MAXILLARY SINUS TUMOR LIKE CONDITION; A RARE LESION, AT UNUSUAL LOCATION.

Presented by: Dr. Vinayak Sharma
JR3, Department of Pathology
Guide: Dr. Sushma Gurawale

Clinical Details

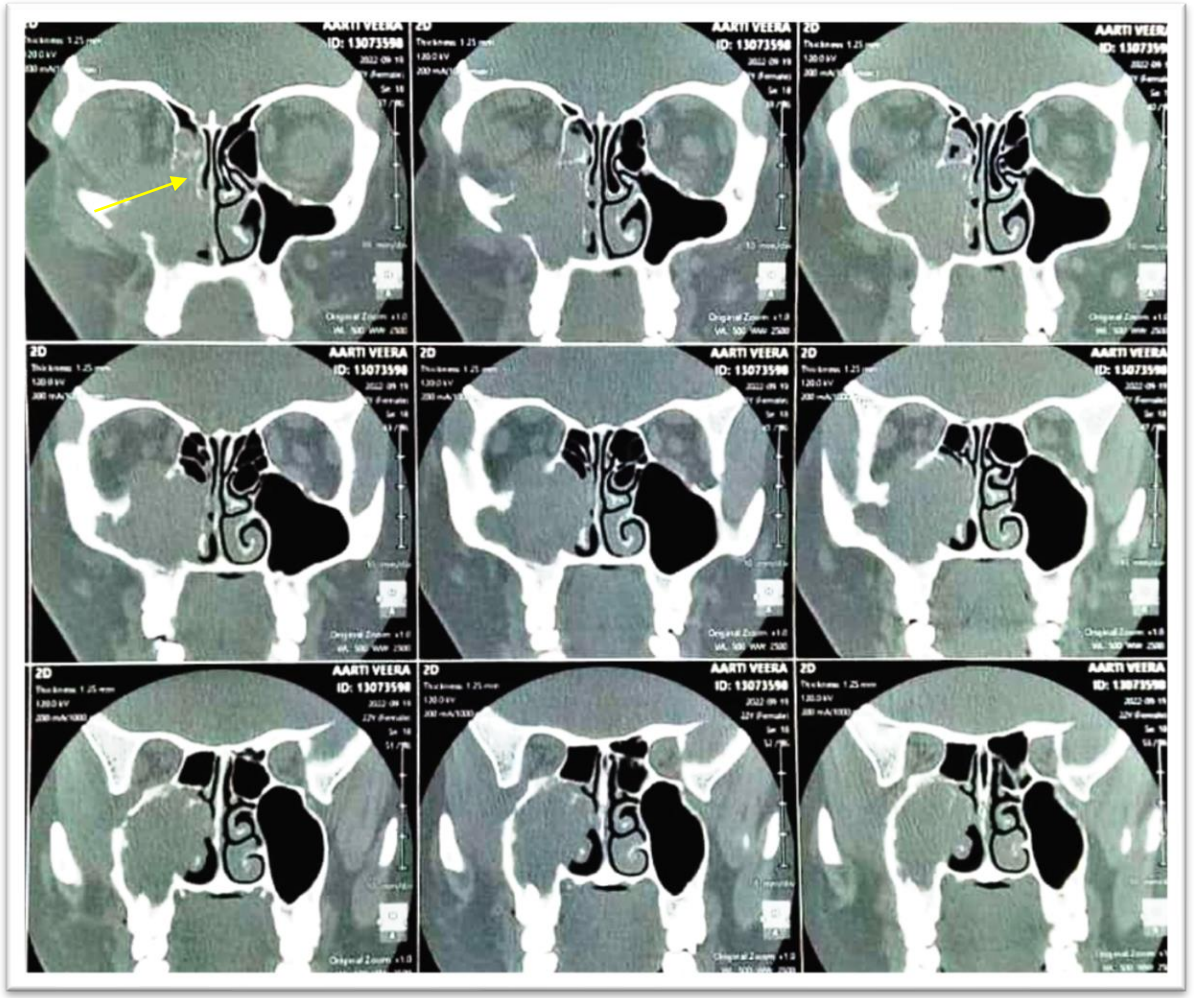
- 22 Year old female presented with;
  - Right sided nasal obstruction, swelling over right cheek, associated with epistaxis and pain since 1 month.
  - TMJ pain and dental pain was present.
  - Pain was relieved temporarily upon medication.
- Symptoms were Insidious in onset, gradually progressive in nature.
- No history of recurrent URTI, trauma, rhinorrhoea, allergic eruptions.

Examination

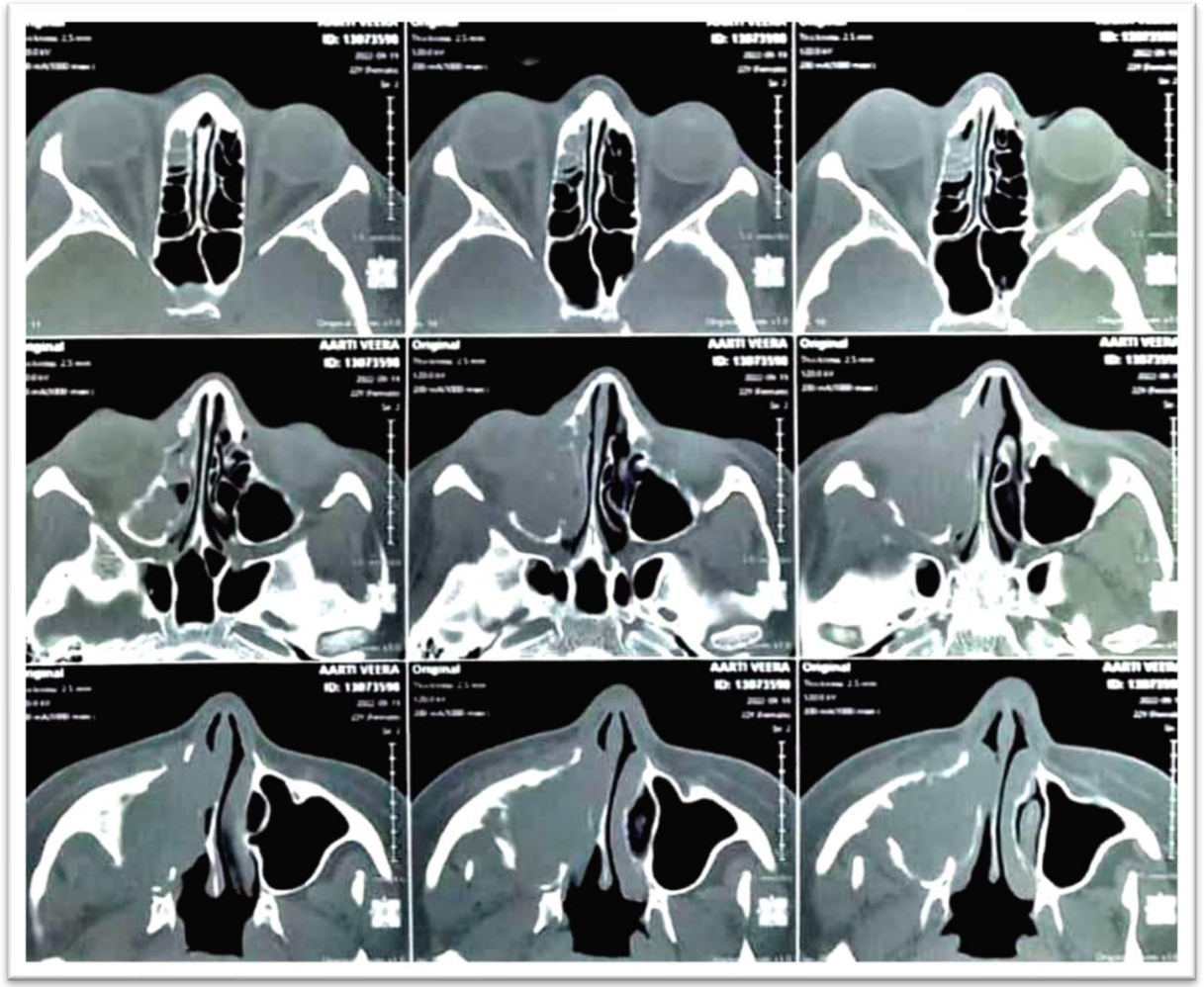
- **Nose**: Minimal swelling over right side nose and cheek.
- **Anterior rhinoscopy**: Mass seen behind the inferior turbinate protruding into right nasal cavity with gross deviation of nasal septum to right.
- Right frontal and ethmoidal sinus tenderness was present.
- **Throat examination**: Bilateral grade 2 tonsillar enlargement.

Radiology

Coronal



Axial



Large soft tissue mass of approx. size 37 X 32 mm in right maxillary antrum completely filling it and extending in nasal cavity through destruction of medial wall of sinus.

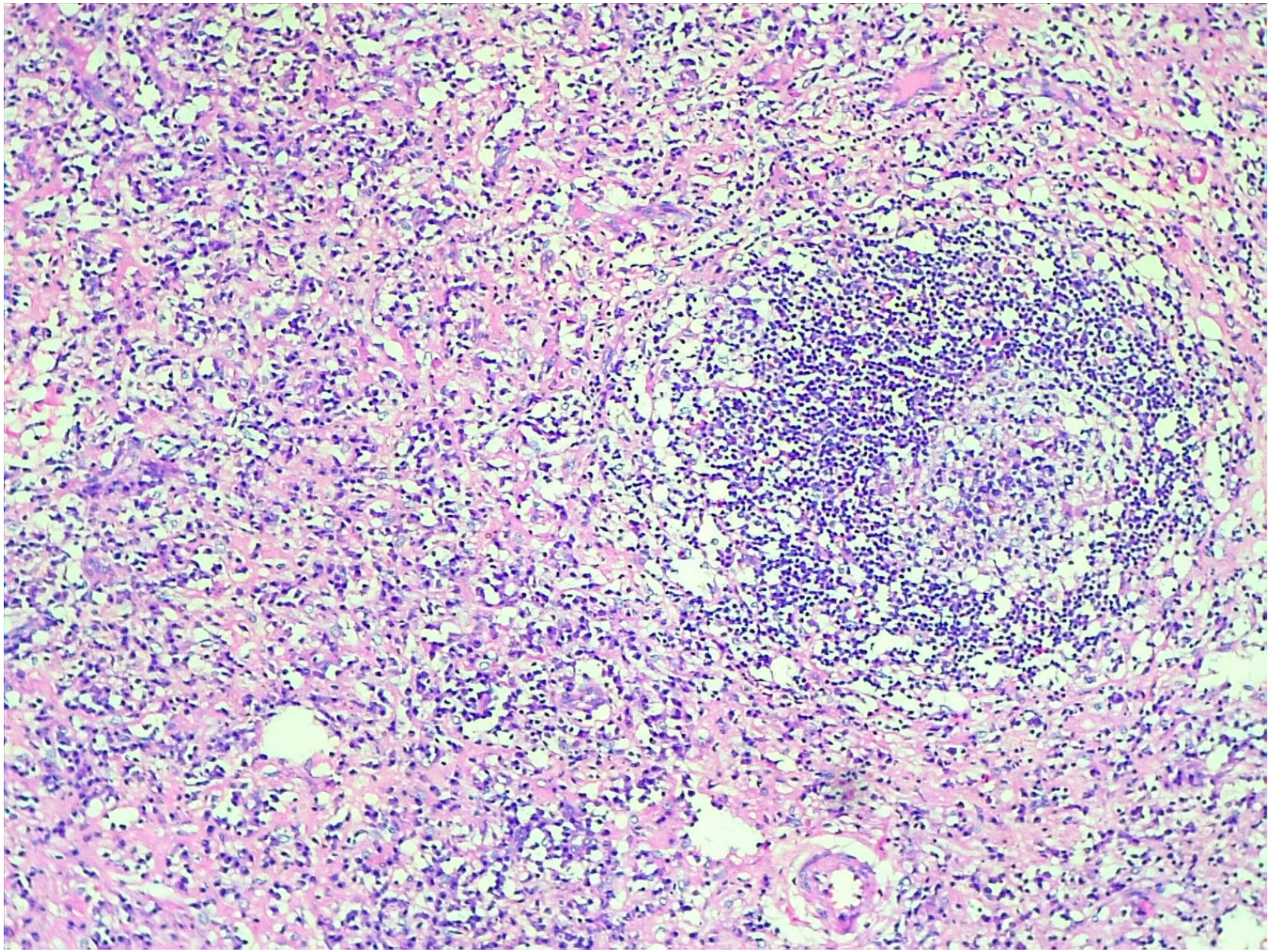
Epicentre appears at floor of sinus with lytic destruction of maxillary alveolus.

- Endoscopic right maxillary clearance and submucosal resection (SMR) was done.

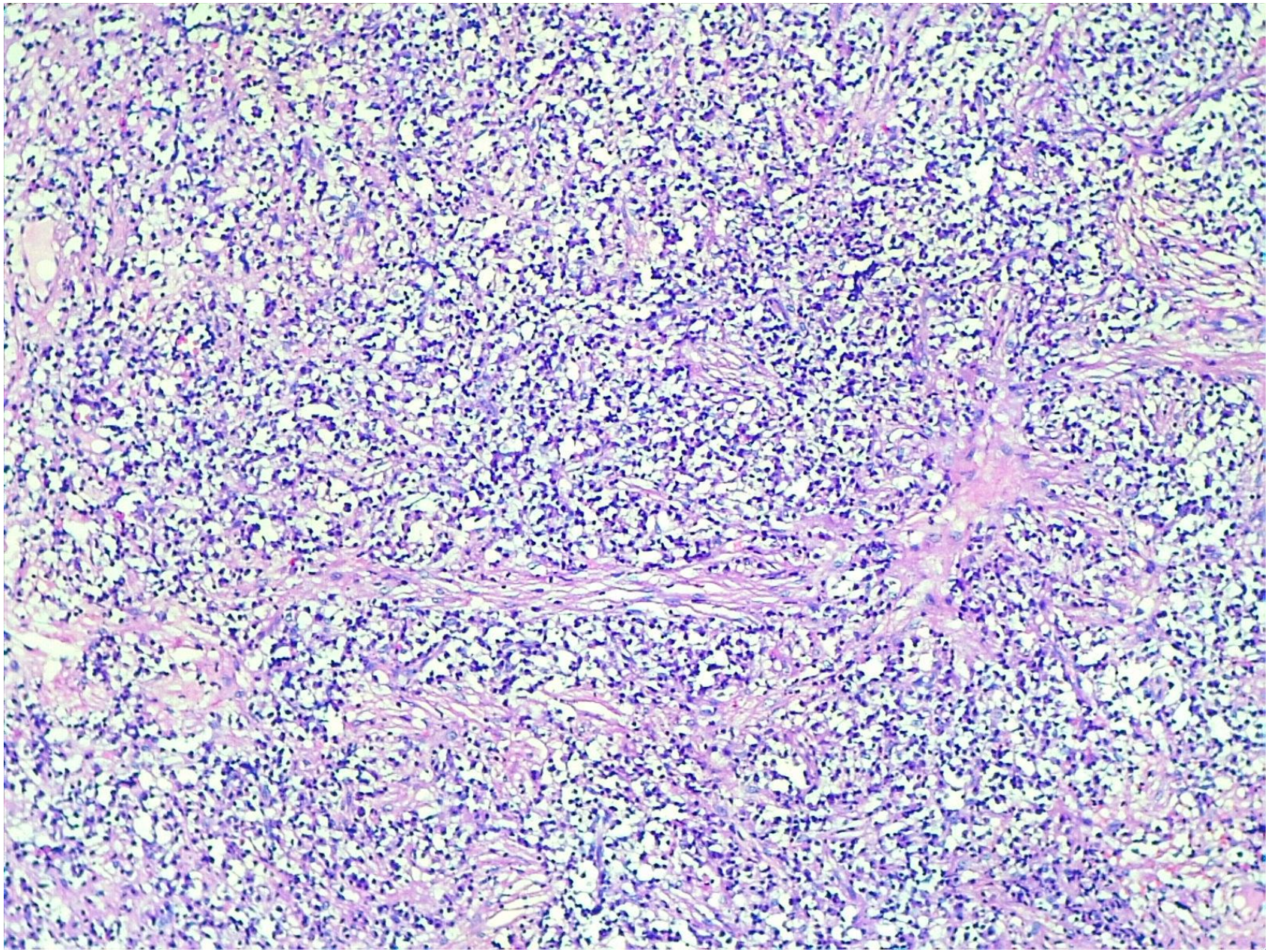
Histopathological Examination

- Gross
  - Received multiple grey brown, soft to firm tissue pieces ranging from 0.5cm to 2.5cm in greatest dimension, aggregating to 6 X 5 X 1.5 cm.

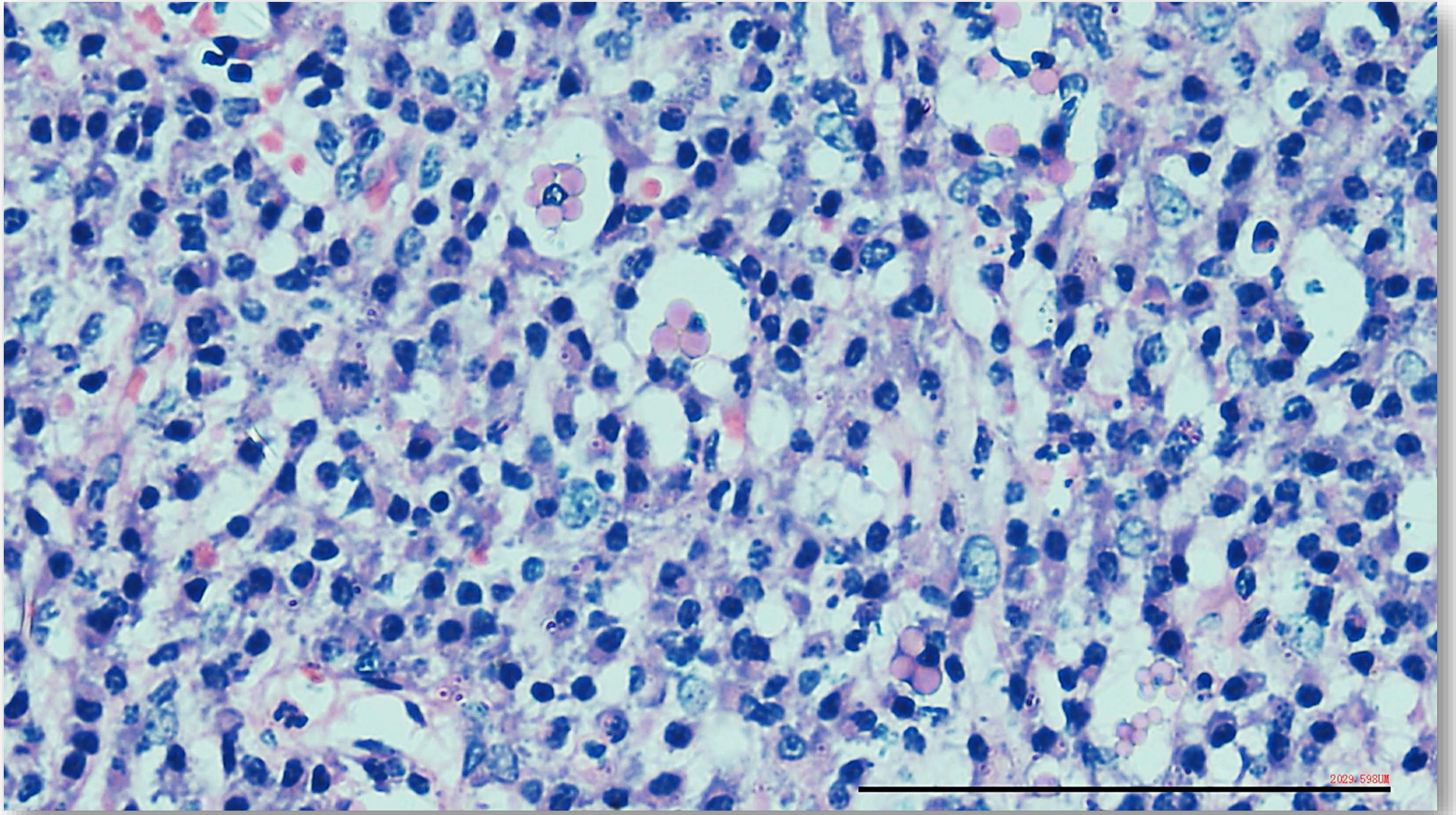
- Microscopy
  - Fragments of nasal mucosa exhibiting extensive lymphoplasmacytic inflammatory infiltrate.
  - Presence of lymphoid follicles
  - Areas of fibrosis with storiform pattern.
  - Infiltration of bony trabeculae.



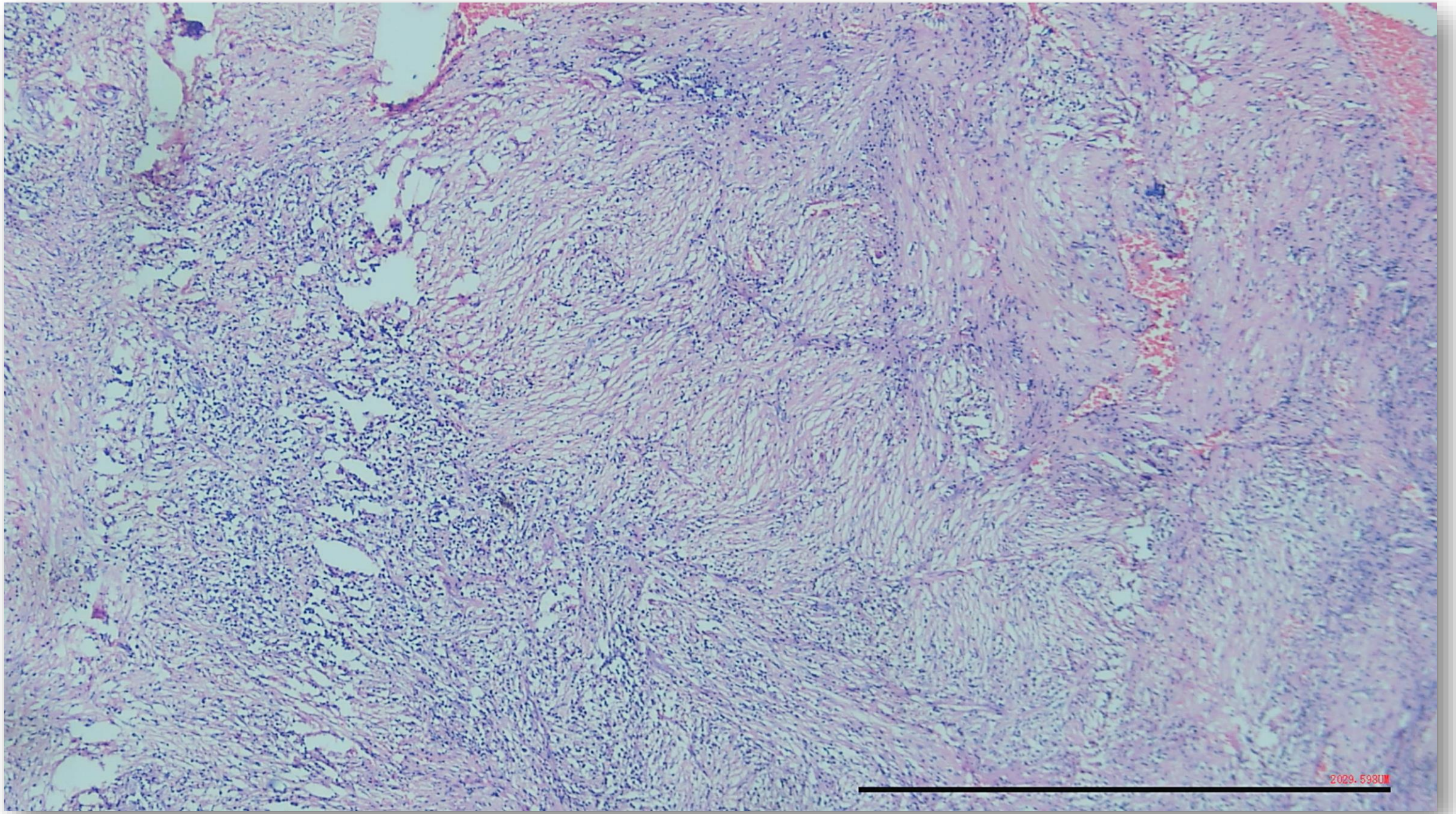
H&E 100X low power



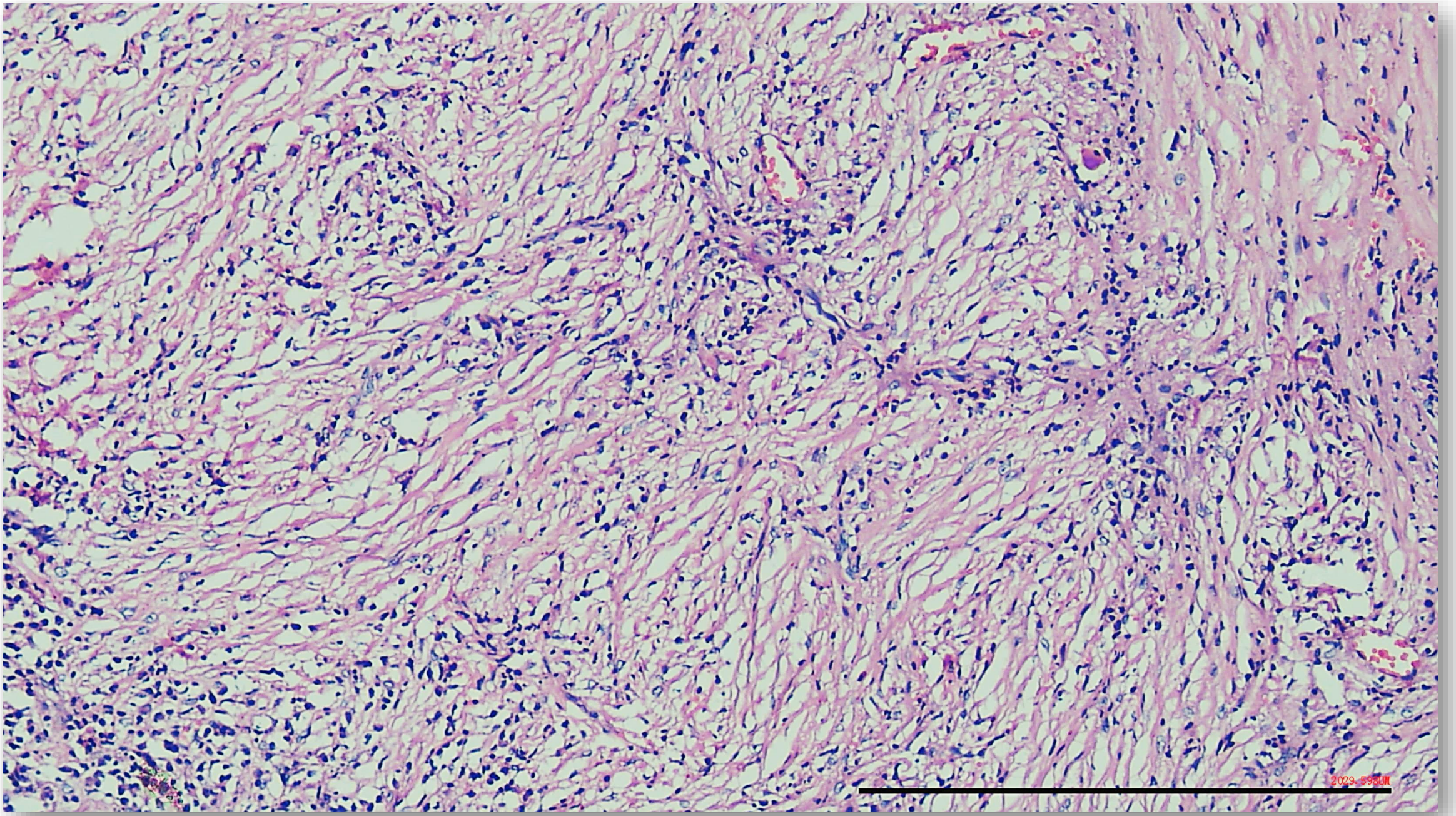
H&E: 200X



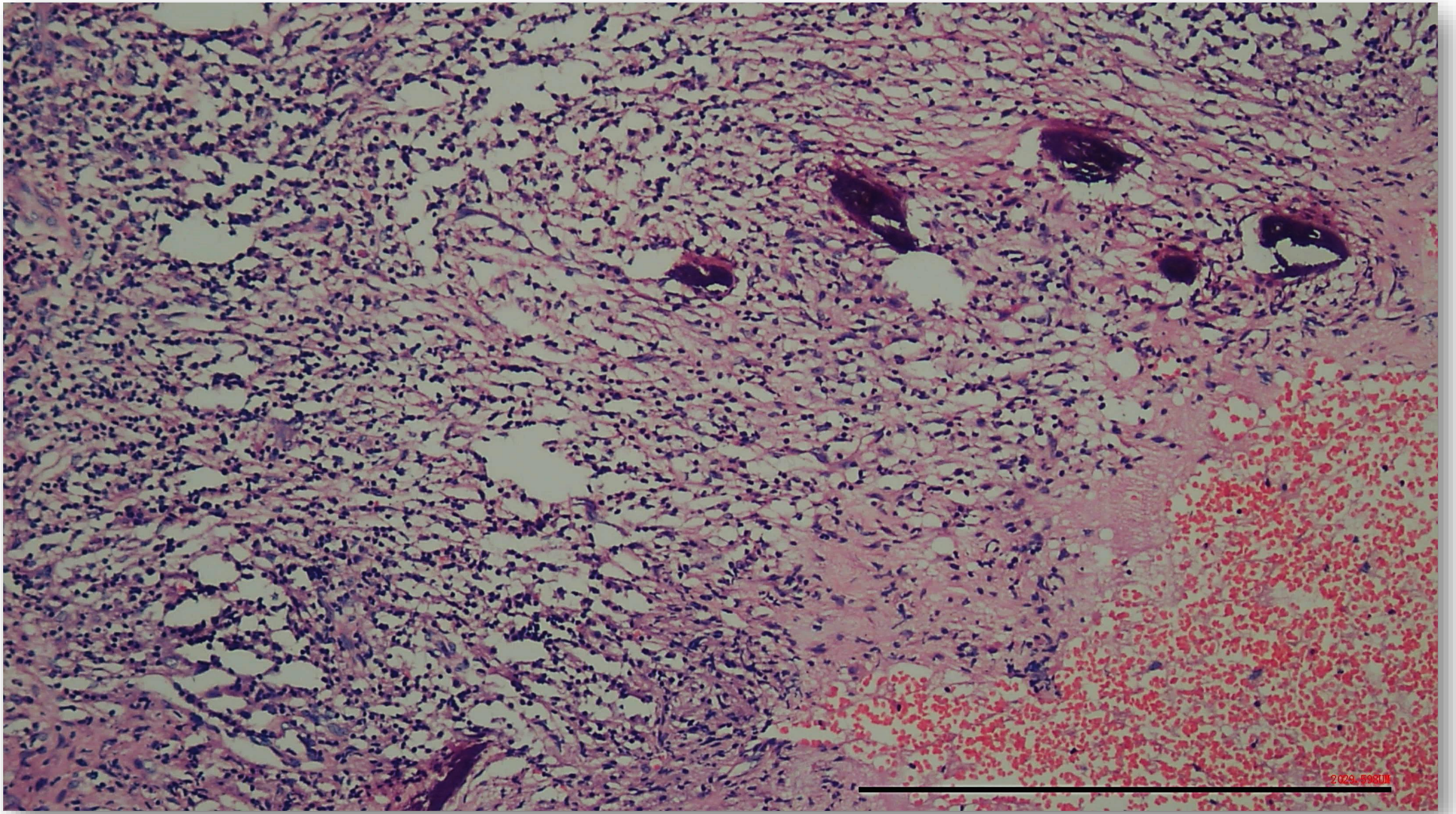
H&E 400X



H&E 100X LOW POWER



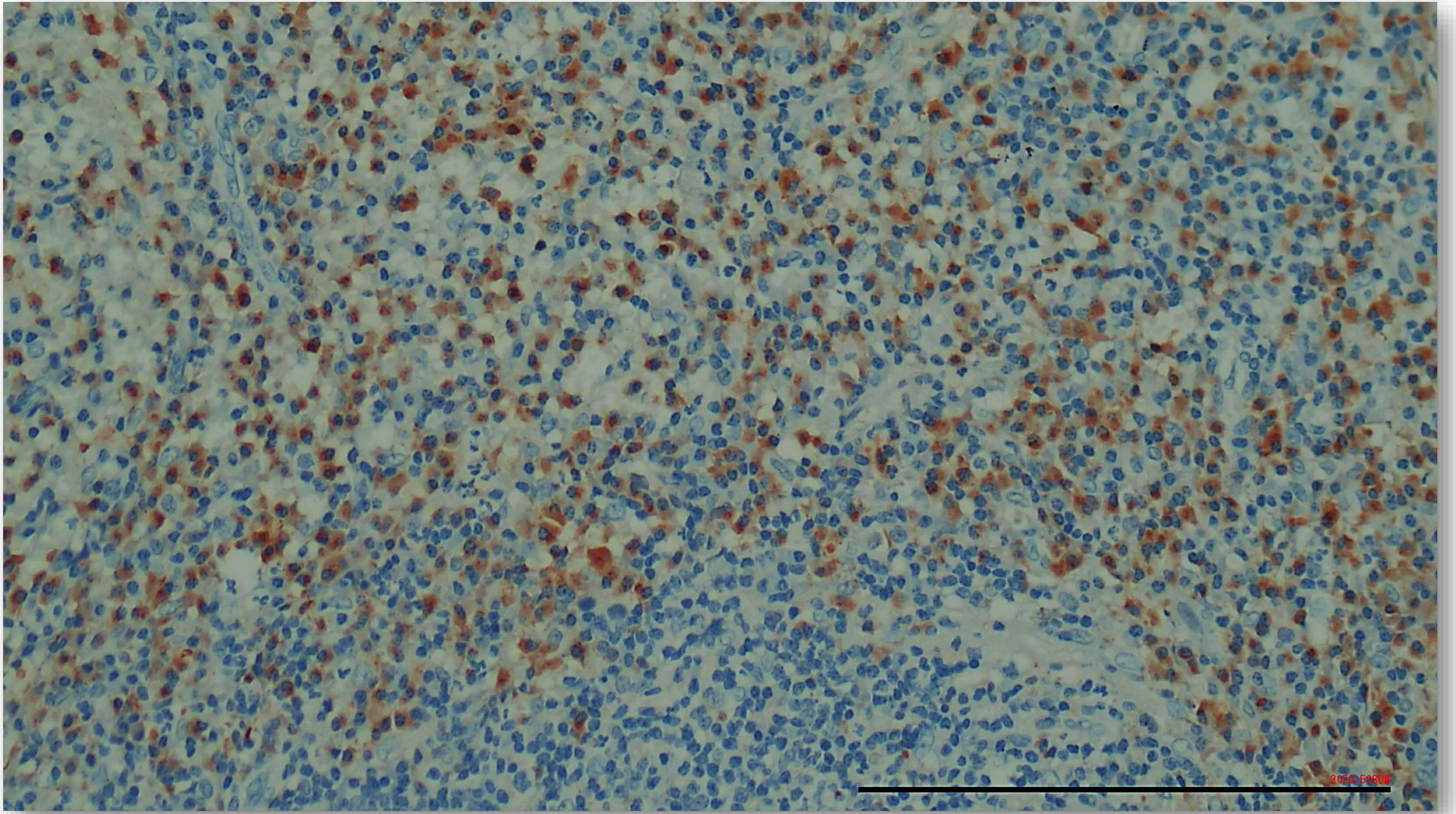
H&E 200X



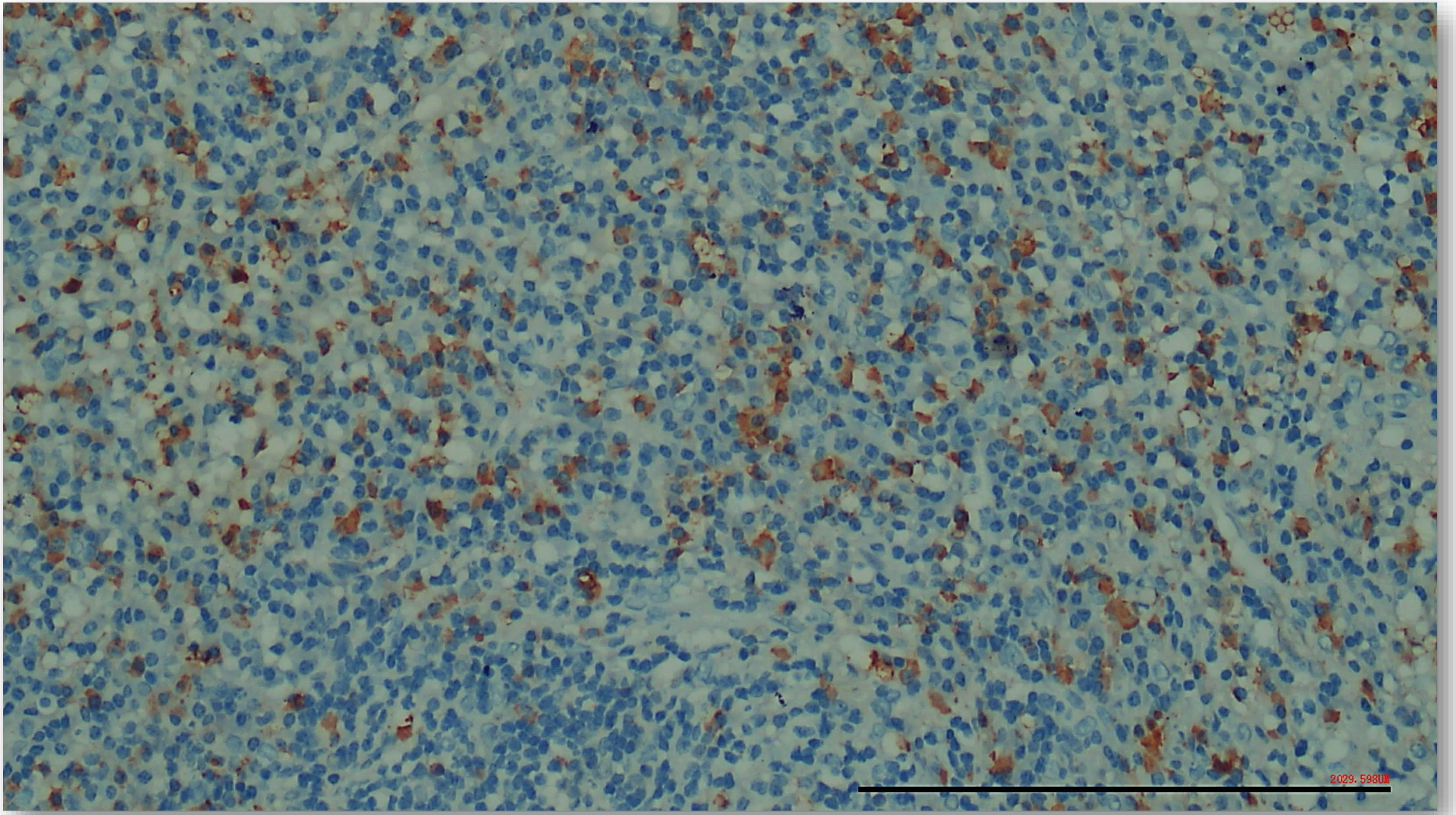
H&E 100X

- Differential diagnosis were made:
  - Plasma cell granuloma.
  - IgG4 Disease.
  - Plasmacytoma.

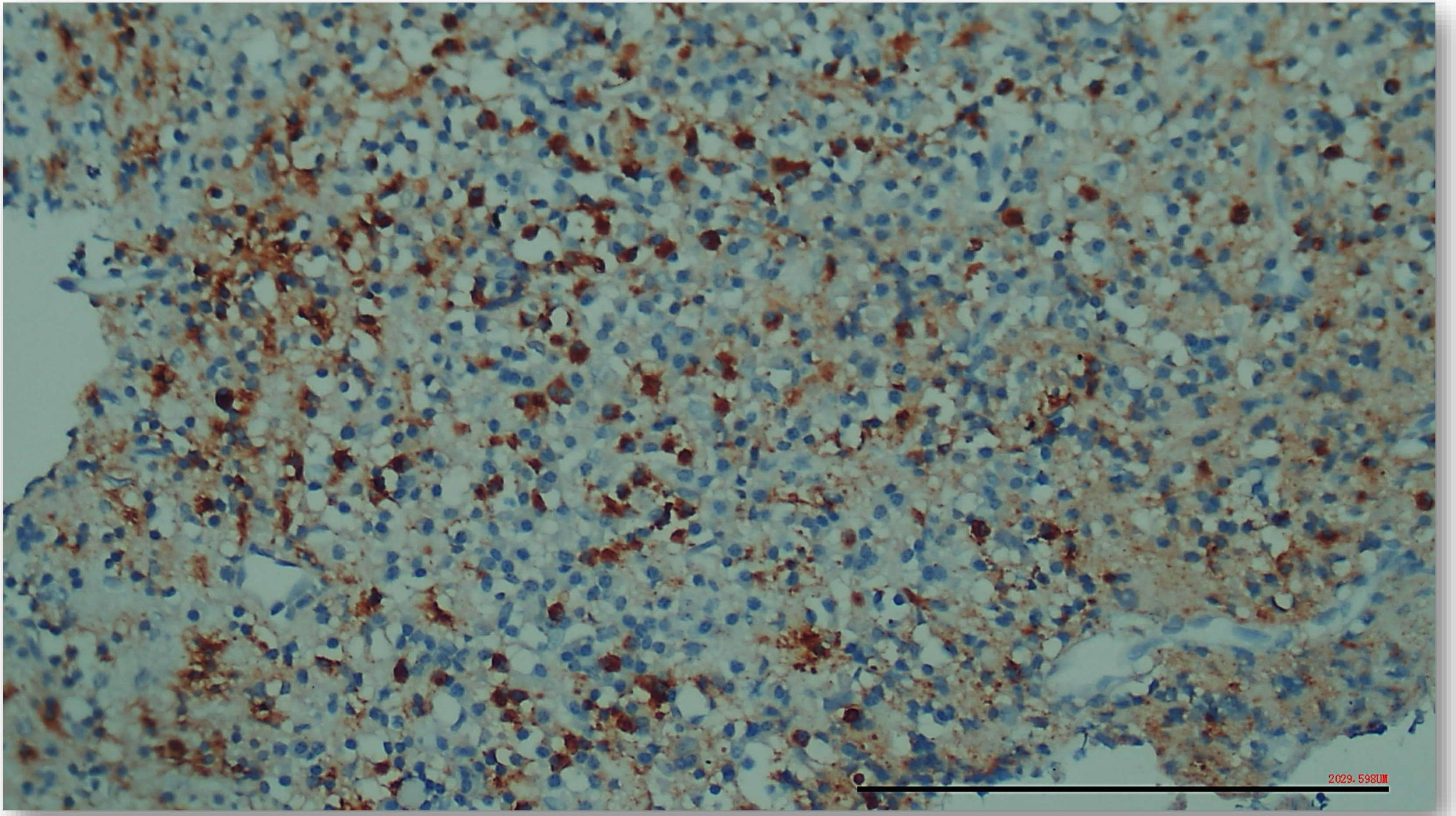
- IHC was used for the diagnosis:
  - Kappa
  - Lambda
  - IGG4



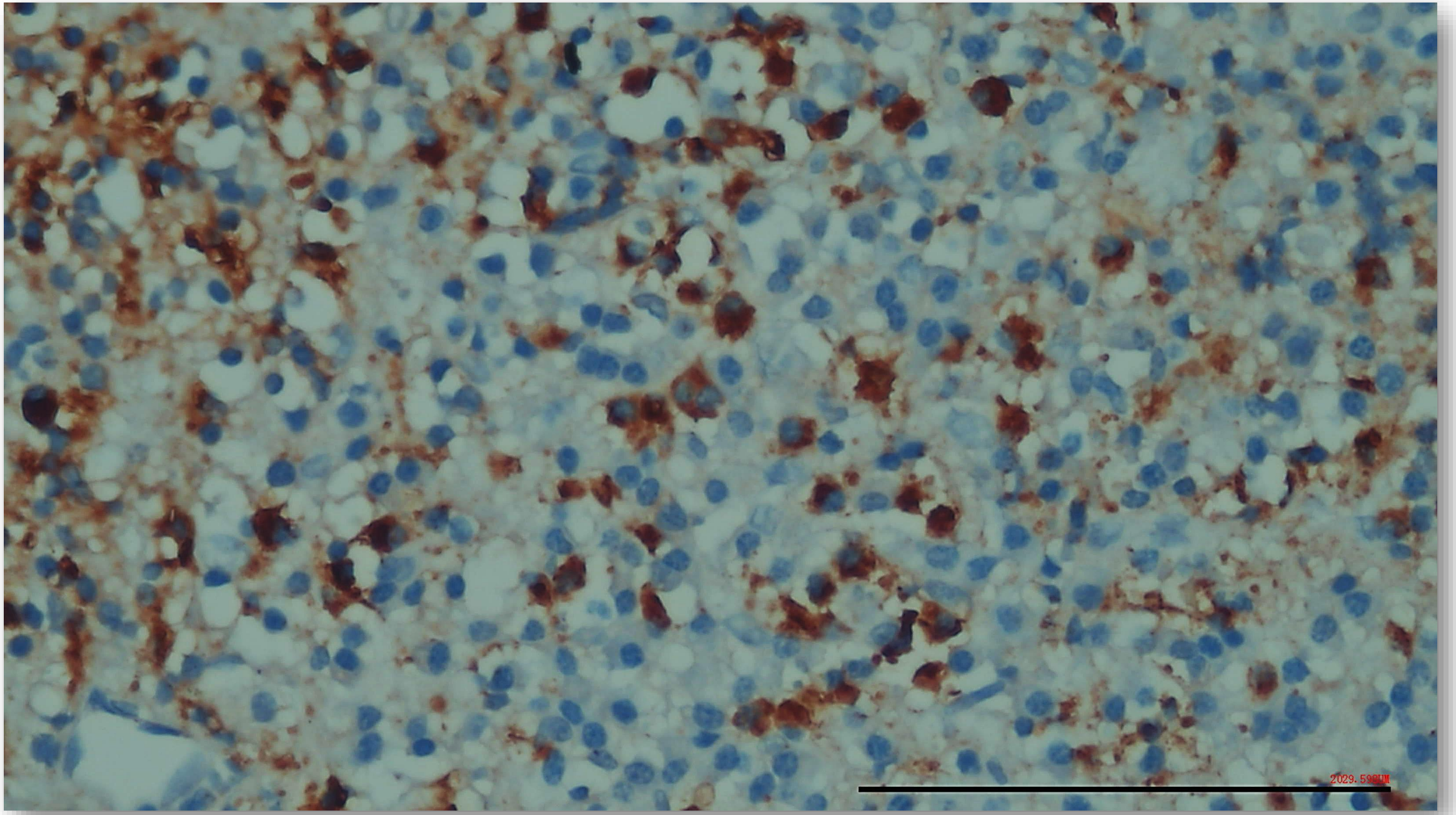
Kappa IHC: 200X



LAMBDA IHC: 200X



IgG4 IHC: 100X



IGG4 IHC: 400X

Final Diagnosis

- Based upon presence of
  - Dense lymphoplasmacytic infiltrate
  - Fibrosis, usually storiform in character
- IgG4 IHC positivity; in >100 cells/hpf

- Polyclonal nature of plasma cells ruled out plasmacytoma and IgG4 positivity ruled out plasma cell granuloma.

Final diagnosis of IgG4 Disease was made.

IgG4 Disease; Discussion

- A component of a systemic immune mediated fibroinflammatory condition.
- Affects almost any organ, including the pancreas, bile duct, lacrimal glands, salivary glands, central nervous system, thyroid, lungs, liver, gastrointestinal tract, kidney, prostate, retroperitoneum, arteries, lymph nodes, skin, breast.
- Also has been called; IgG4 related systemic disease, IgG4 syndrome, IgG4 associated disease, IgG4 related sclerosing disease, IgG4 related systemic sclerosing disease, IgG4 related autoimmune disease.
- Average age 65 years. (range 20 - 80 years)
- Male predominance. (3:1 - 7:1)

The Criteria

- Histologically highly suggestive of IgG4 related disease, requires at least 2 of 3 histologic features:
  - Dense lymphoplasmacytic infiltrate
  - Fibrosis, usually storiform in character
  - Obliterative phlebitis

  And

  - > 40% IgG4/IgG plasma cell ratio
  - Elevated total serum IgG (Normal <200mg/dL) or IgG4 levels (Normal 0.052-1.250 g/L)

  And/Or

  - IgG4+ plasma cells/high power field
    - > 30 in surgical specimen, or
    - > 10 in biopsy specimen

Various cutoffs have been assigned as follows

Site	IgG4 Positive plasma cells
Meninges	**>10**
Lacrimal gland	>100
Salivary gland	>100
Lymph node	>100
Lung (surgical specimen)	>50
Lung (biopsy)	>20
Pleura	>50
Pancreas (surgical specimen)	>50
Pancreas (biopsy)	>10
Bile duct (surgical specimen)	>50
Bile duct (biopsy)	>10
Liver (surgical specimen)	>50
Liver (biopsy)	>10
Kidney (surgical specimen)	>30
Kidney (biopsy)	>10
Aorta	>50
Retroperitoneum	>30
Skin	>200

Treatment Protocol

- Surgical resection of the affected tissue and treatment with systemic glucocorticoids, and immunosuppressive agents remains the mainstay.

Take home message

- IgG4-related disease is a recently recognized multi-organ system condition with pathological features that are largely consistent across a wide range of organ systems.
- Although the precise role of IgG4 in this disease is unknown, its presence in tissue in association with plasma cells provides a robust biomarker for diagnosis when interpreted in the proper histopathological and clinical contexts.
- The diagnosis of IgG4-related disease requires collaboration between the pathologist and the treating physician.

References

- Umehara, Hisanori et al. “Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011.” Modern rheumatology vol. 22,1 (2012): 21-30. doi:10.1007/s10165-011-0571-z

- Deshpande, V., Zen, Y., Chan, J. et al. Consensus statement on the pathology of IgG4-related disease. Mod Pathol 25, 1181–1192 (2012). https://doi.org/10.1038/modpathol.2012.720

- Peng F, Zuckerman JE. IgG4 related disease. PathologyOutlines.com website. https://www.pathologyoutlines.com/topic/kidneyigg4related.html. Accessed November 1st, 2022.

Thank you!