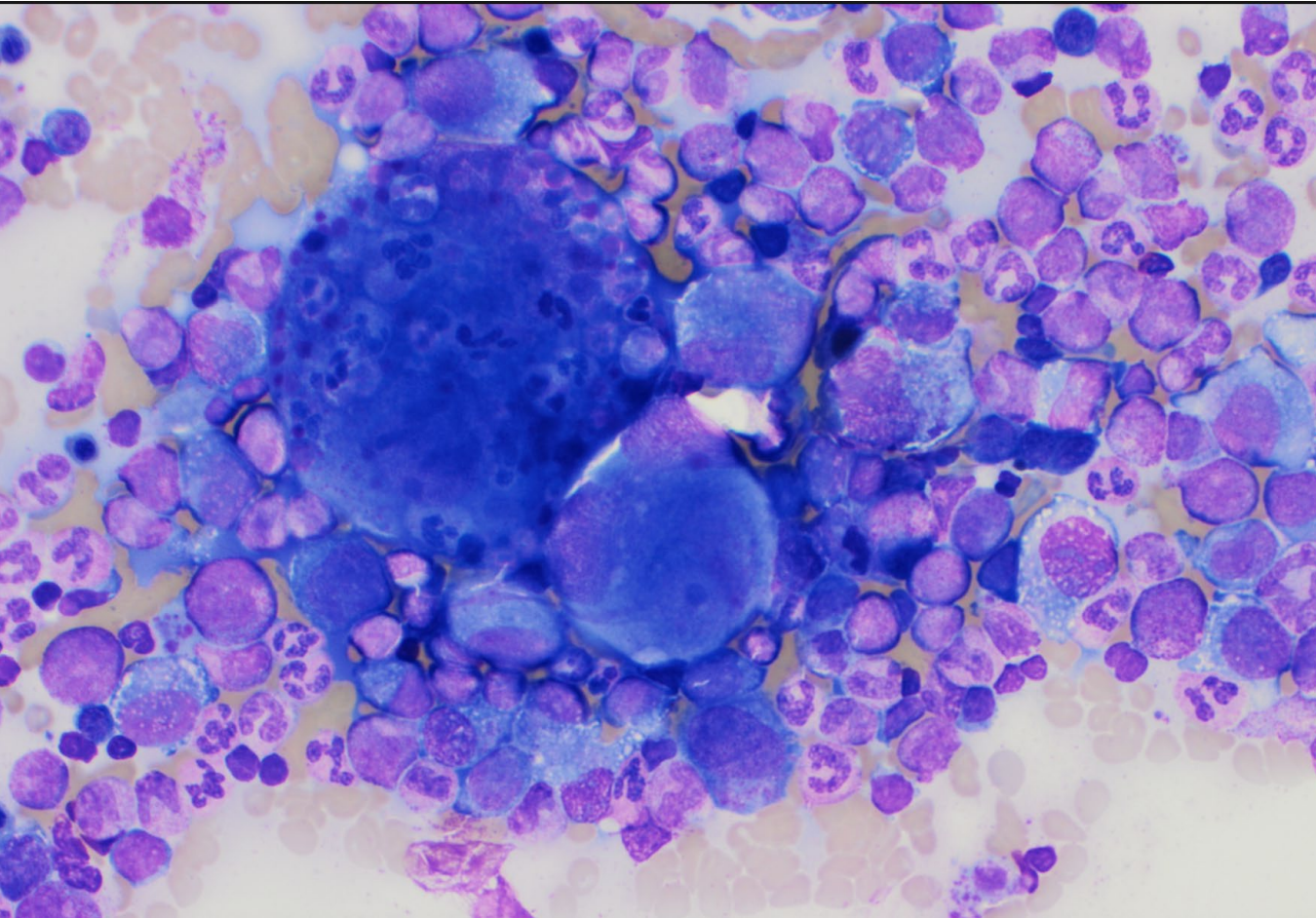


Bone Marrow Aspirate x 40

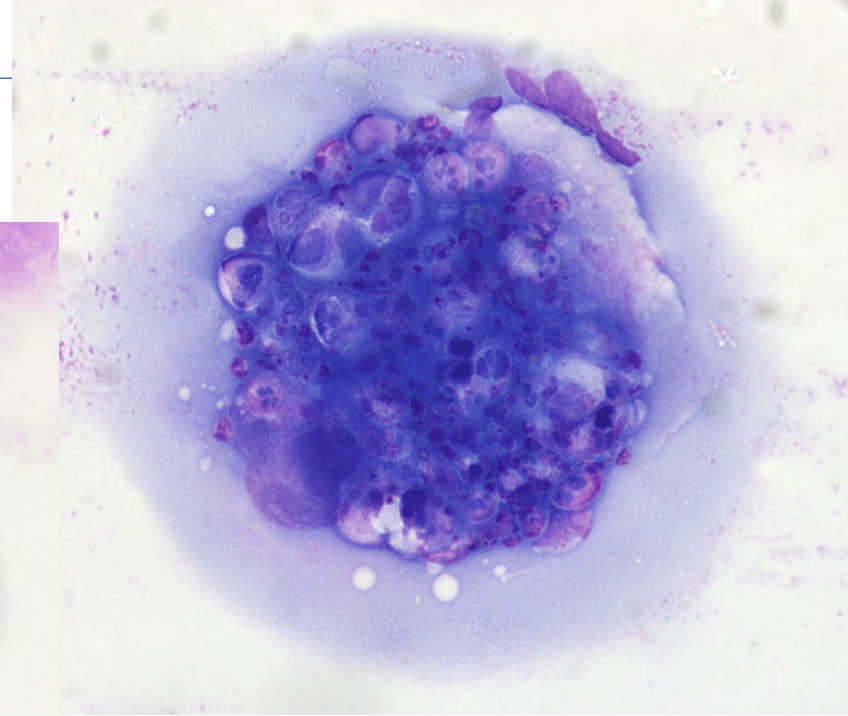
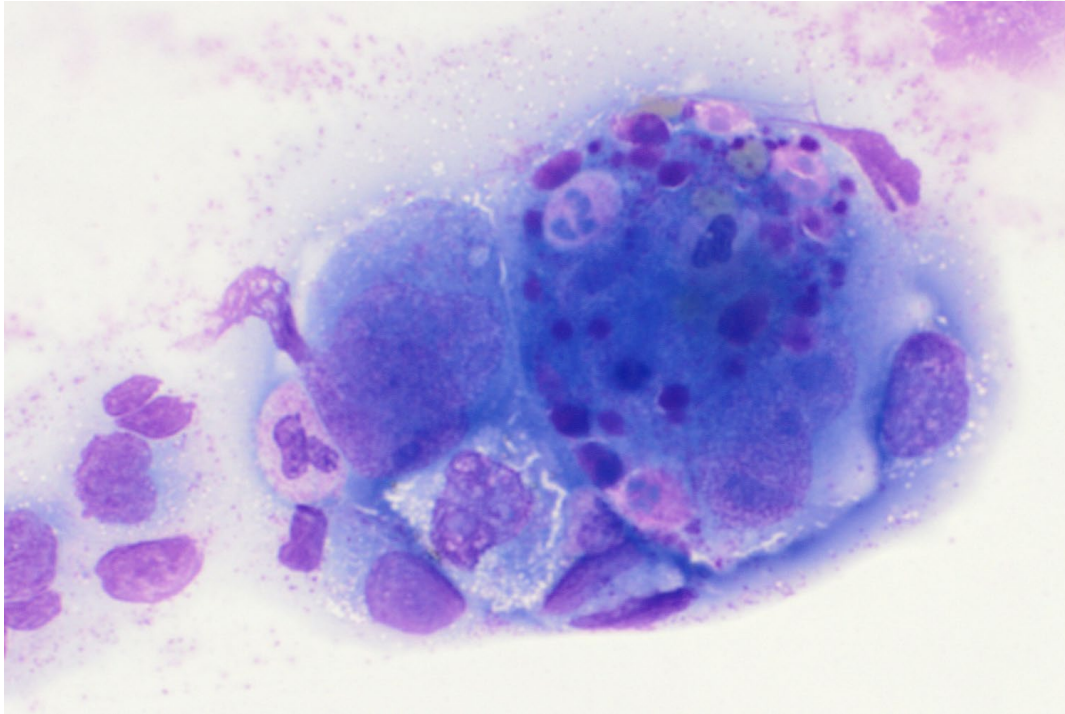




Bone Marrow Aspirate x60



Bone Marrow Aspirate x100



Bone Marrow Aspirate Flow Cytometry

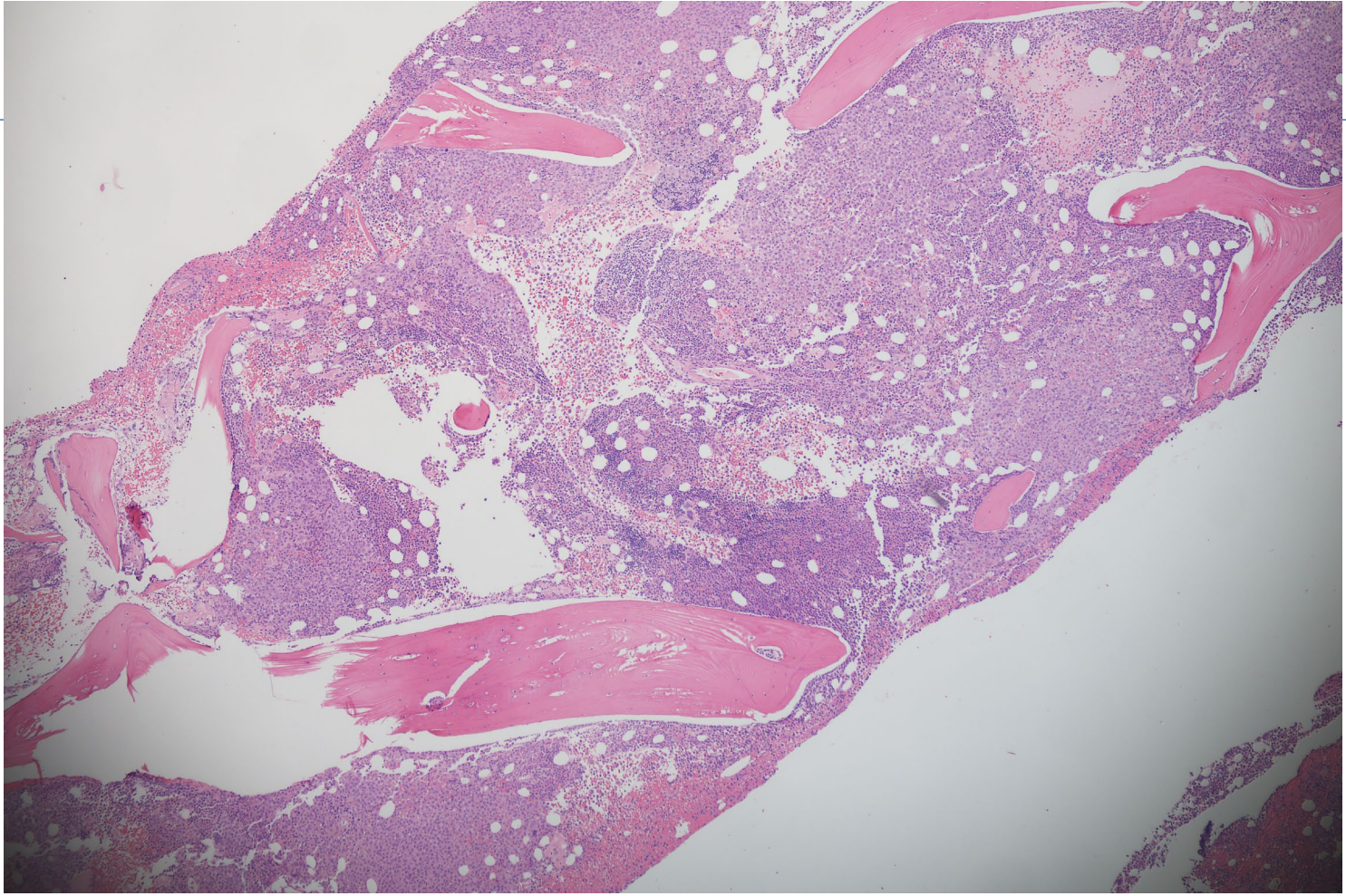
- 1.1% CD19+CD20+ B-cells with skewed Kappa+ light chains.
- The B- cell population is CD20+ CD79b+ CD10+.

What does the bone marrow aspirate show?

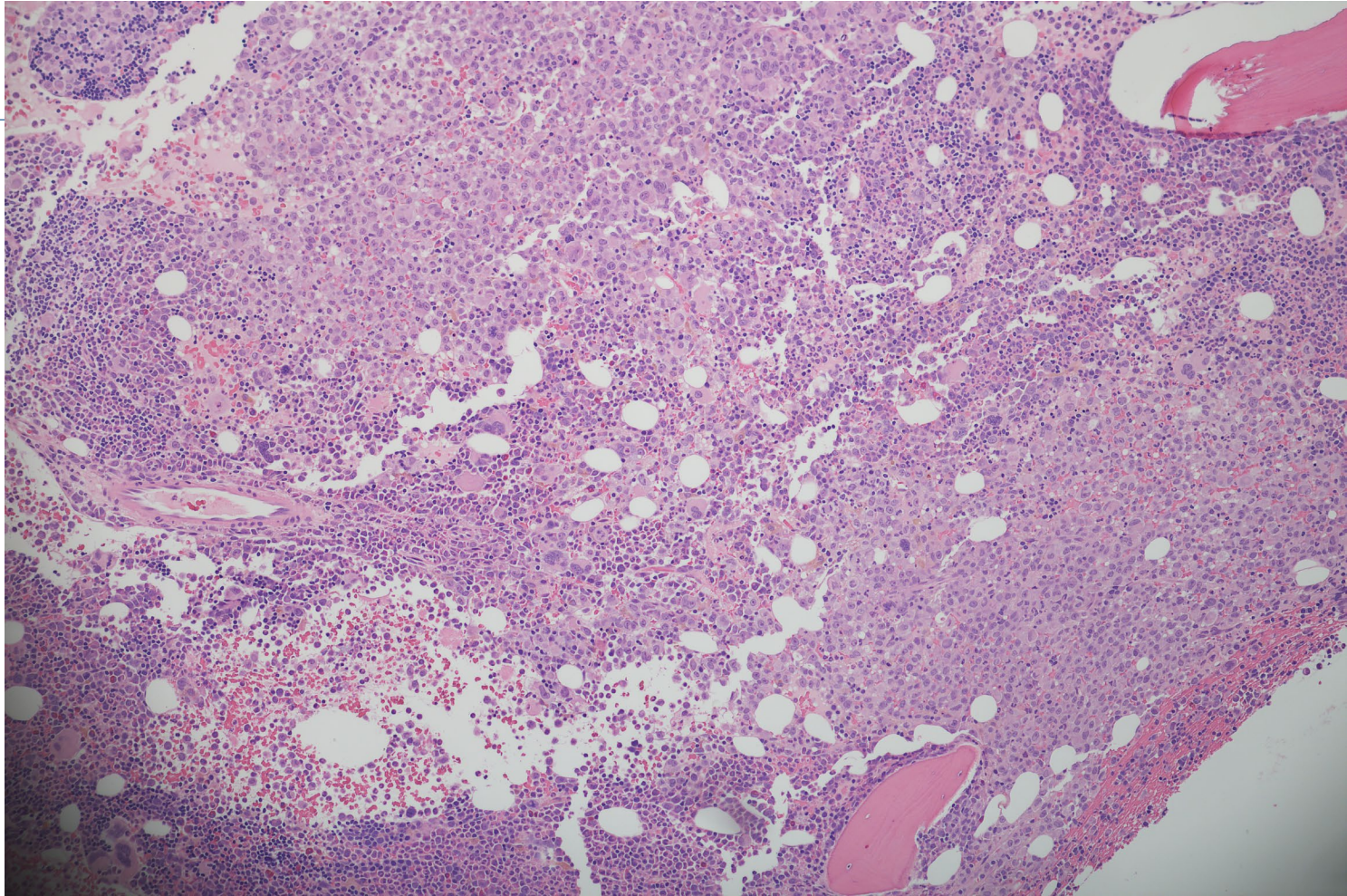
- A) Large atypical B cells
- B) Reed-Sternberg cells
- C) Non-haemopoietic cells
- D) Histiocytes
- E) Small B-cells this is the patient's known Follicular lymphoma
- F) Unable to make a diagnosis yet



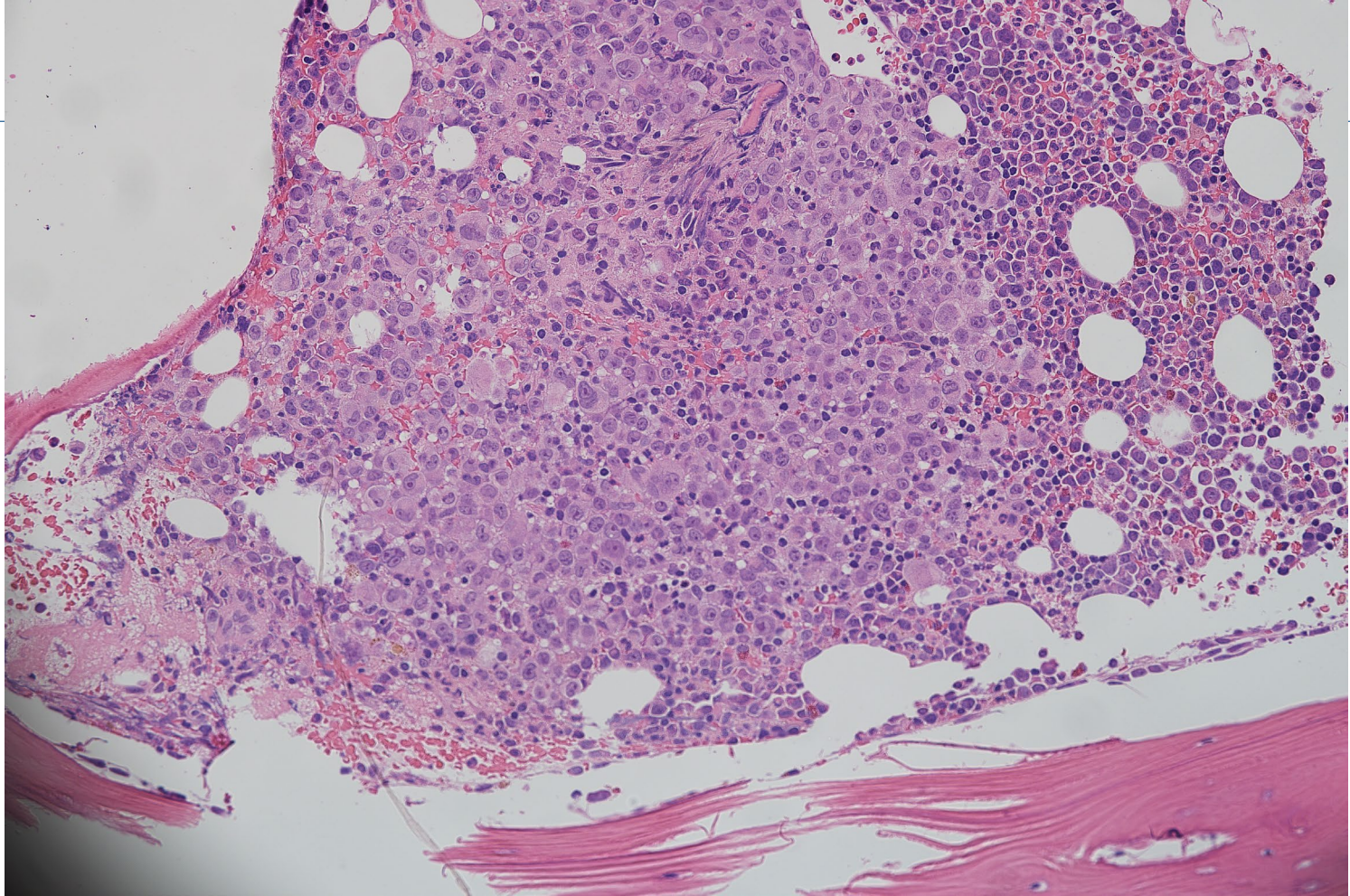
Bone Marrow Trephine x4



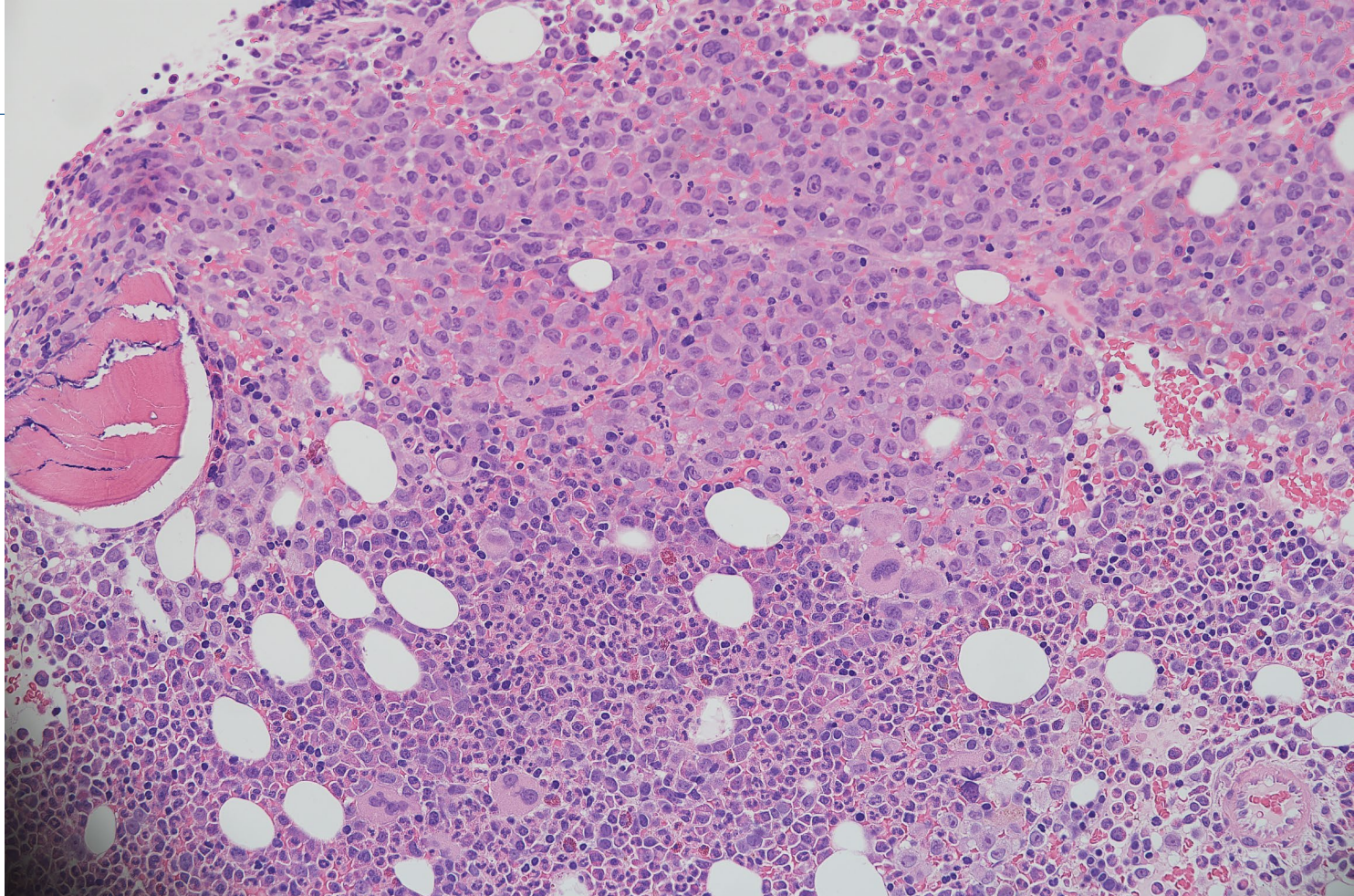
Bone Marrow Trephine x10



Bone Marrow Trephine x20



Bone Marrow Trephine x40



What is the most likely diagnosis?

- A) Follicular lymphoma
- B) Diffuse large B cell lymphoma
- C) Follicular lymphoma transformed to DLBCL
- D) Follicular lymphoma transformed to histiocytic sarcoma
- E) Dual pathology with follicular lymphoma and histiocytic sarcoma



Immunohistochemistry of the Large cell Infiltrate in the Bone Marrow Trephine

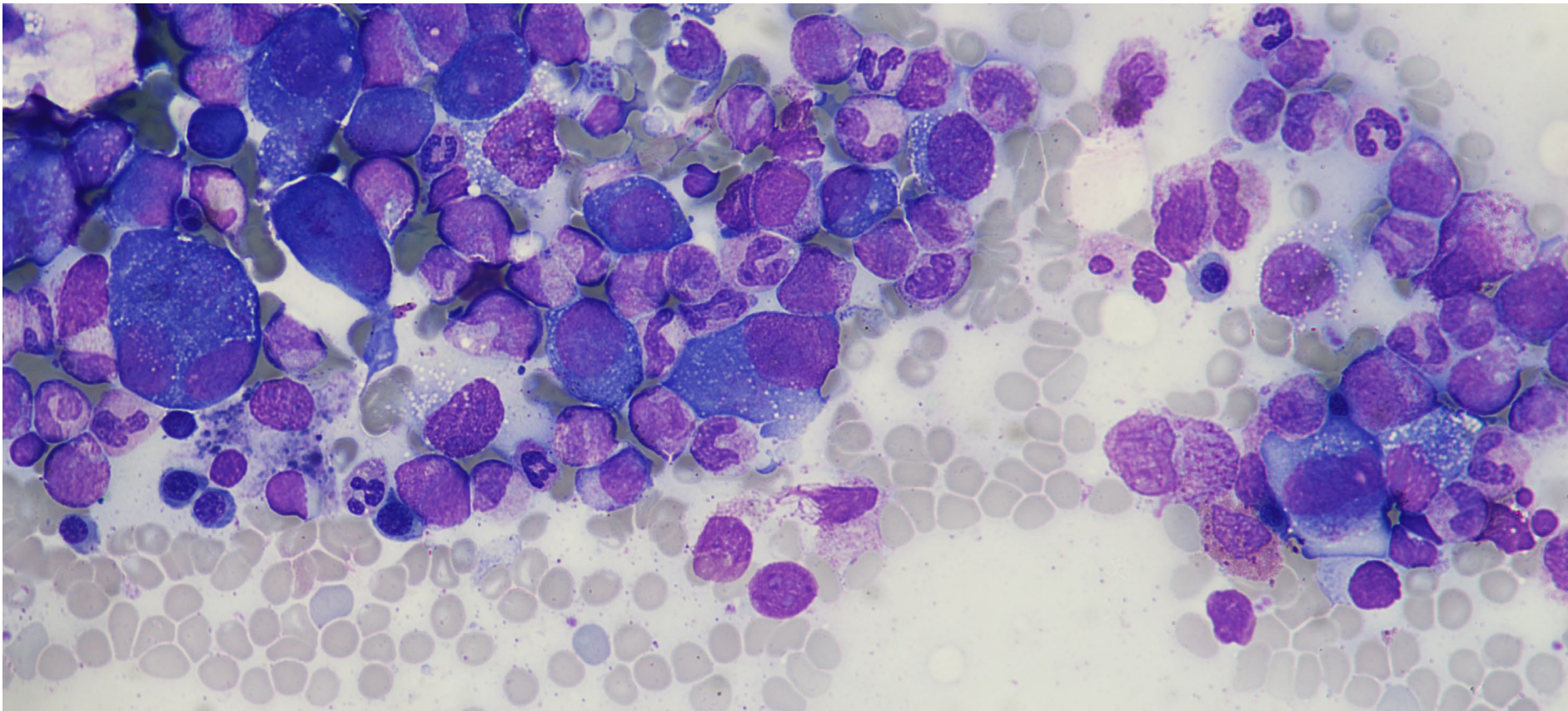
- CD68+
- CD168+
- CD4+
- S100+
- CD1a-, Langerin-

Additional Investigations

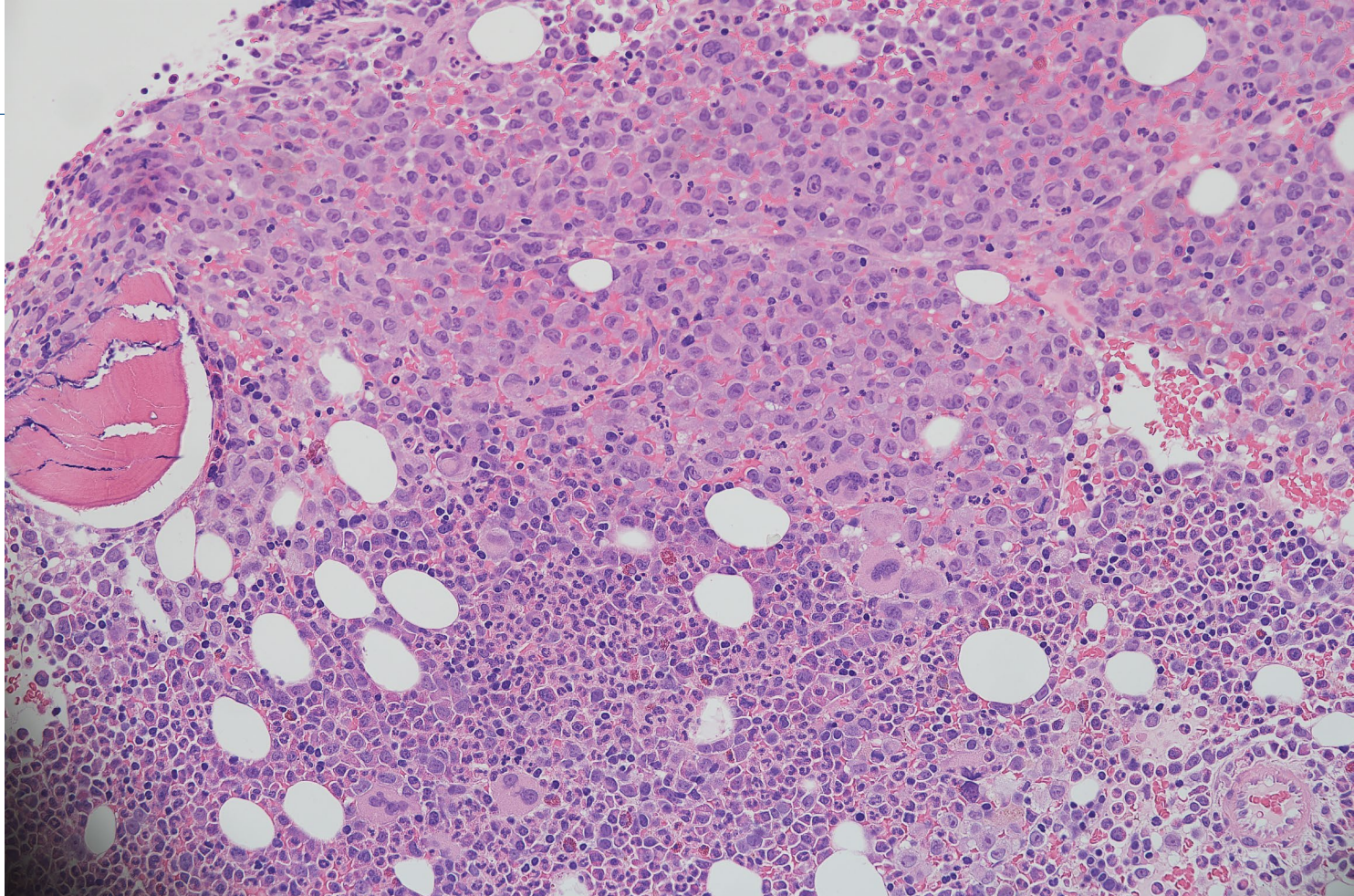
- Clonality studies were performed on the lymph node and clonal IGH rearrangement and 2 IGH translocations including IGH::BCL2 were detected.
- However, the studies were performed on tissue that included the histiocytic disease and the FL components making it is not possible to definitively conclude a clonal relationship, without sequencing on single selected cell.

Diagnosis

- *Concurrent follicular lymphoma and histiocytic sarcoma*
- *This was considered as transdifferentiation of follicular lymphoma to histiocytic sarcoma*
- *However it is difficult to conclusively prove a clonal relationship unless sequencing is performed on selected cells.*



Bone Marrow Trephine x40



Management

- Discussed at the national histiocytic disorders MDT
- CHOP chemotherapy
- Developed TLS, CRS and multi-organ failure leading to his death

Literature & Take-home Messages

- Histiocytic sarcoma is a rare aggressive haemopoietic neoplasm arising from mononucleated phagocytic cells and accounts for less than 1% of haemopoietic tumours ^[1].
- The disease can arise de novo or transdifferentiate from a low-grade B-cell lymphoma such as follicular lymphoma, the diagnosis can be made concurrently or sequentially ^[2].
- The reported cases in the literature show a clonal relationship between the two diagnoses as proven by the detection of BCL2/IGH [t (14;18)] translocation in both malignancies ^[3].

Acknowledgments

- Dr Reem Ahmed- Haematology registrar
- Dr Nisar Ahmed- MIRHO
- Dr Fiona Clark- MIRHO
- Dr Yi Lin Hock- Histopathology consultant, QEH
- Dr Bindu Vydianath- Histopathology consultant, QEH

References

- 1) Yin P. Hung, MD, PhD; Xiaohua Qian, MD, PhD *Arch Pathol Lab Med* (2020) 144 (5): 650–654. <https://doi.org/10.5858/arpa.2018-0349-RS>
- 2) Feldman AL, Arber DA, Pittaluga S, Martinez A, Burke JS, Raffeld M, Camos M, Warnke R, Jaffe ES. Clonally related follicular lymphomas and histiocytic/dendritic cell sarcomas: evidence for transdifferentiation of the follicular lymphoma clone. *Blood*. 2008 Jun 15;111(12):5433-9. doi: 10.1182/blood-2007-11-124792. Epub 2008 Feb 13. PMID: 18272816; PMCID:PMC2424145.
- 3) Sarah Haebe, Debra K. Czerwinski, Anuja Sathe, Susan Grimes, Tianqi Chen, Brock Martin, Hanlee Ji, Ronald Levy, Tanaya Shree; Co-Occurrence of Clonally Related Follicular Lymphoma and Histiocytic Sarcoma. *Blood* 2023; 142 (Supplement 1): 6090. doi: <https://doi.org/10.1182/blood-2023-189712>

Thank you!

