CASE OF THE MONTH
JUNE 2019

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FELLOW ONCOPATHOLOGY
RGCIRC

Clinical History

- 34 year old male, chronic Alcoholic presented with pain in abdomen on and off since 6 month ,increased in severity since last 2 months.
- Abdominal distension, Pedal edema since last 2 months.
- Significant weight loss since last 2 months

Examination

- No pallor
- Icterus present
- No palpable lymphadenopathy
- B / L chest : Clear
- Abdomen distended
- Fluid thrill present.

Investigations:

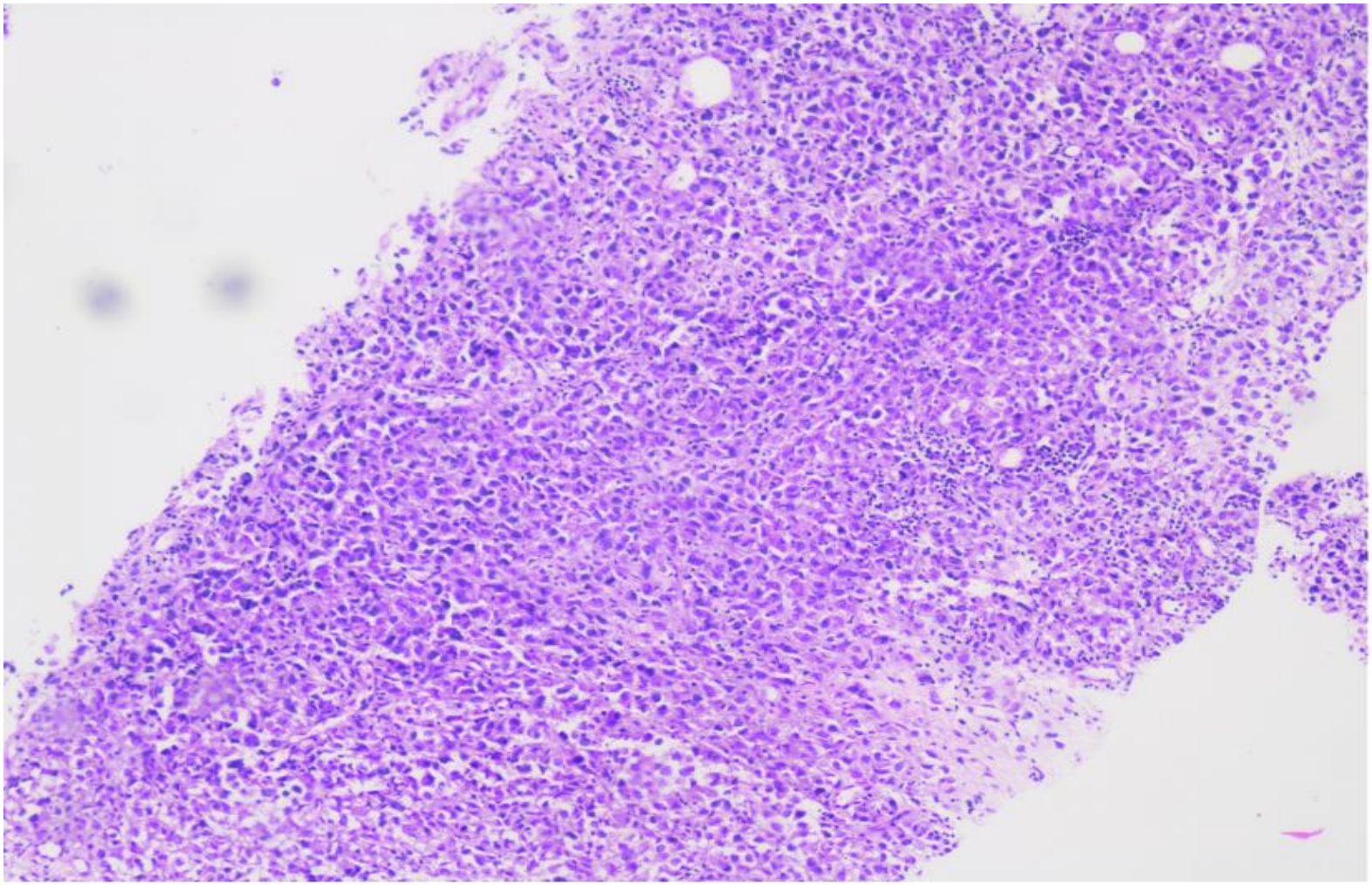
- LDH was raised.
- CECT whole abdomen: Moderate to gross ascites with large omental, mesenteric deposits, largest of size 8 * 4 cm, omental caking, abdominal lymphnodes.
- Ascitic fluid tapping done.
- Ascitic fluid cytology : Immature lymphoid cells suggestive of NHL

- Gene Xpert for TB : negative
- HCV, HBsAg, HIV : negative
- FNA from omental deposits : s/o lymphoproliferative disorder.
- PET –CT : Multiple FDG avid Bilateral cervical, B/L supraclavicular, mediastinal, Abdomino-Pelvic lymph nodes with diffuse FDG avid omental and peritoneal thickening.

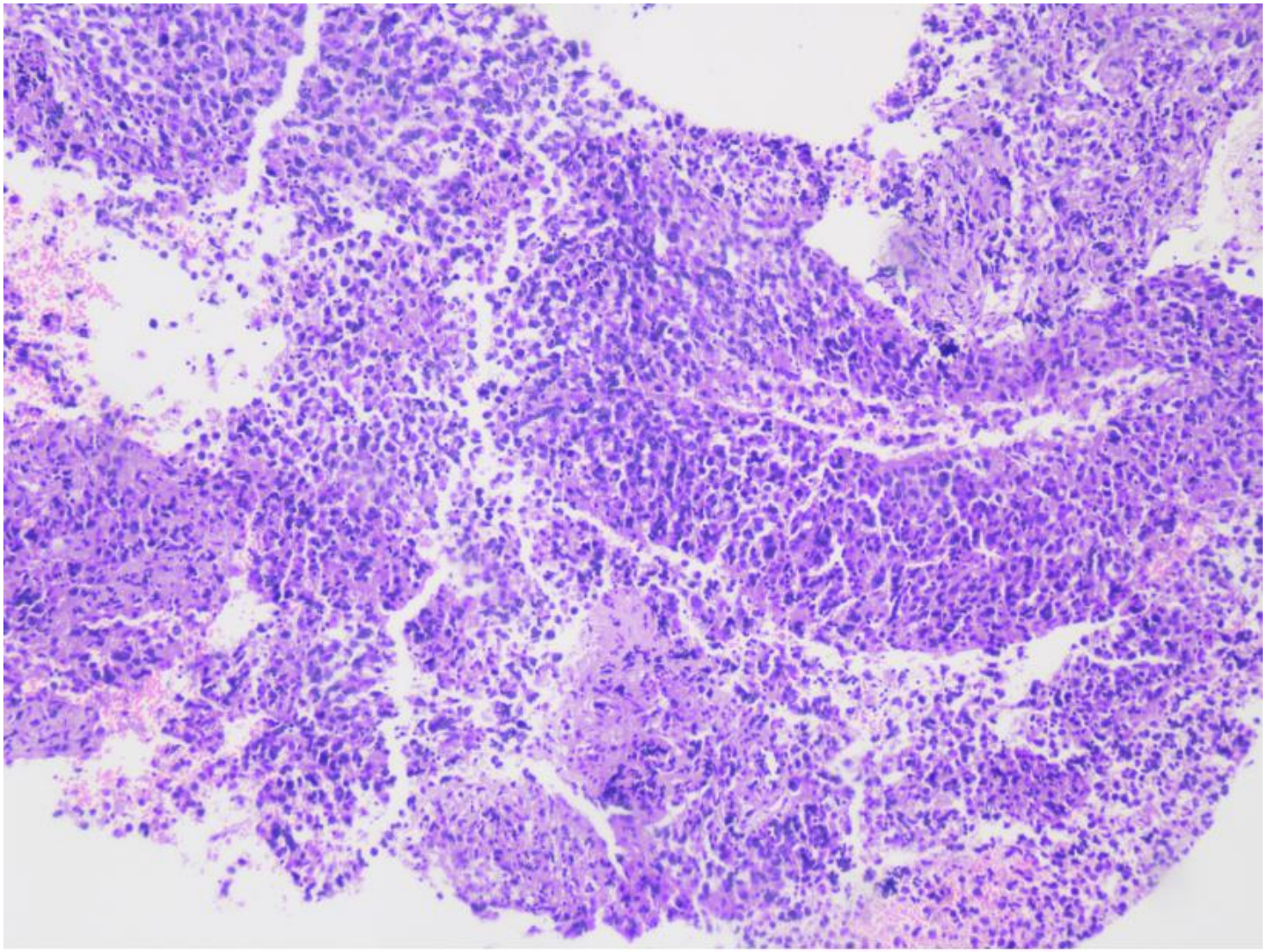
HISTOPATHOLOGY

- **Omental** and **periportal** lymph node biopsy s/o Hematolymphoid malignancy

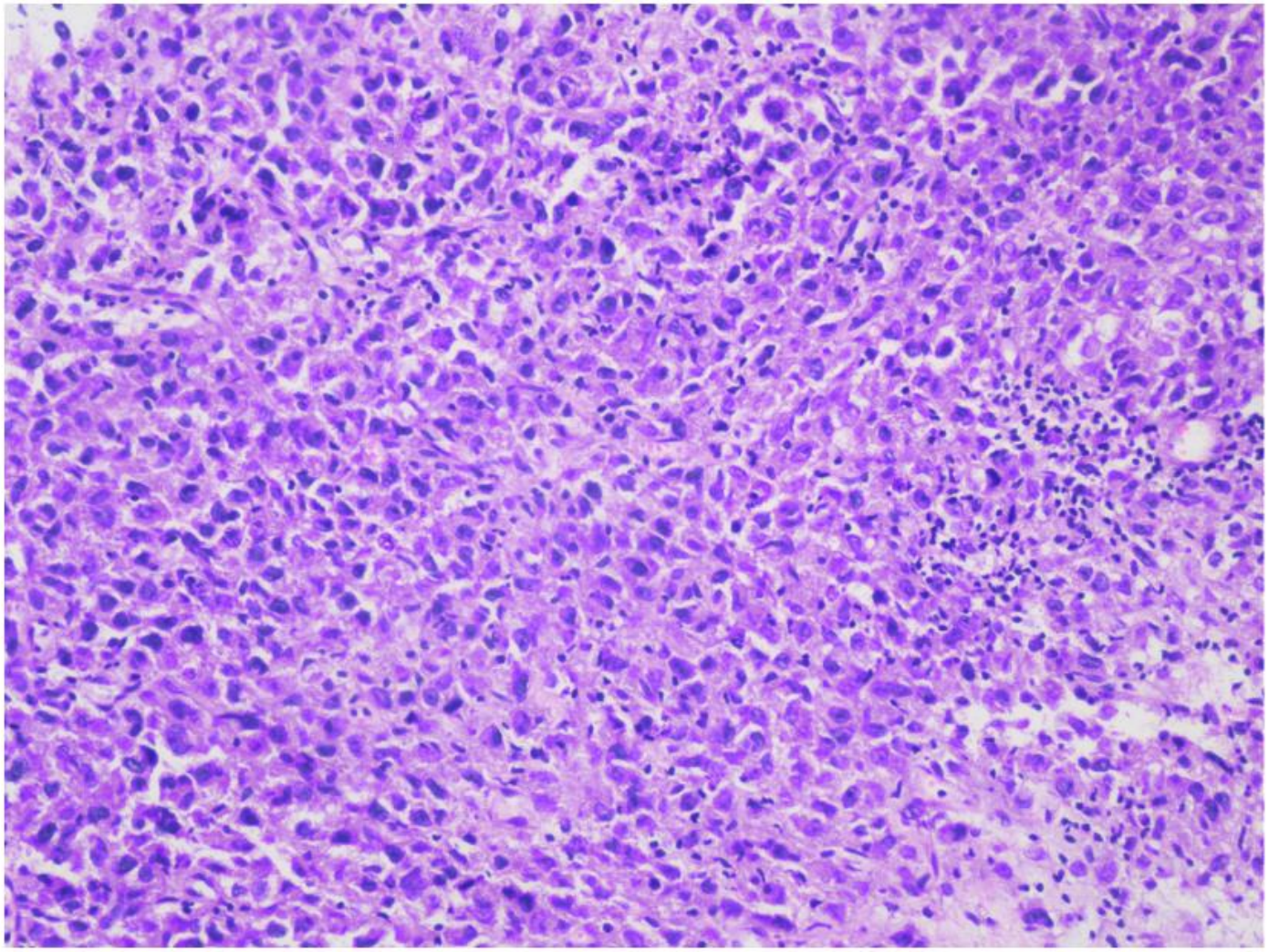
- Microscopic Examination of omental biopsy revealed diffuse sheets of atypical neoplastic cells large in size with vesicular convoluted nuclei prominent nucleoli with brisk apoptotic and mitotic activity.



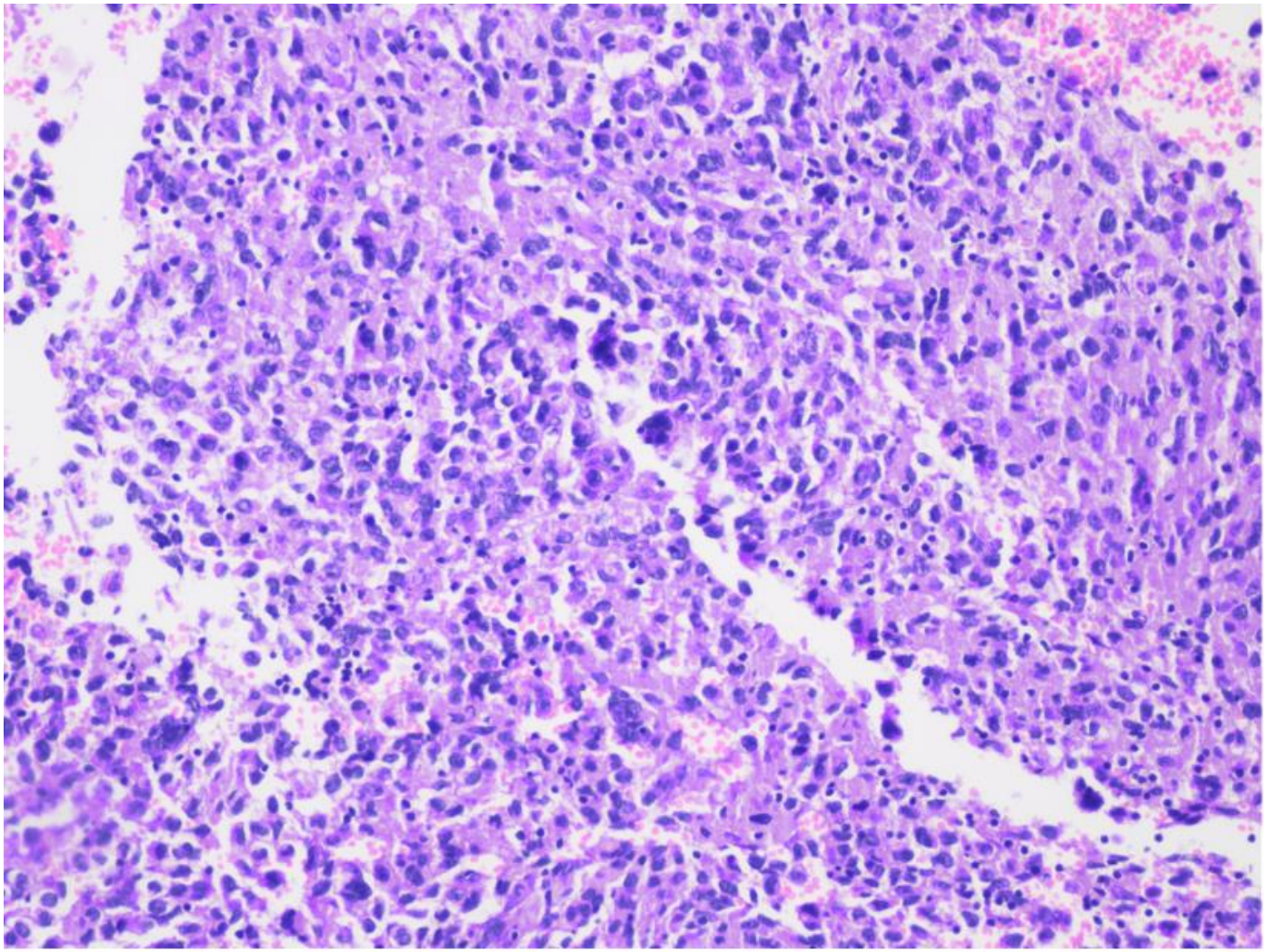
Diffuse sheets of atypical neoplastic cells (100X)



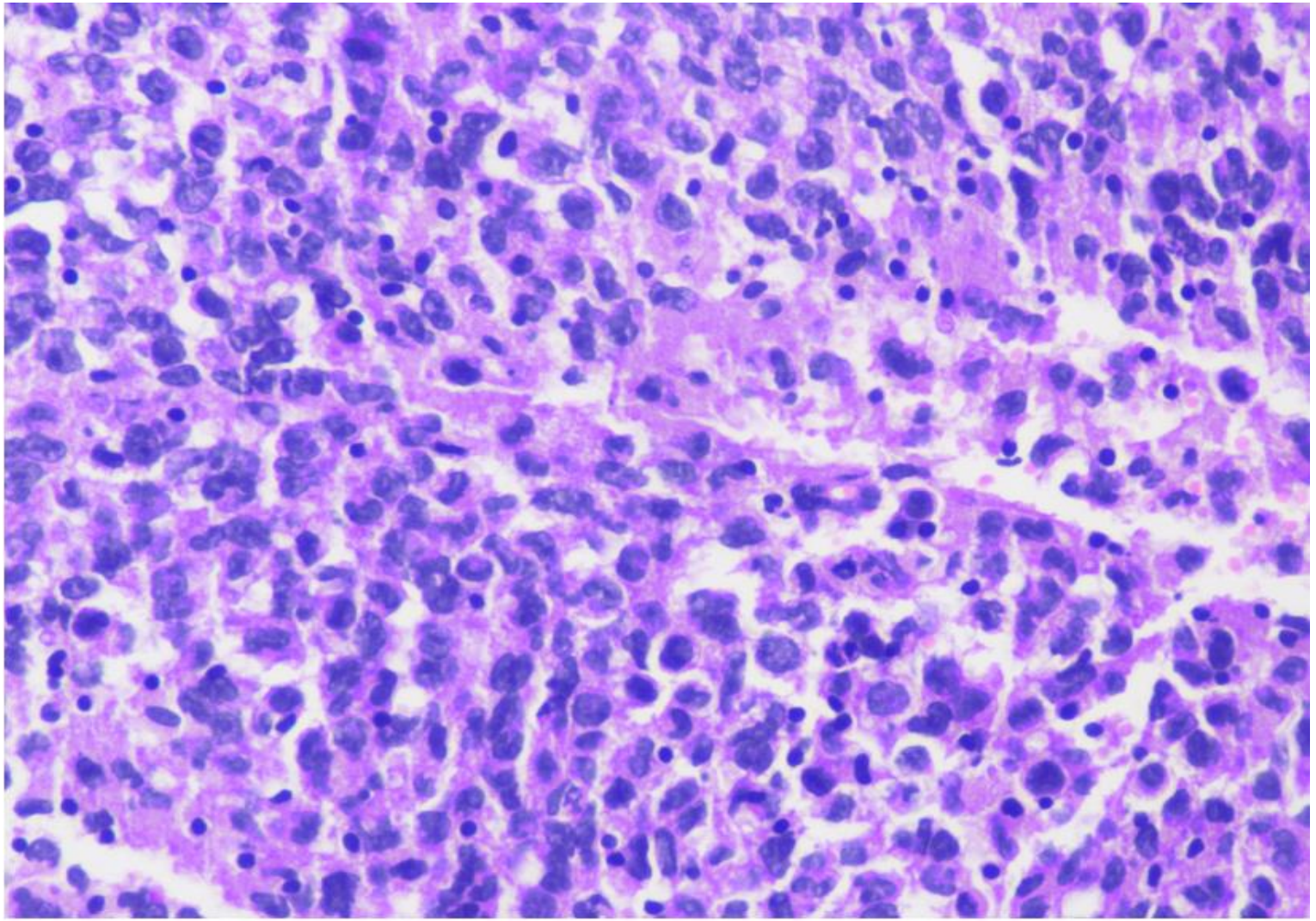
100X



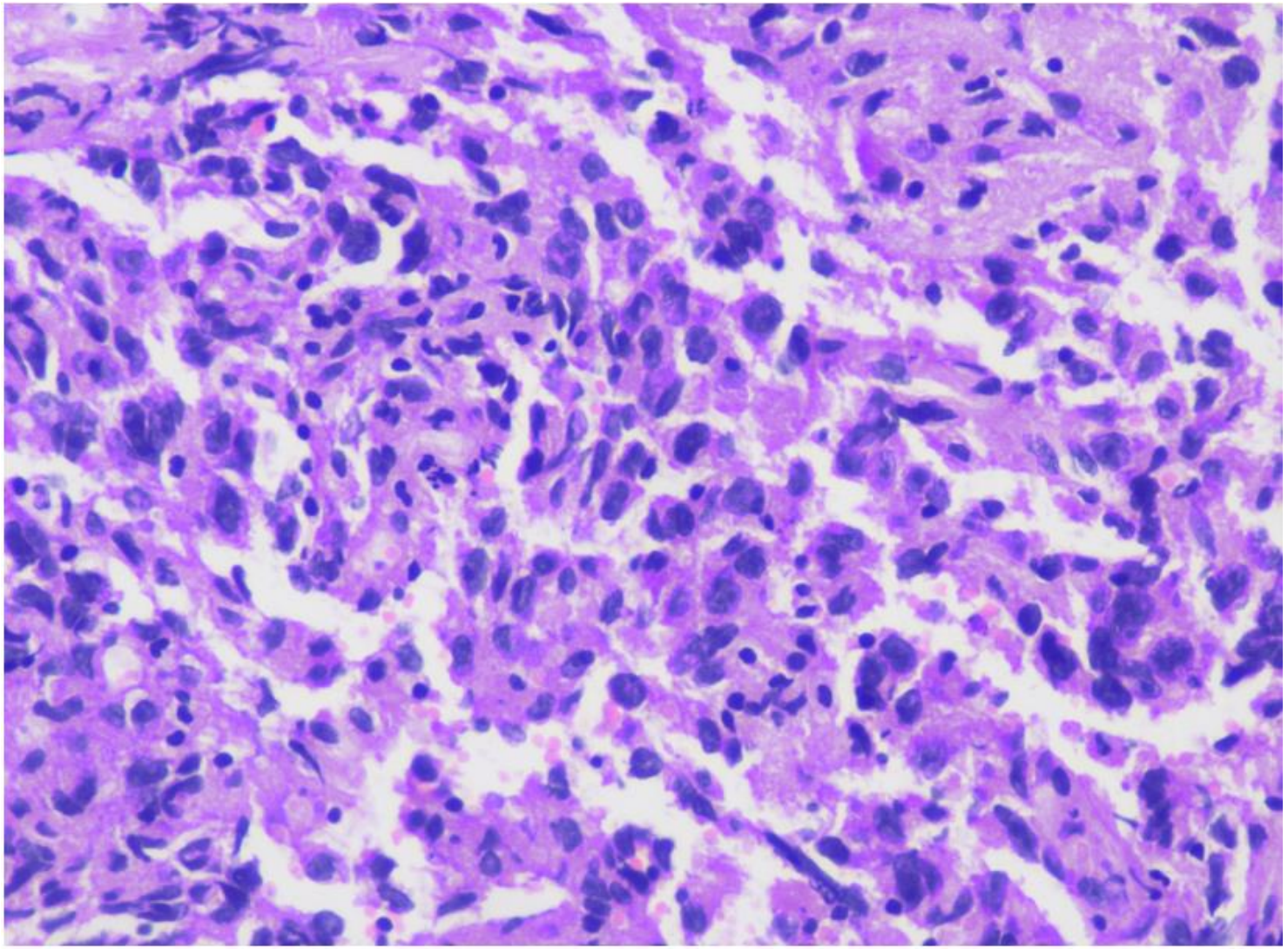
Neoplastic cells are predominantly large with vesicular, convoluted nuclei and prominent nucleoli.(200x)



Brisk apoptotic and mitotic activity noted.(200x)



Neoplastic cells are predominantly large with vesicular, convoluted nuclei and prominent nucleoli. (400x)

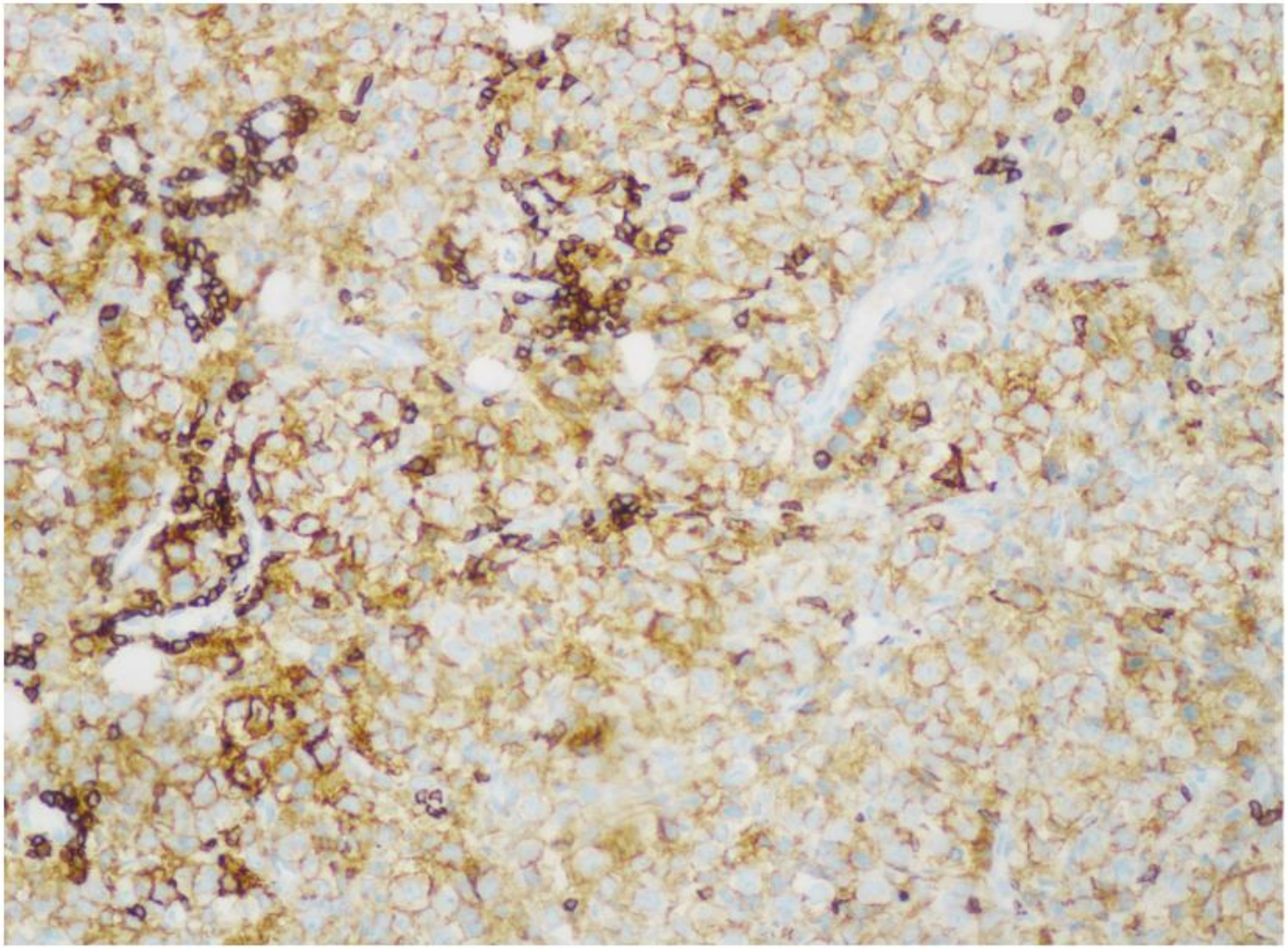


400x

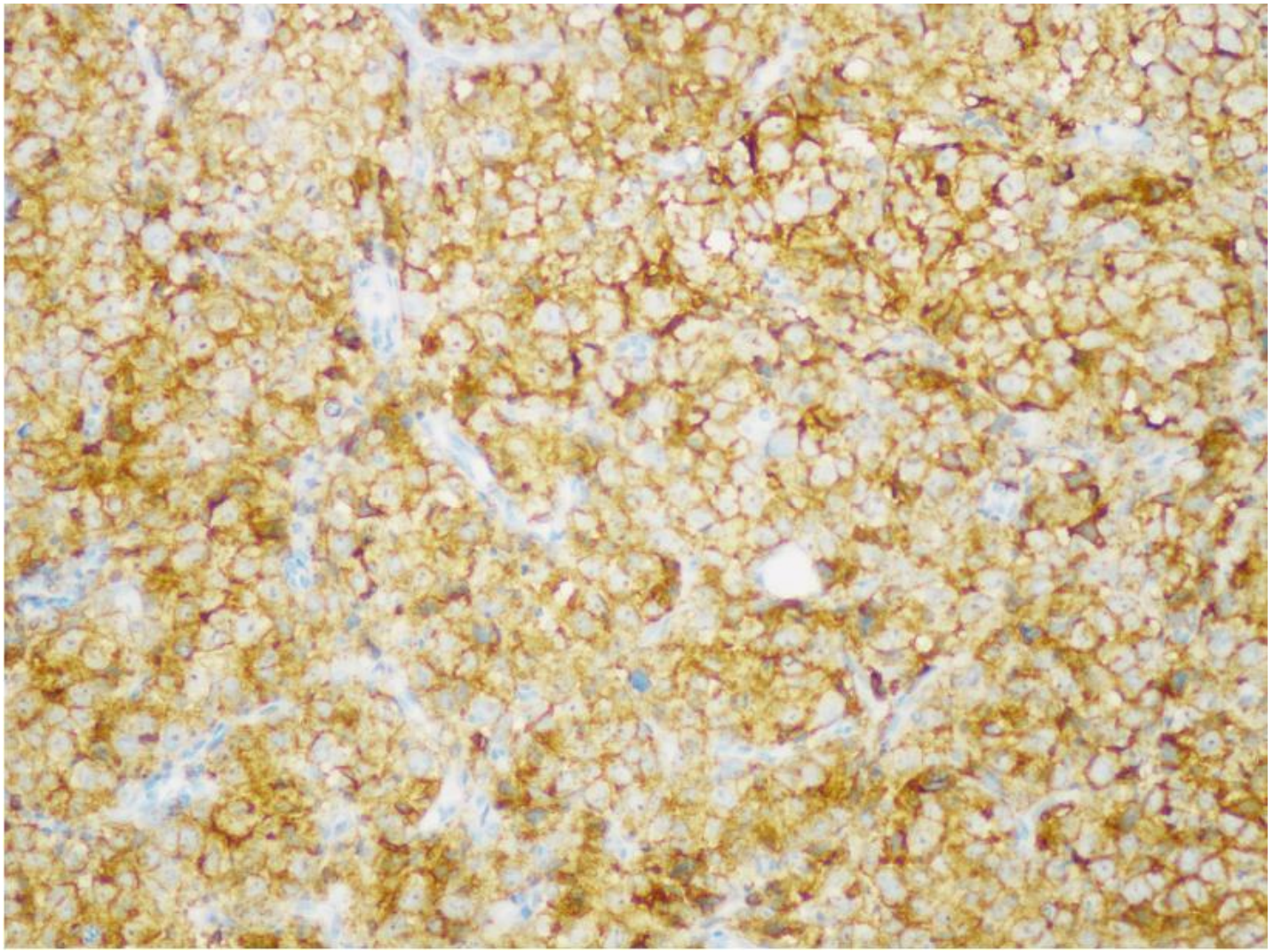
Differential diagnosis

- Metastatic poorly differentiated carcinoma
- Hematolymphoid malignancy
- Melanoma

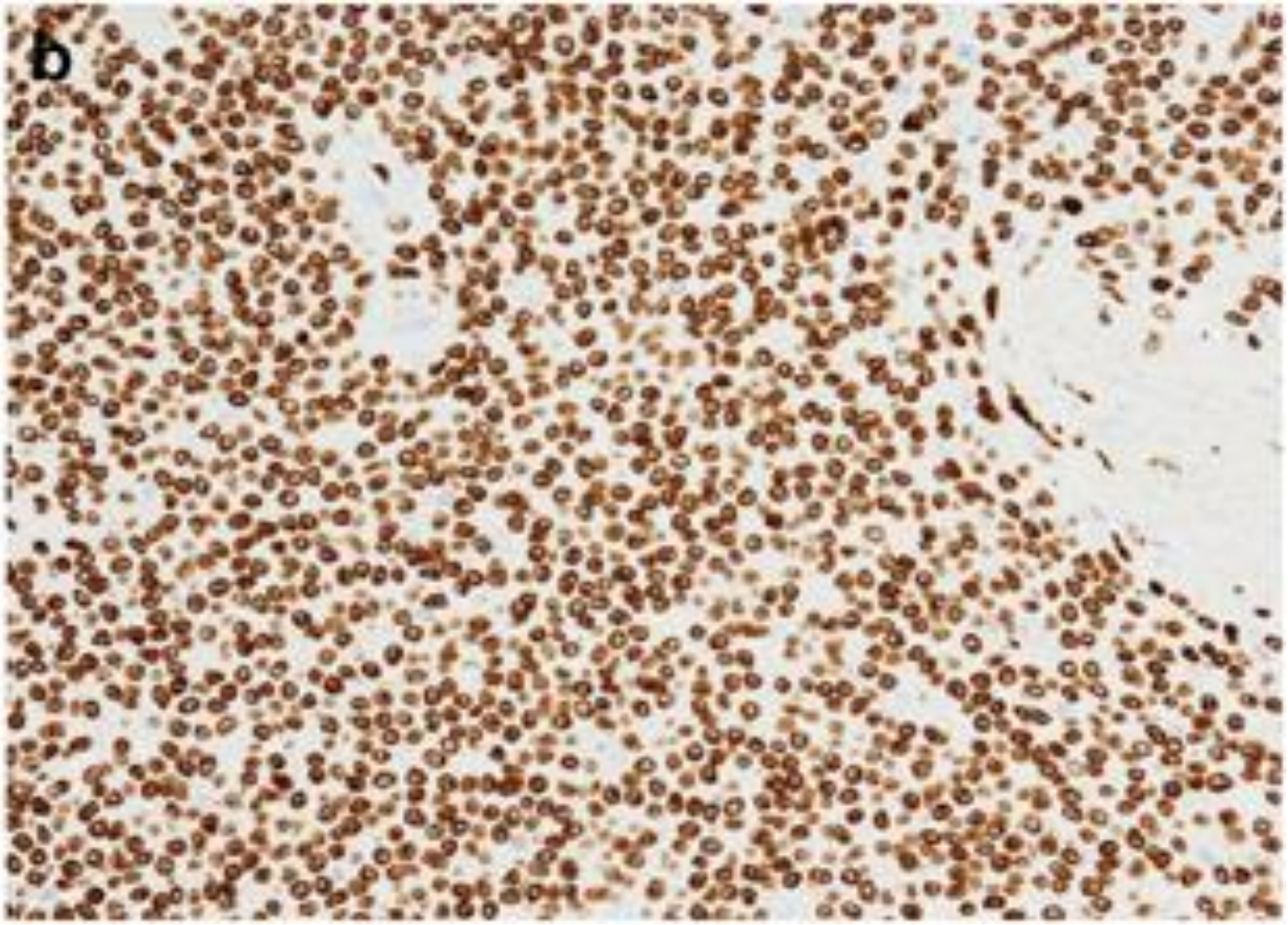
ImmunoHistochemistry



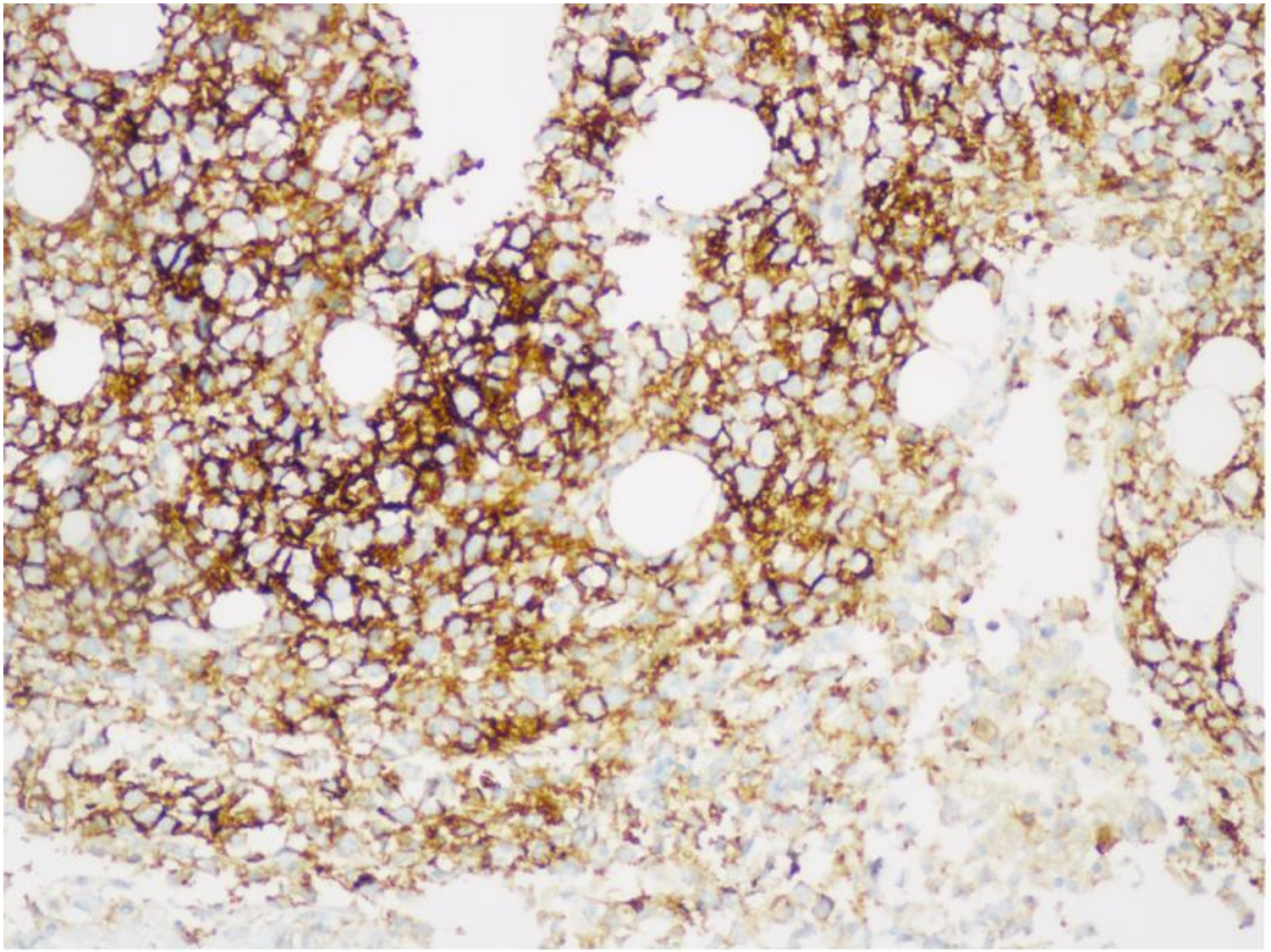
LCA – POSITIVE (400x)



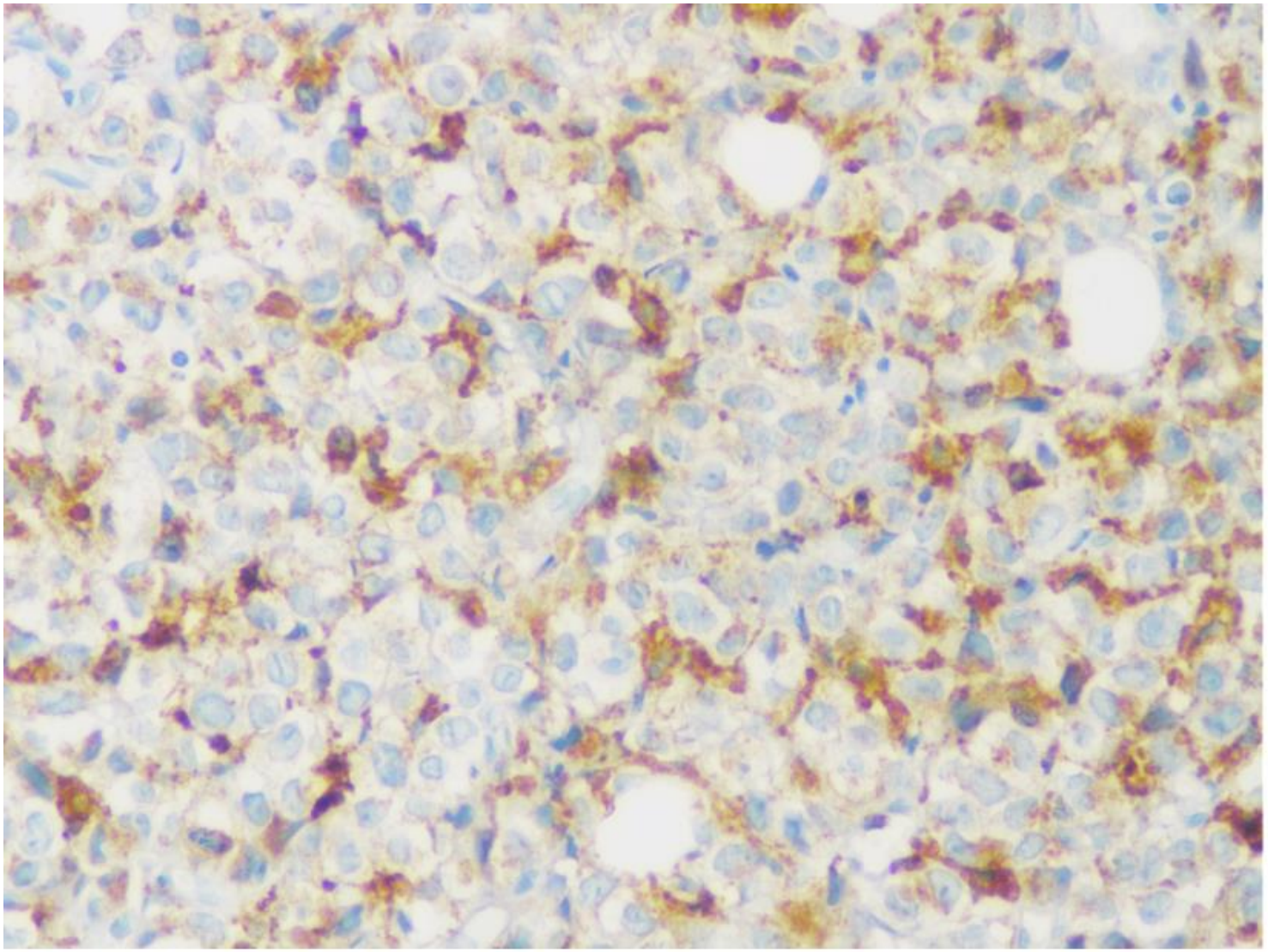
CD4 – POSITIVE (400x)



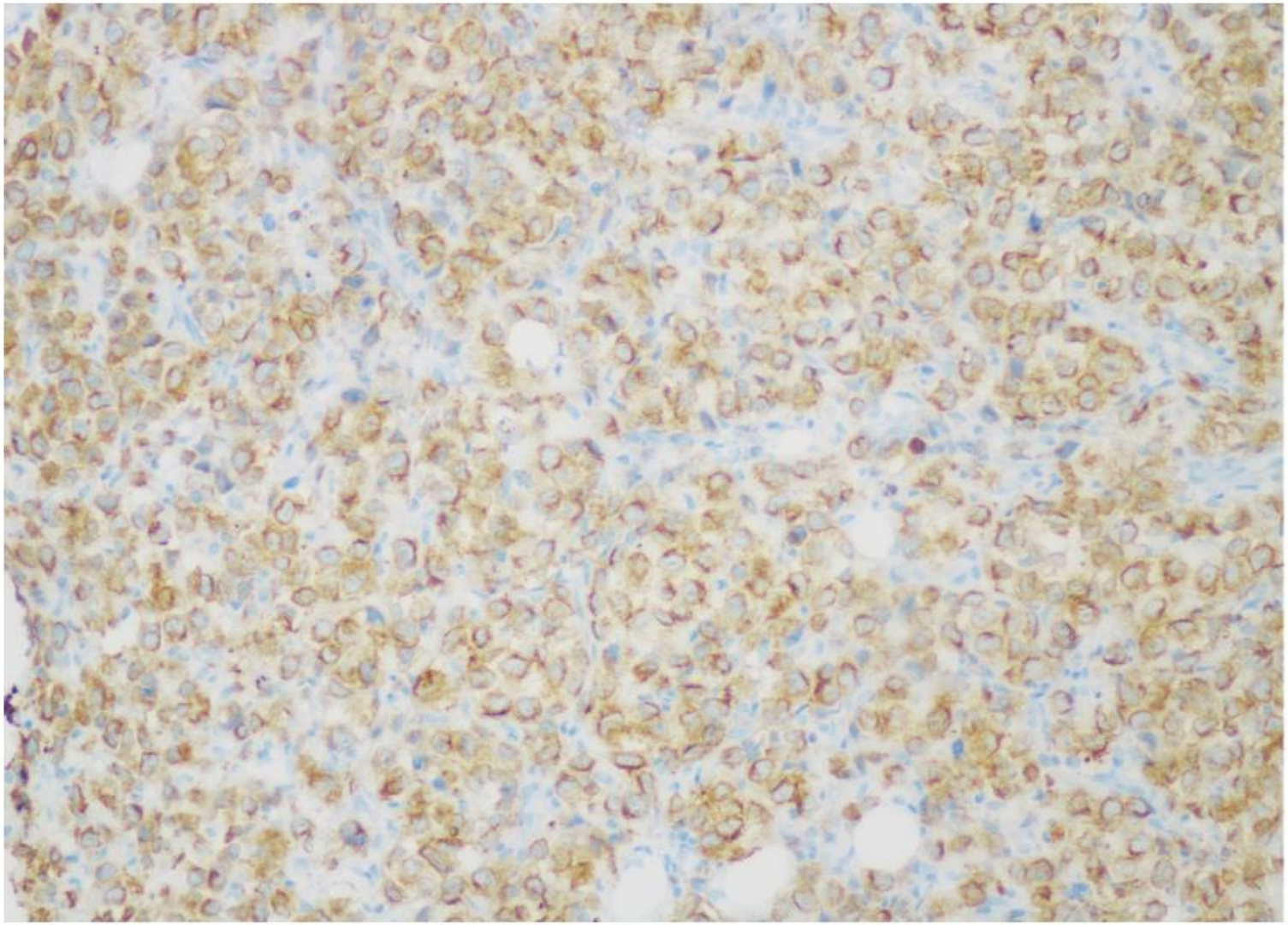
MNDA – POSITIVE (400x)



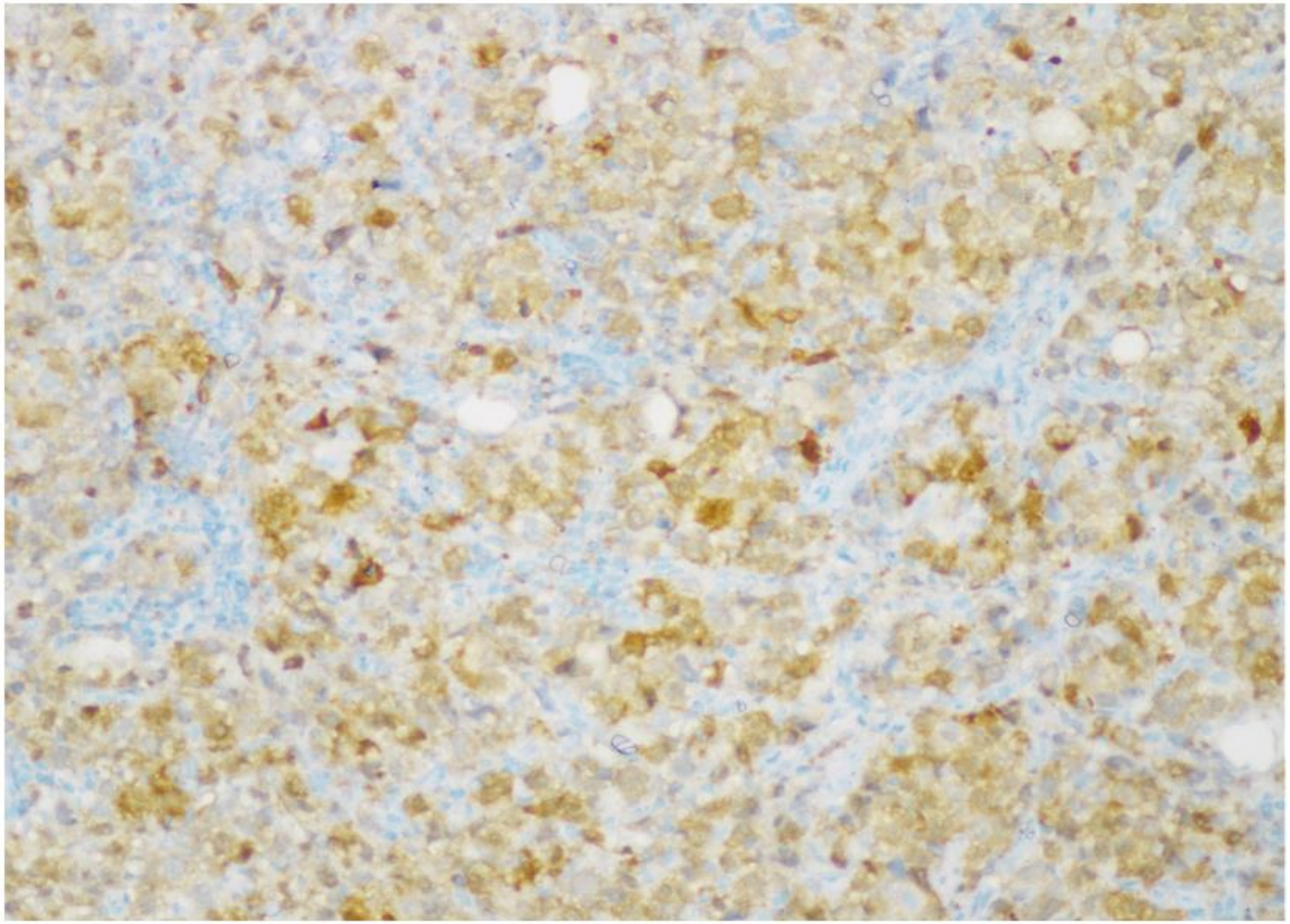
CD43 – POSITIVE (400x)



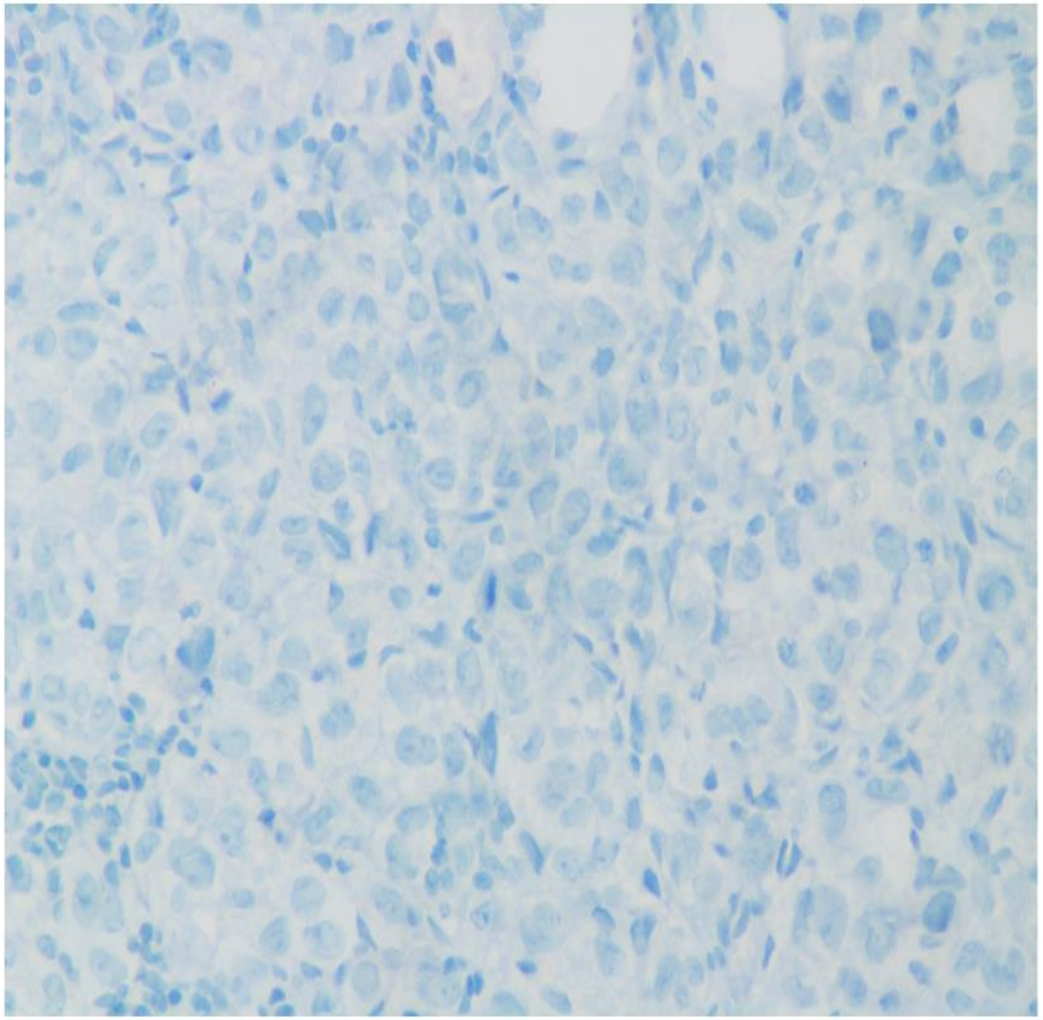
CD68 – POSITIVE (400x)



CK – POSITIVE (400x)



S 100 – POSITIVE (Focal) 400x



CD20, PAX5, CD2, CD3, CD5, CD30, EMA, CD79a, TdT, CD138, C –kit,CD163, CD34, CD123, CD56, CD99, MPO, ALK-1 – NEGATIVE

Summary of IHC

POSITIVE	NEGATIVE
LCA	CD20, PAX5, CD79a
CK	CD2, CD3, CD5, TdT
CD4	CD30, ALK-1
CD43	EMA, c-Kit, CD34, CD99, CD56, MPO
MNDA	CD138, CD163, CD123
CD68	
S 100 (Focal)	

Flow cytometry

- Flow cytometry examination was done on ascitic fluid malignant cells

- Antibodies used were : CD45 , CD5,CD7, CD33, CD117, CD64, HLA-DR, CD16, CD56, CD22,CD34a

ANALYSIS

- There was marked cellular degeneration
- Cells were gated in bright CD4 region which showed positivity for CD45, CD4,CD33, CD13, CD64 and negative for CD3, CD16, CD56, CD117, CD22,CD34, CD5, CD7

- Bone marrow aspiration and Bone marrow biopsy : no involvement.
- Peripheral smear examination was normal with no blasts.

Final diagnosis

- **Based on Overall histopathological features along with IHC and flow cytometry findings, final diagnosis of Myeloid sarcoma with monocytic differentiation was made.**

Discussion

- Myeloid sarcoma (MS) is a rare disease entity identified as a variety of manifestations defined by the occurrence of extramedullary myeloid cell masses with or without bone marrow involvement.

- Byrd *et al.* examined all known MS cases in the literature as of 1995 and found that of the 154 published isolated MS cases, 46% had initial erroneous diagnoses often mislabeled as large cell lymphoma.

J Clin Oncol. 1995 Jul; 13(7):1800-16.

- Mature or immature types of MS can be confused with Hodgkin lymphoma, T-cell lymphomas, extramedullary hematopoiesis (myeloid metaplasia), or infectious processes.
- Additionally, blastic types of MS can be readily confused with non-Hodgkin lymphoma, lymphoblastic lymphoma, poorly differentiated carcinoma, or melanoma.

Guidelines for diagnosing myeloid sarcoma (MS).

Differential diagnosis	– Mature or immature types of MS: Hodgkin lymphoma, T-cell lymphoma; extramedullary hematopoiesis (myeloid metaplasia), or infectious processes; – Blastic type of MS: non-Hodgkin's lymphoma, lymphoblastic lymphoma; poorly differentiated carcinoma, melanoma
Anatomical locations	Varied
Associated hematological malignancies	Acute myelogenous leukemia; chronic myelogenous leukemia; multiple myeloma; myelodysplastic syndrome; myelofibrosis
Histology (varied)	Mature and immature myelocytes; blasts; lack of bileneage or trileneage differentiation; extensive infiltration of surrounding tissue, or quite distinct
Cytochemistry: recommended stains	Myeloperoxidase; lysozyme; naphthol AS-D chloroacetate esterase; non-specific esterase
Immunophenotyping: recommended markers	– Most common: CD43 and CD68/KP1 – Other common: CD4, CD15, CD30, CD34, CD56, CD99, CD117, tdt, Glycophorin A, CD61/linker of activated T-lymphocyte/von-Willebrand antigen – Myeloid markers: CD68/KP1, CD117 – Monocytic markers: CD56, CD68, CD163 – T-cell markers: CD3, CD4, CD43, CD45, LCA – B-cell markers: CD20, CD79a
Cytogenetics: recommended evaluations should include	– Evaluate for monosomies, trisomies, translocations, and inversions: monosomy 7, monosomy 16, trisomy 8, trisomy 11, t(8;21)(p22;q22), inversion 16 – Particular deletions: 16q, 5q, 20q – Mutations commonly associated with acute myeloid leukemia: NPM1, FLT3-ITD

Hagen, P. A., Singh, C., Hart, M., & Blaes, A. H. (2015). Differential Diagnosis of Isolated Myeloid Sarcoma: A Case Report and Review of the Literature. *Hematology reports*, *7*(2), 5709. doi:10.4081/hr.2015.5709

- The most common sites of presentation include the skin, lymph node, soft tissue, and bone;however, nearly all anatomical locations have been described including the ovaries, myocardium, pineal body, and other locations.

- Most patients who are diagnosed with isolated MS develop traditional AML with bone marrow involvement within five to nine months, but some develop AML up to 9 years after the initial MS diagnosis

- CD68/KP1 and CD43 are the most commonly encountered makers, both present in 75-100% of cases but the non-specificity of CD43 in particular should be noted.

- Once a diagnosis of MS has been made, moving quickly to induction therapy is important because median survival is quite poor ranging from 1-2 years but as low as 8 months.