Case of the Month

MAY 2020

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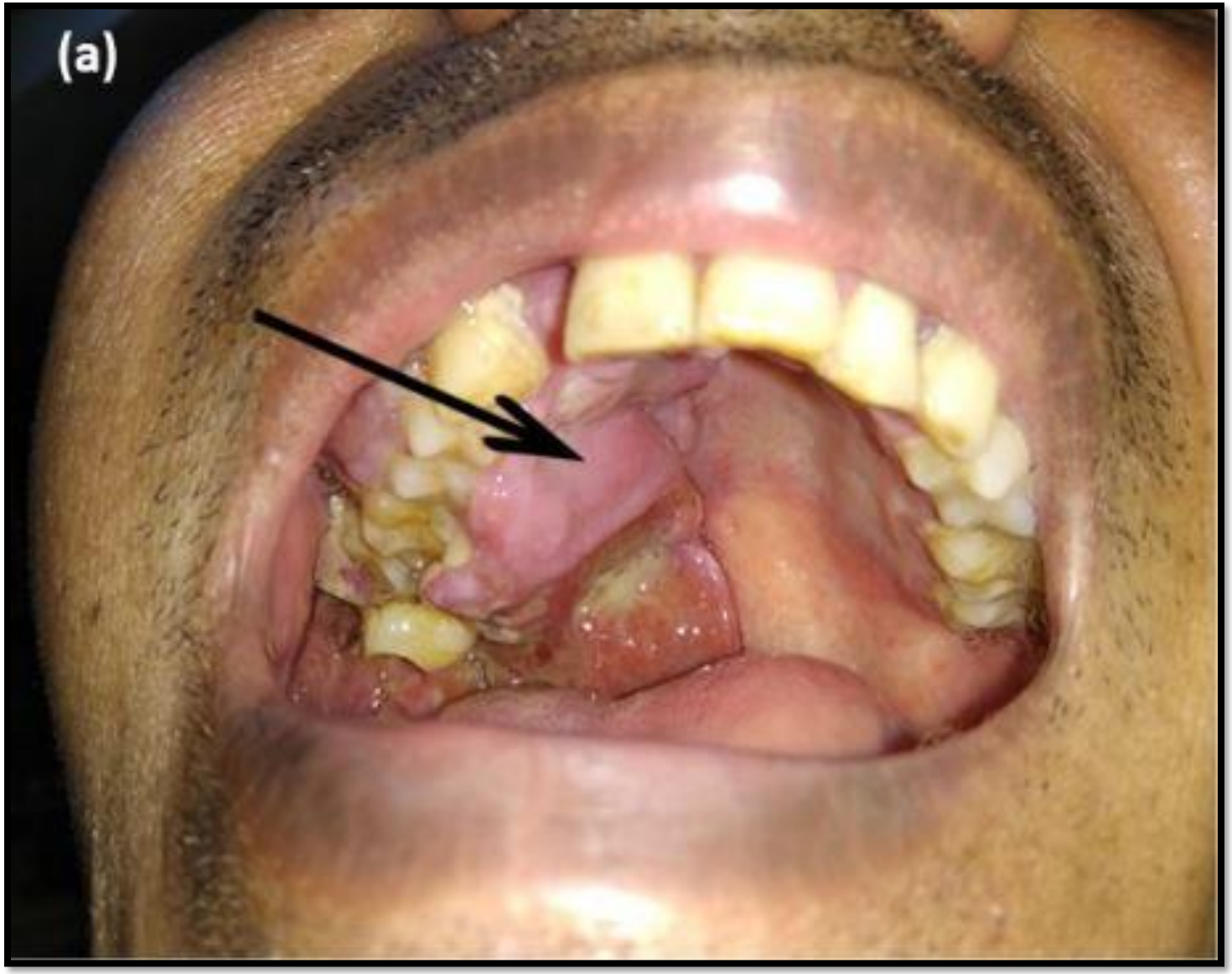
Case

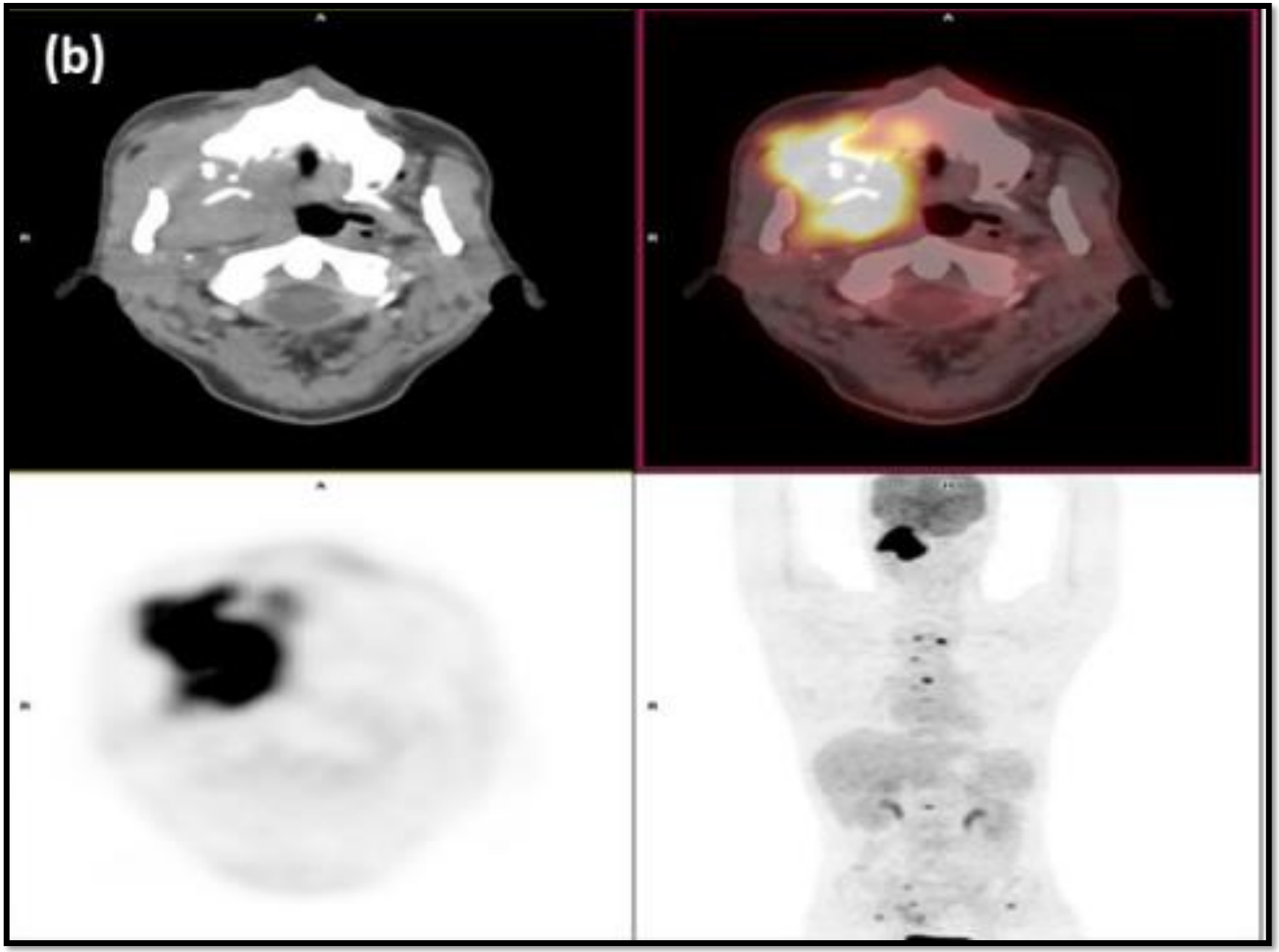
- A 44-year-old male, a chronic tobacco chewer with no significant history, presented with complaints of ulcer in the right upper alveolus for 1 month.

- On examination, an ulcero-proliferative growth was noted involving the right-side upper alveolus/hard palate reaching upto midline and extending to tonsillar pillar and GB sulcus.



Ulcero-proliferative involving the right-side upper alveolus and hard palate

Imaging



Findings

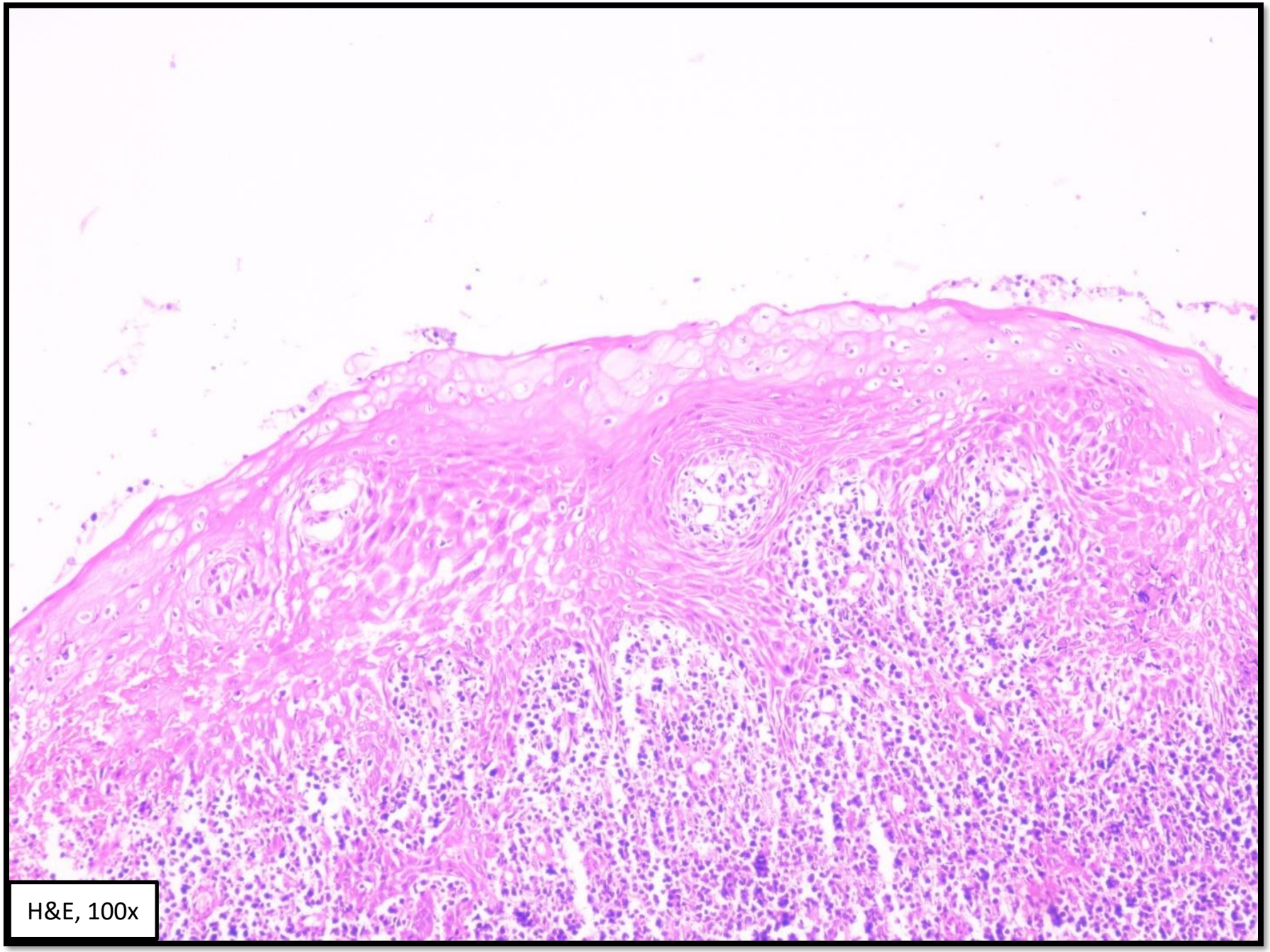
- PET-CT showed FDG avid osteolytic lesion with large soft tissue component, measuring 6.8x4.9x5.8cm involving right maxillary sinus.

- The mass was seen infiltrating the masticator space, pterygoid space and muscle, soft palate, RMT, right buccal mucosa and GB sulcus.

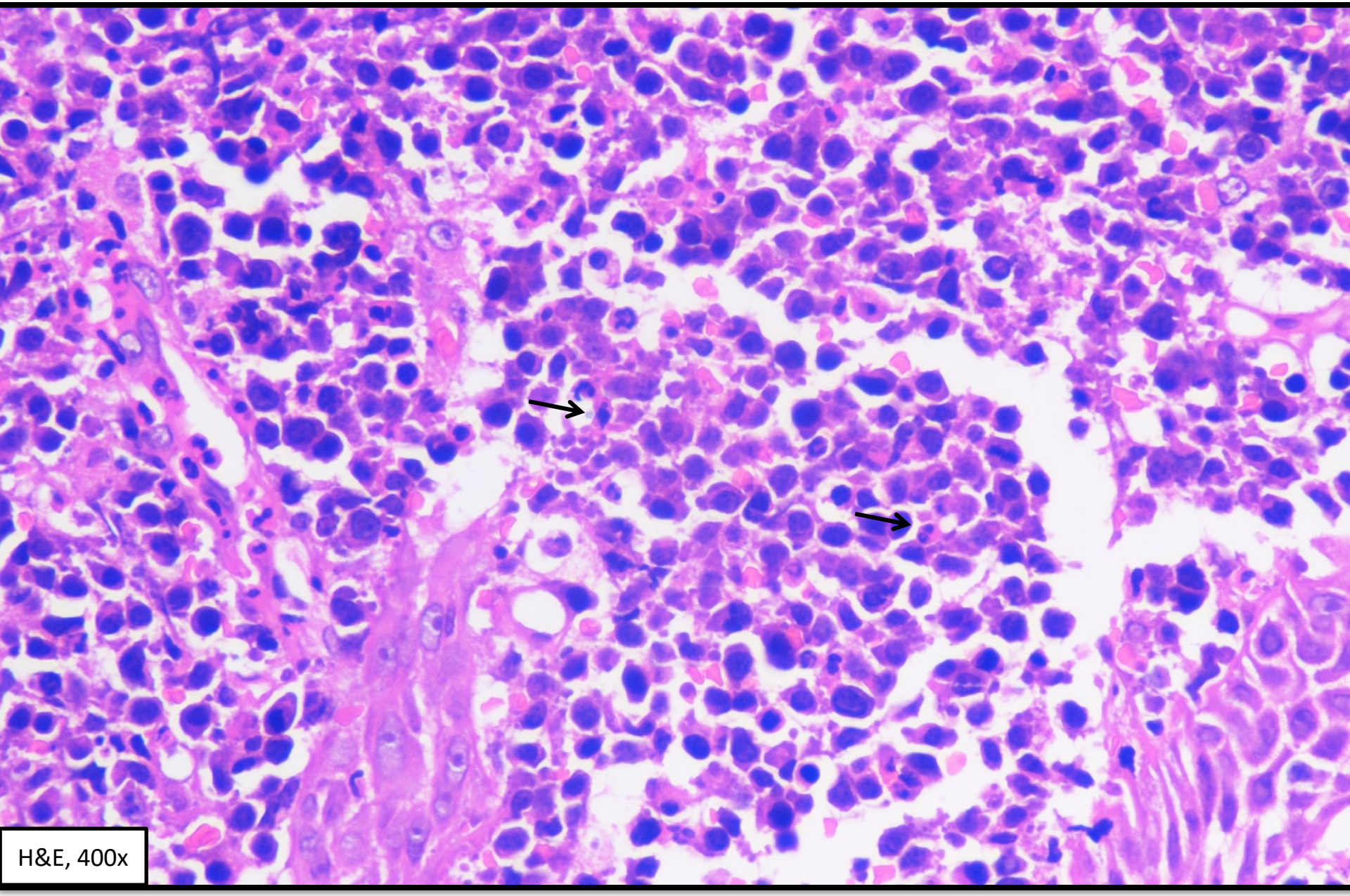
- Multiple FDG avid lytic lesion and marrow deposits were seen in sternum, few dorso-lumbar vertebrae, sacrum and bilateral iliac bones

- A working clinico-radiologial diagnosis of carcinoma of right upper alveolus was made.

- Subsequently, a biopsy from the alveolar mass was performed.

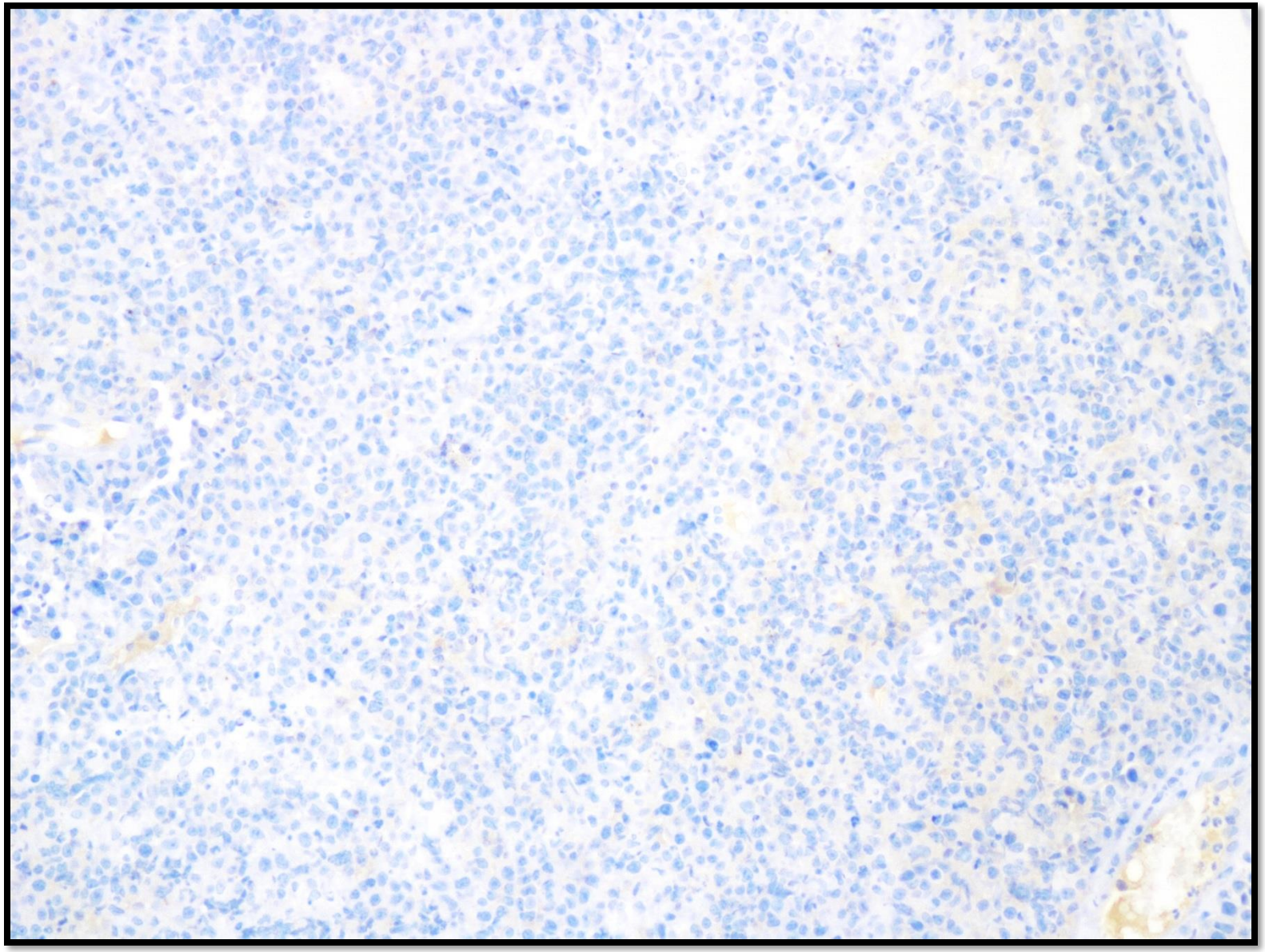


Biopsy showed partially ulcerated mucosa with diffuse infiltration by neoplastic cells

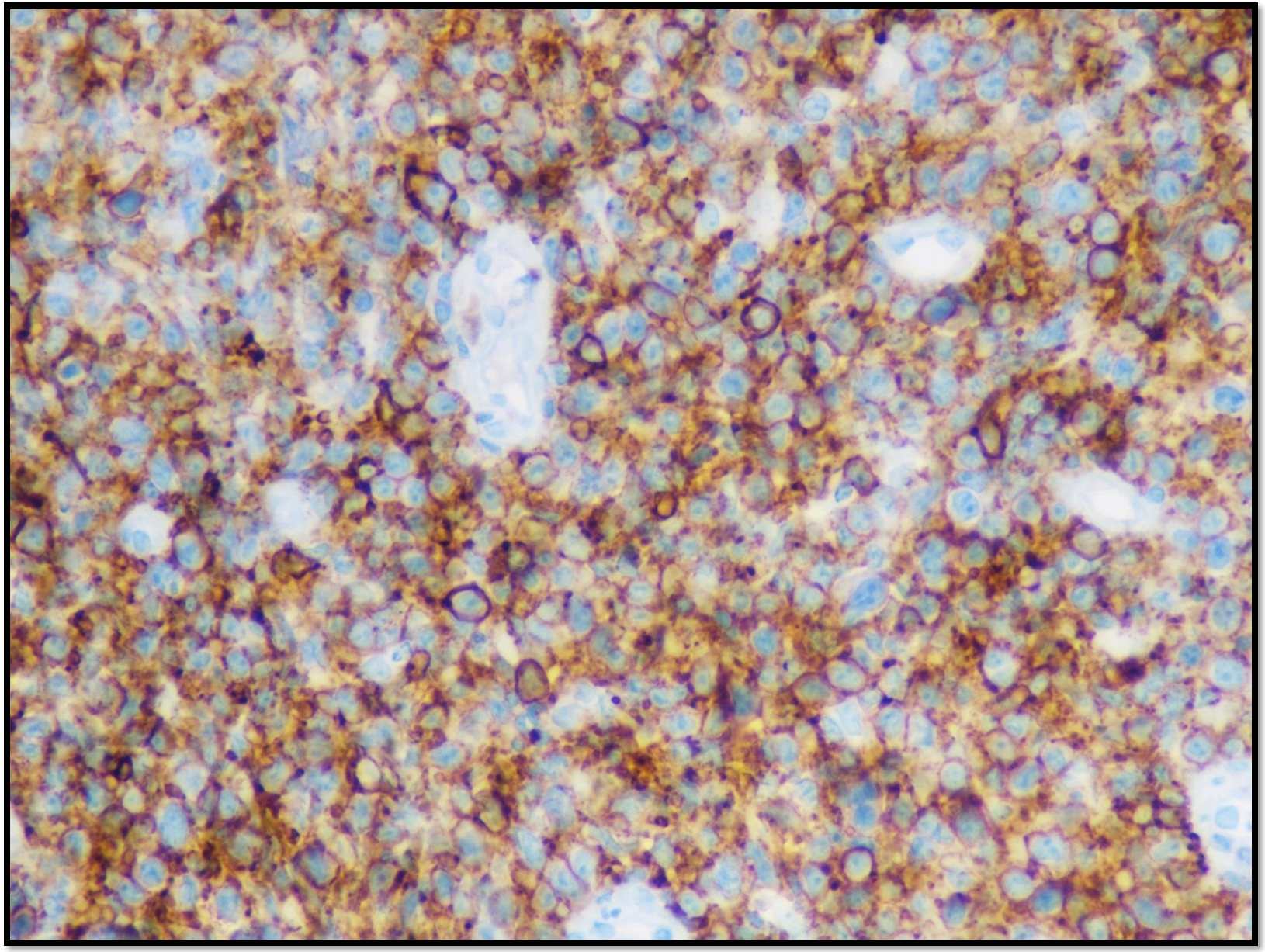


The neoplastic large round cells have plasmacytoid nucleus, prominent nucleoli and exhibit significant mitosis (Arrow)

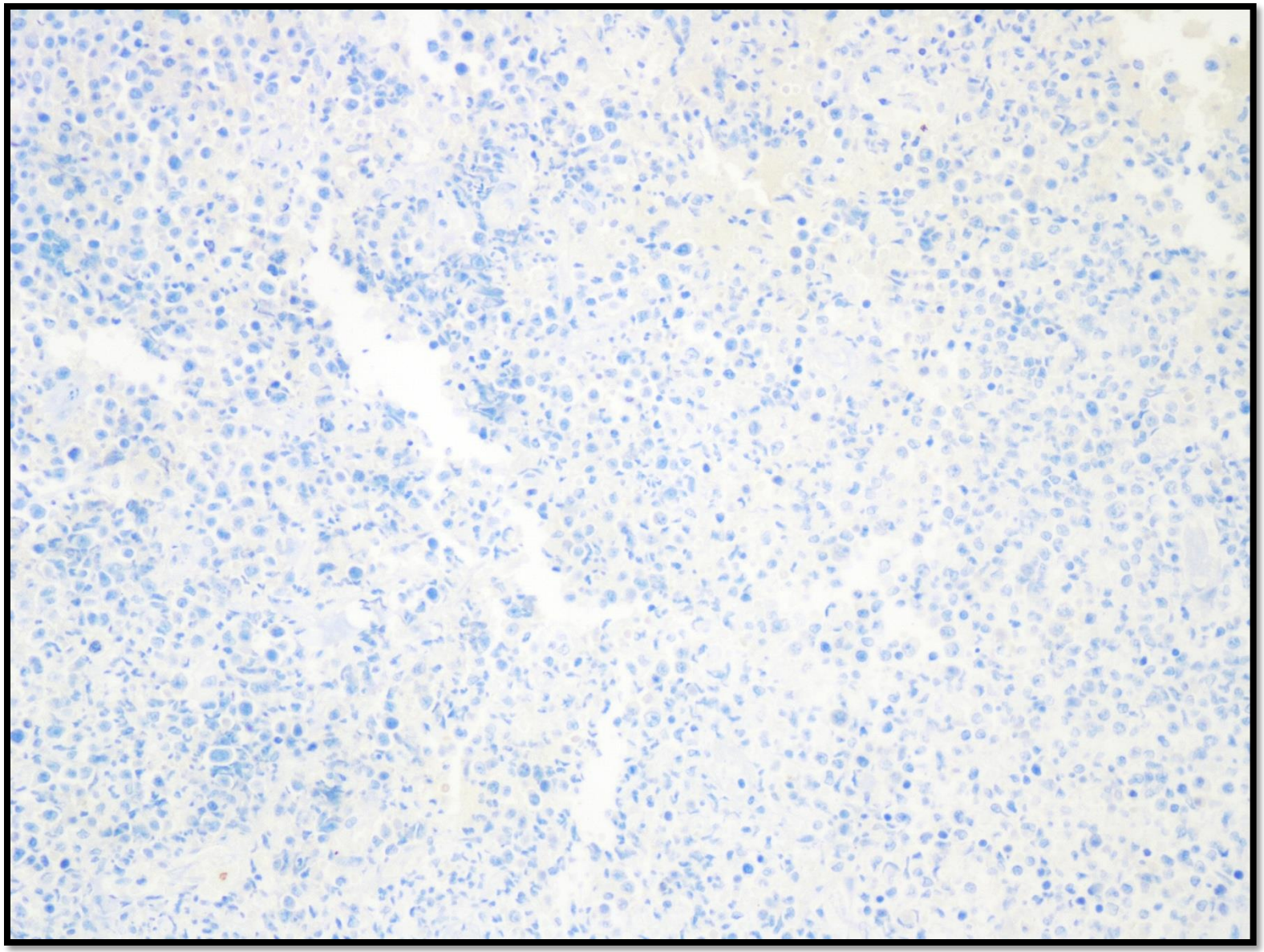
CK: Negative



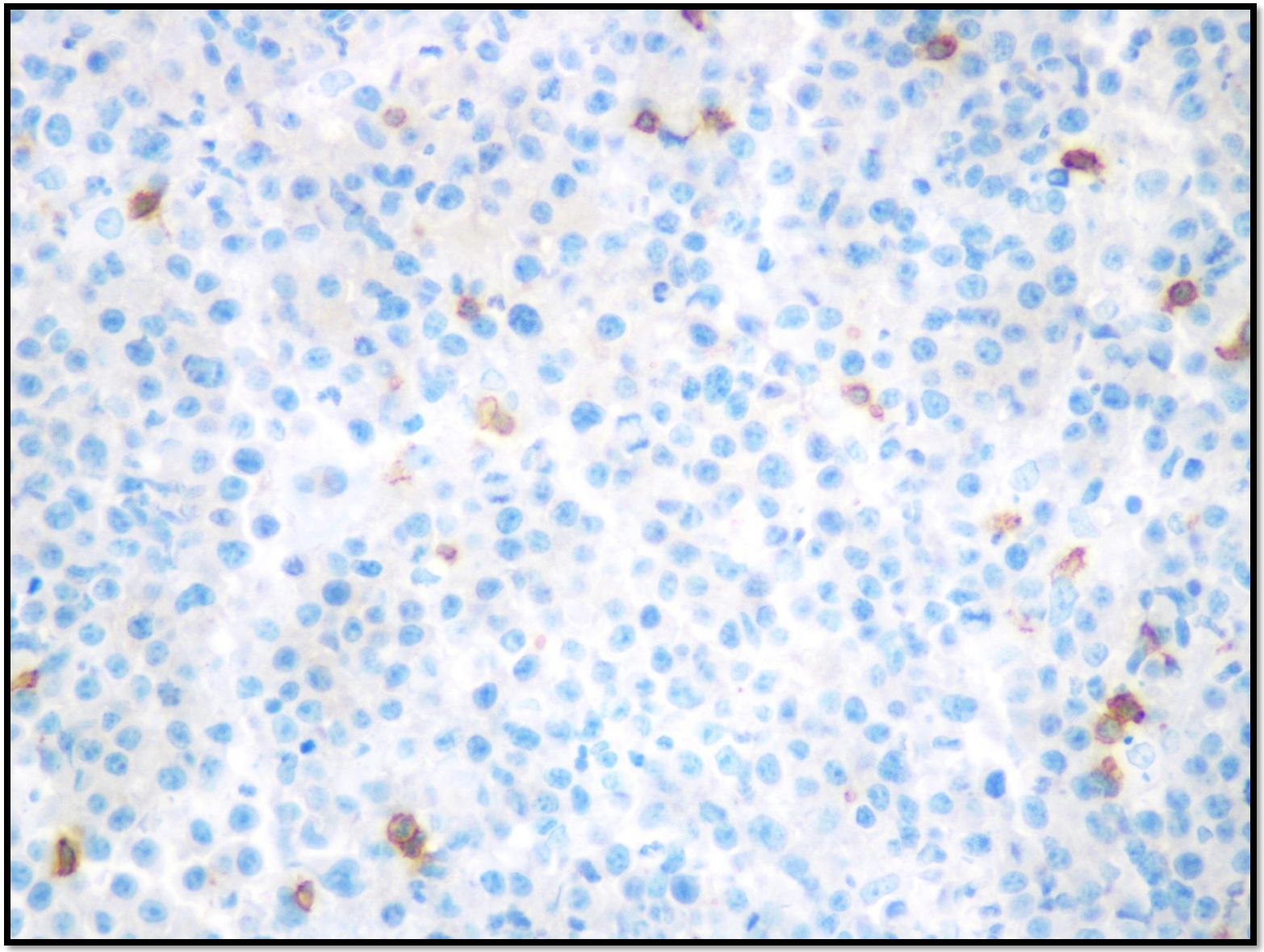
LCA: Positive



CD20: Negative



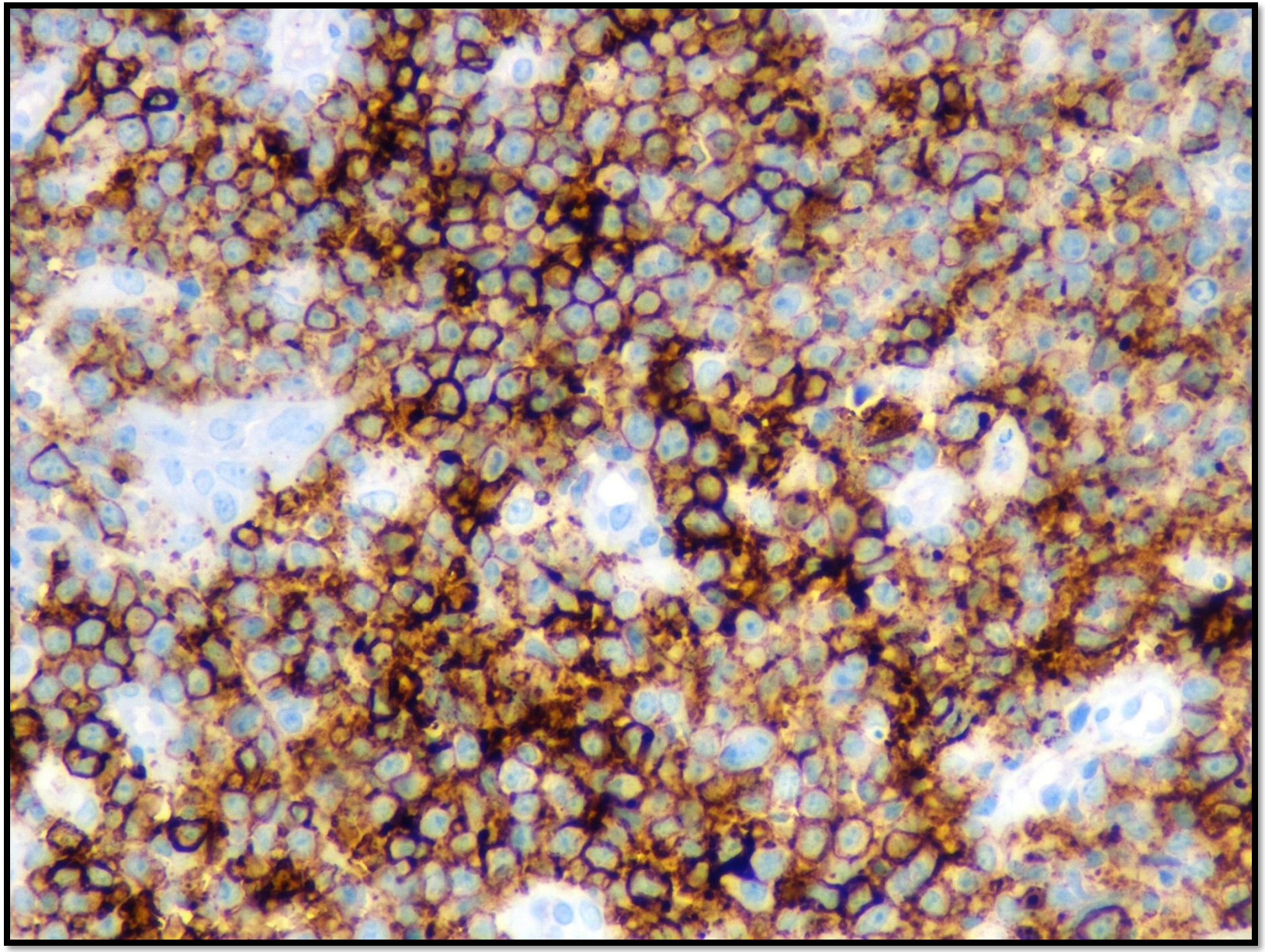
CD3: Negative



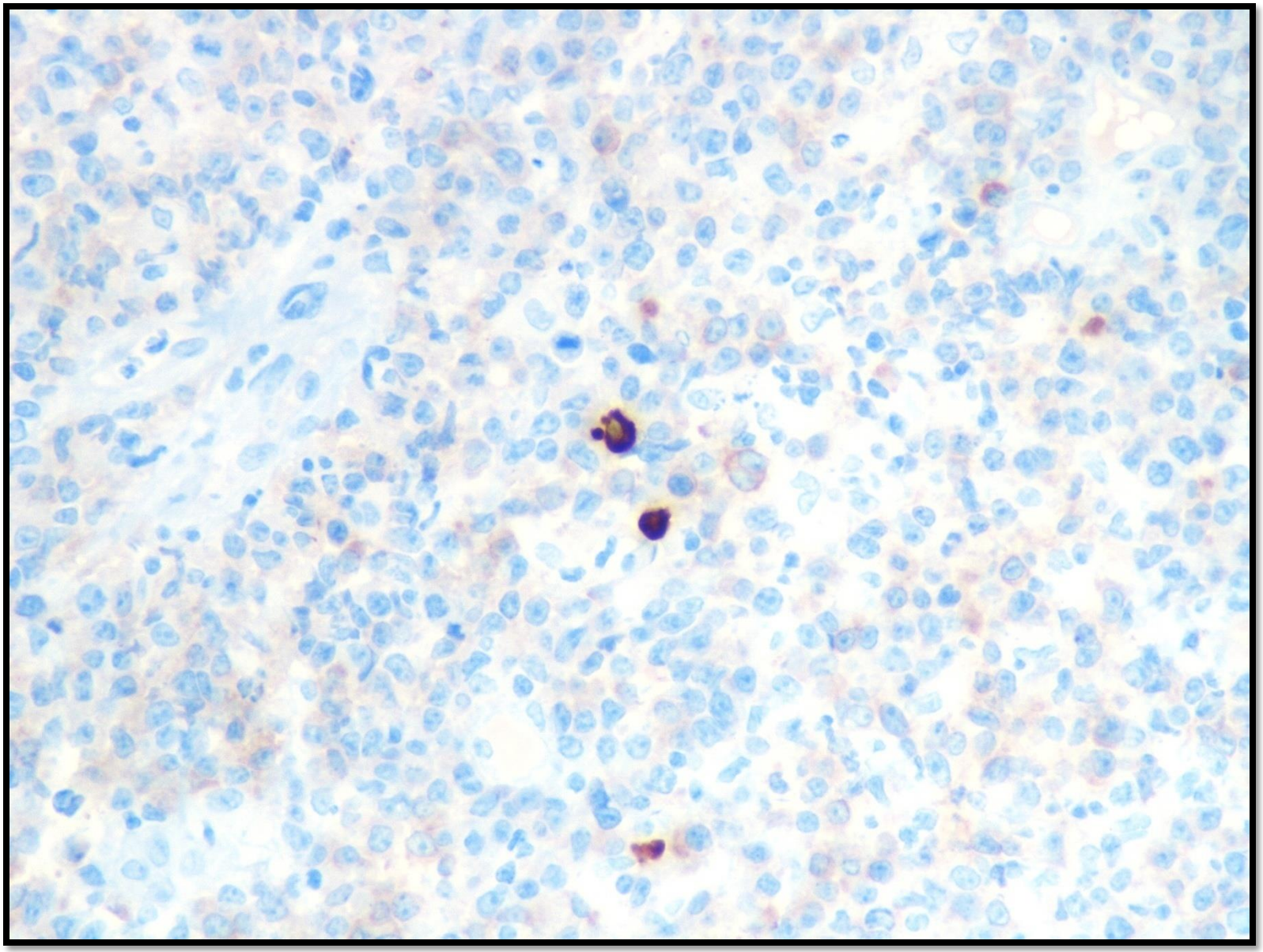
Differentials after primary IHC panel

- Myeloid sarcoma
- ALK positive large B-cell lymphoma
- B lymphoblastic lymphoma
- HHV8 positive primary effusion lymphoma
- Myeloma with plasmablastic diffrentiation
- Plasmablastic lymphoma

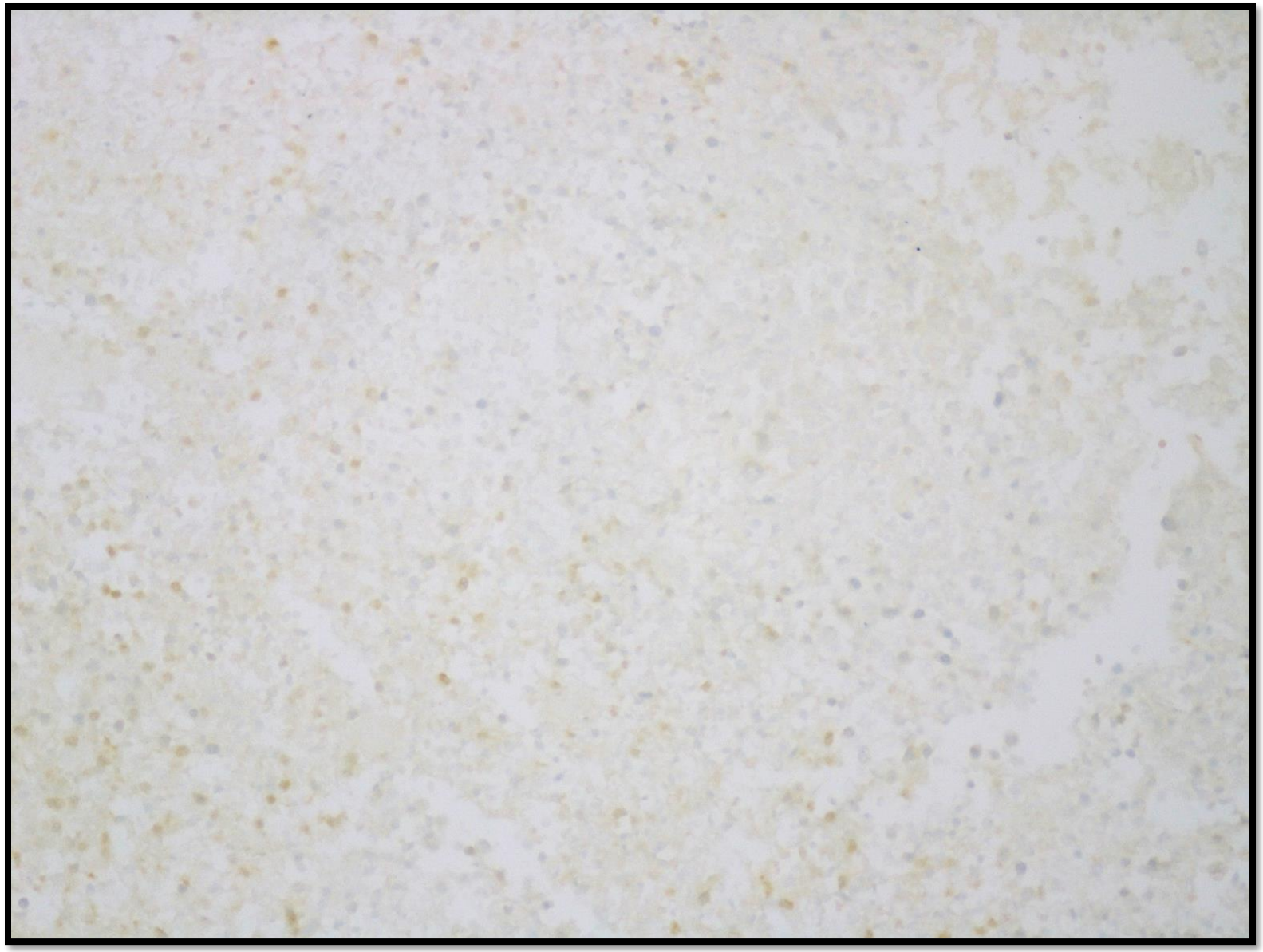
CD38: Positive



CD79a: Focal



ALK: Negative

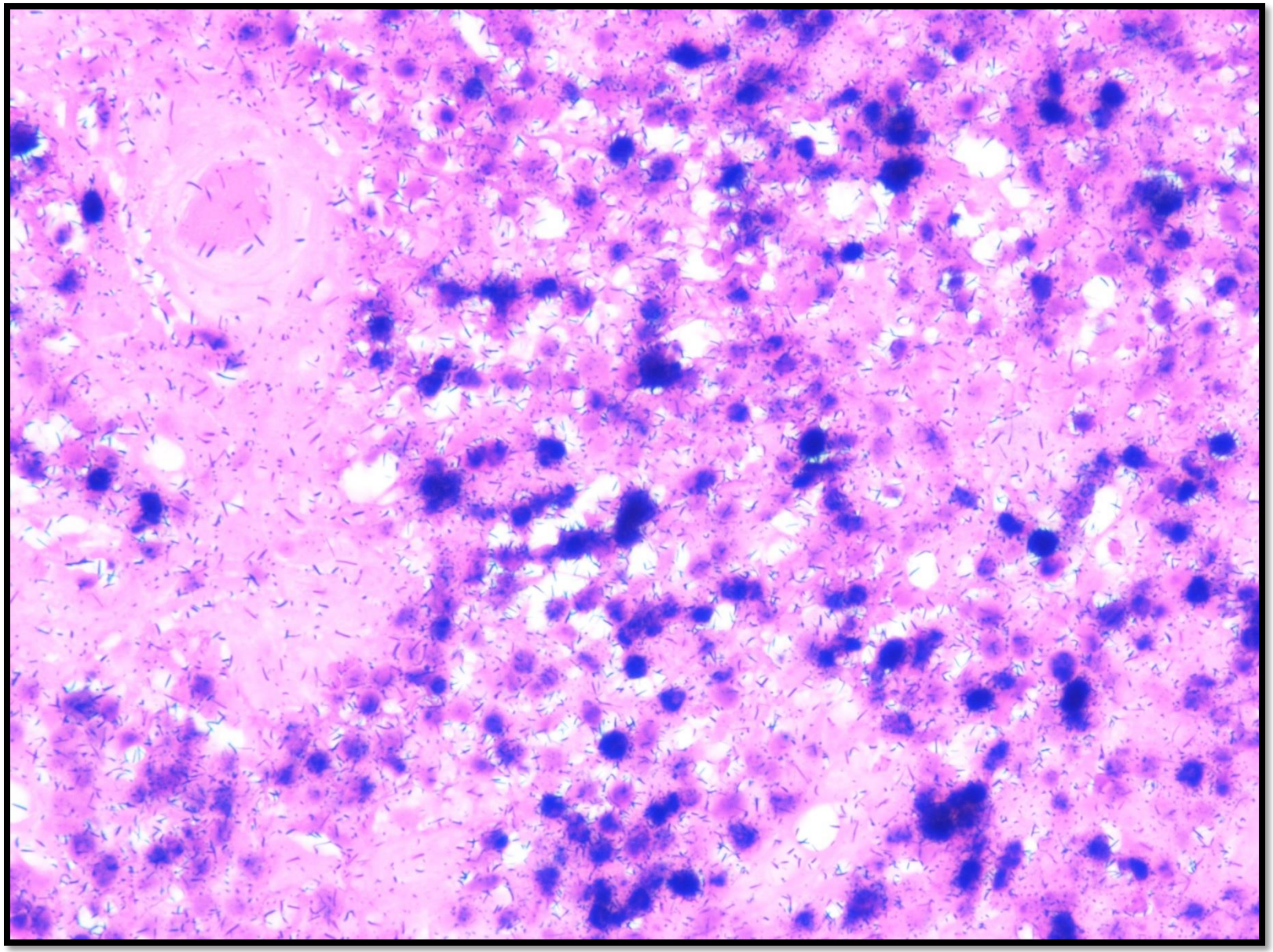


In view of LCA, CD79a and CD38 positivity

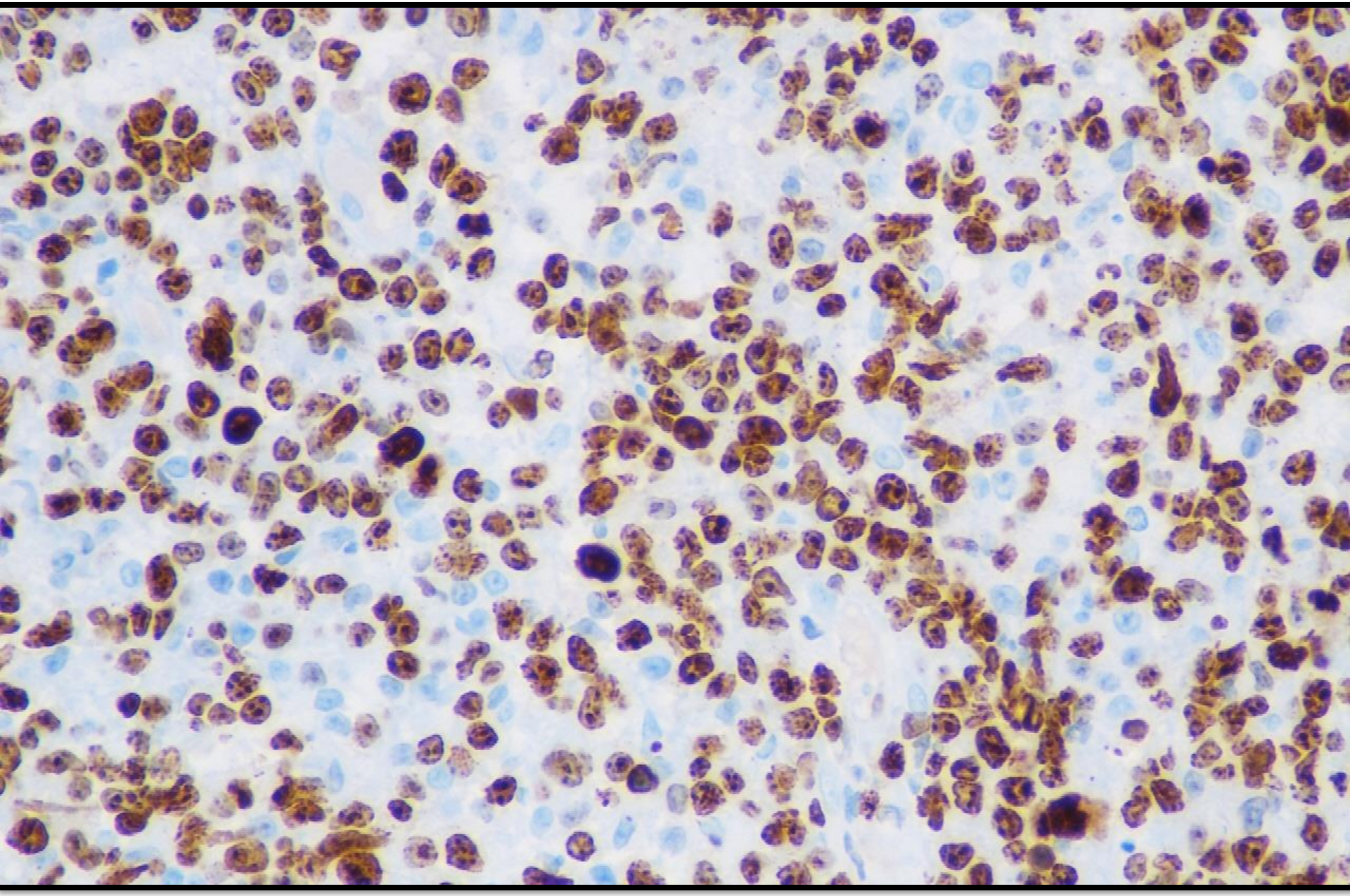
Differentials remaining in following order

- Multiple Myeloma
- Plasmablastic lymphoma

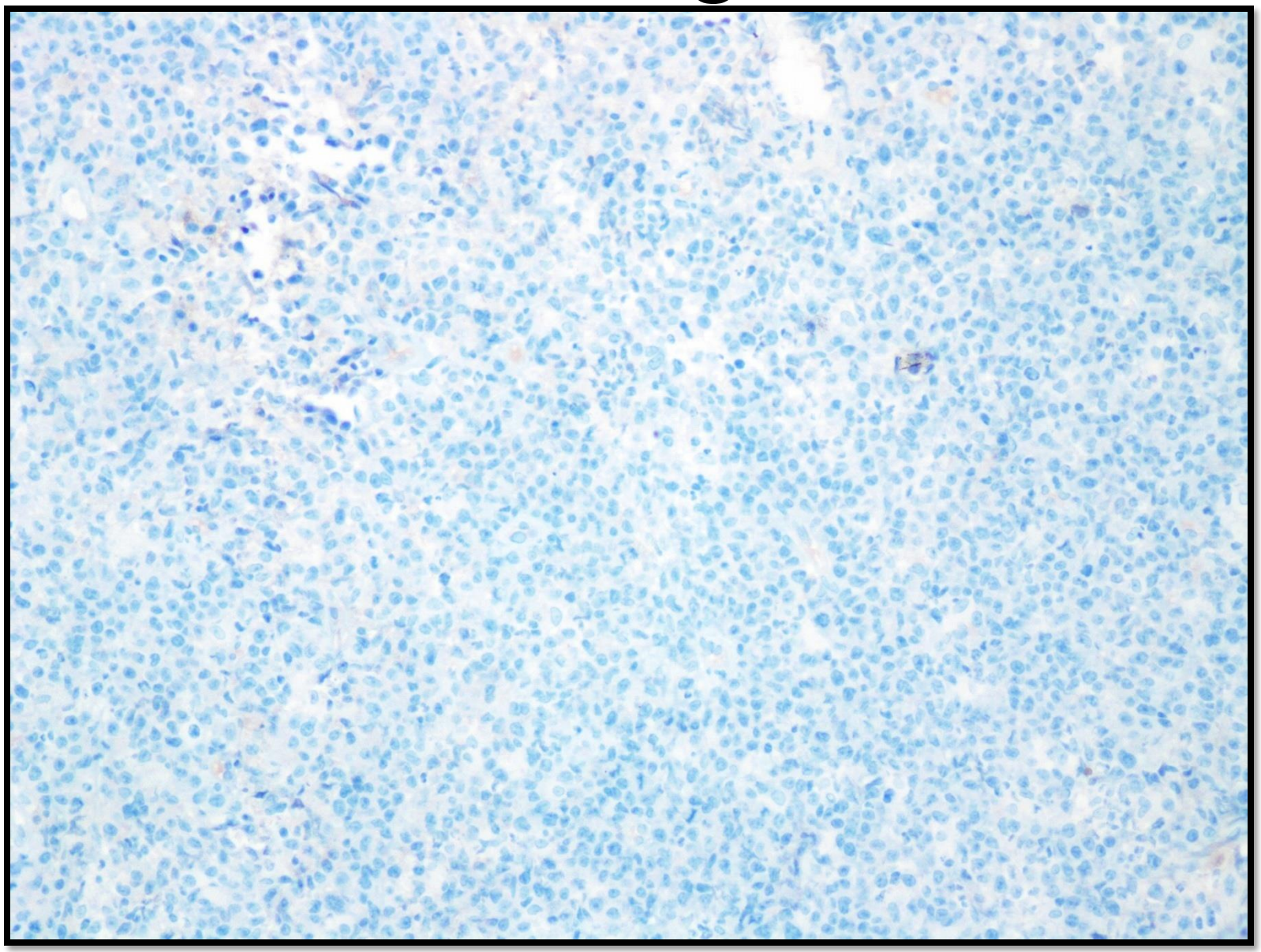
EBER-ISH: Positive



Ki-67: ≈90%



CD56: Negative



Final Diagnosis

- In view of EBER positivity, Ki-67 index of ≈90% and CD56 negativity,

- A diagnosis of Non-Hodgkin Lymphoma, B-cell type favouring **Plasmablastic lymphoma (PBL)** was rendered.

Further investigations

- Viral markers were reactive for HIV
  - Absolute CD4 count was 25/uL
  - HIV viral load was 983749 copies/mL

- Bone marrow biopsy was suggestive of marrow involvement

- CSF cytology were unremarkable

Management

- Patient was initially started on anti-retroviral therapy (ART).

- Subsequently, patient was planned and treated with CHOP based chemotherapy along with intrathecal methotrexate.

Discussion

- PBL is an aggressive variant of B-cell lymphoma commonly involving the oral cavity and mostly seen in HIV positive patients.

- In fact, World Health Organization describes PBL as an AIDS defining lesion.

- However, In India, PBL is rarely seen in HIV patients as majority of them die of immuno-compromised state due to infections.

- PBL is a NHL of B-cell immunophenotype with an unusual immunohistochemistry profile.

- It shows a negative expression for B-cell markers like CD20 and PAX5, while CD79a can be weakly expressed as evident in this case; though cancer cells express markers of plasmacytic differentiation like CD38, CD138, or MUM1/IRF4.

- Immunophenotypically, PBL is difficult to distinguish from myeloma with plasmablastic features.
  - EBER-ISH positivity and Ki67 index >90% favours the diagnosis of PBL, as in this case

- PBL in HIV seropositive patients have an aggressive course and a poor prognosis due to relapsing nature of disease and persistent immunosuppressive state of the patient .

- Antiretroviral therapy in addition to combination chemotherapy forms the basis of its treatment.

- Lymphoma specific chemotherapy regimens like cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP) or aggressive regimens like EPOCH (Etoposide, prednisolone, vincristine, cyclophosphamide and doxorubicin) are recommended.

Conclusion

- PBL is an aggressive variant of NHL which presents a diagnostic challenge due to its overlapping histological and immunophenotypic features with other lymphomas and myeloma.
- B cell markers like CD20 and PAX5 can be negative in few B cell lymphomas and may lead to their exclusion from the differential diagnosis.
- This highlights the role of CD79a, another B cell marker, which can help clinch the diagnosis when the suspicion of lymphoma is high.
- Features such as a high Ki67 index, presence of EBV infection, immunodeficient state and extra-nodal disease may point to PBL diagnosis.

THANK YOU