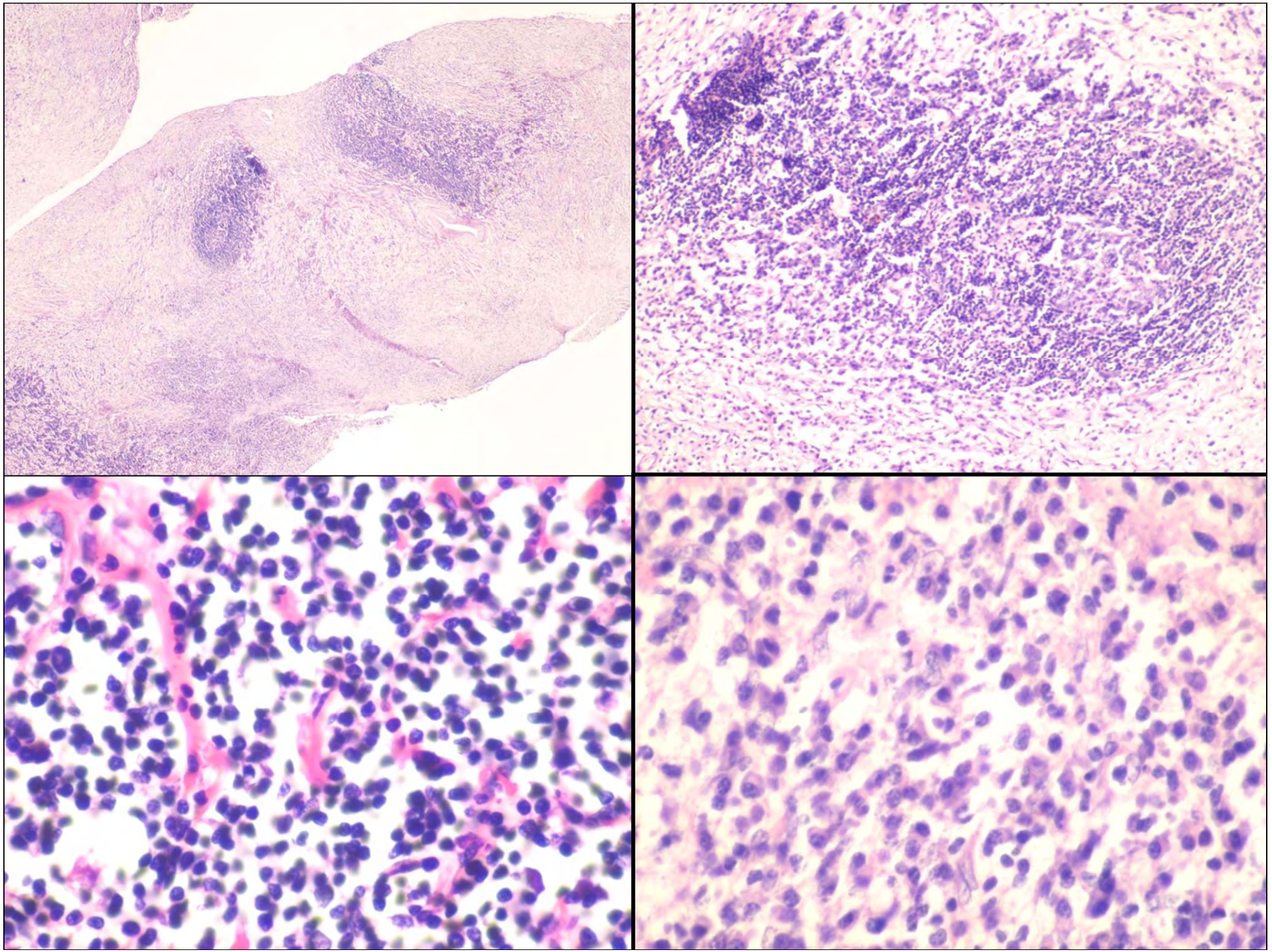
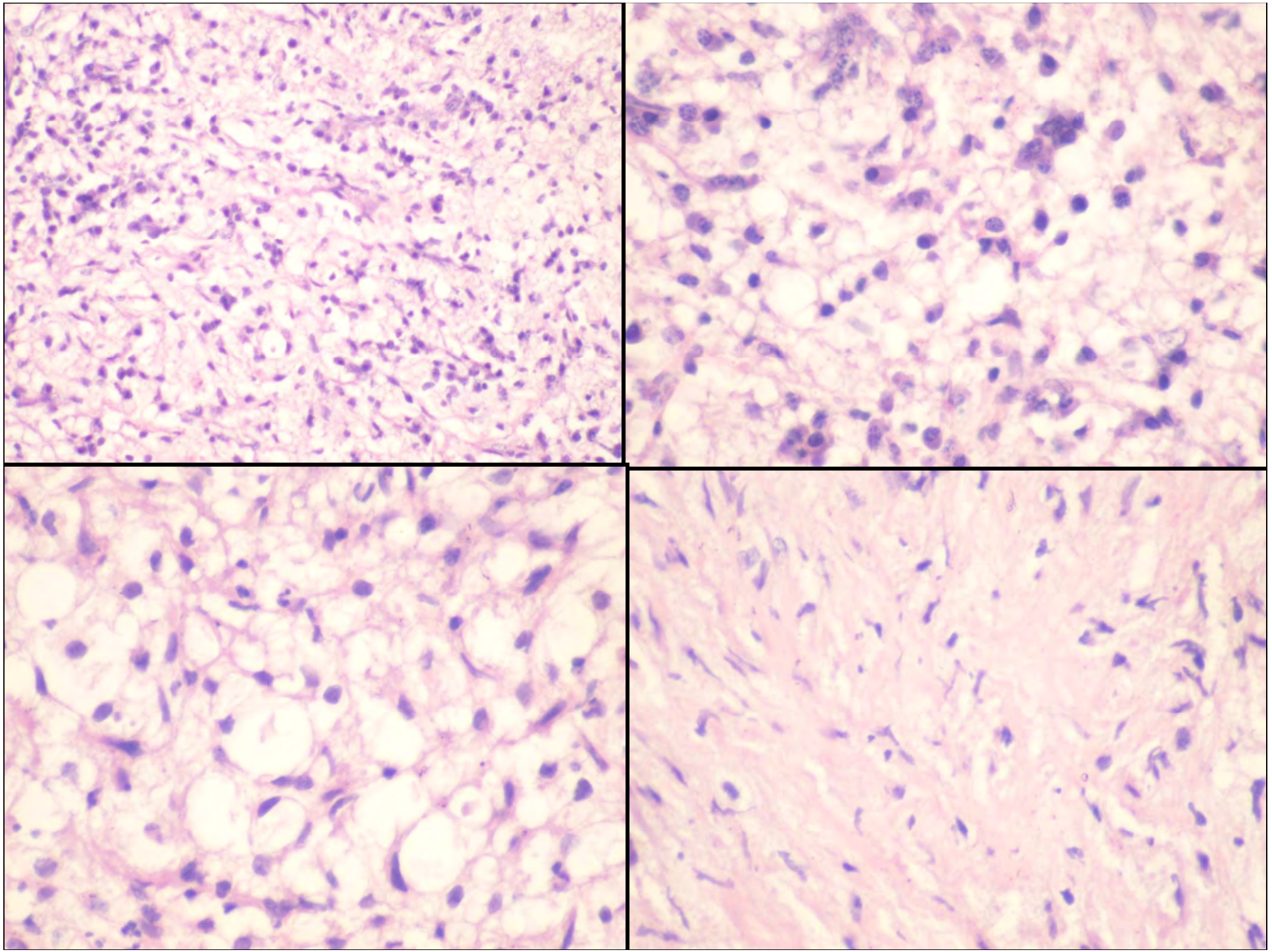




Summary of Microscopic Features

- Granulomatous inflammation
- Marked infiltration of lymphocytes and plasma cells
- Lymphpoid aggregation
- Large amount of xanthomatized histiocytes (foamy cytoplasm)
- Hyalinization and fibrosis

Differential Diagnosis

- **Granulomatous lesions**
  - **Xanthogranuloma**

    xanthomatized histiocyte, lymphoid follicles
  - **Tuberculosis, Leprosy**
  - **Sarcoidosis**
- **Histiocytic lesion – Langerhan cell histiocytosis**
- **Benign lymphoepithelial lesions**
  - **Sjogren syndrome, Mikulicz's diseases**

    epi-myoepithelial islands

Histopathological Report

- **Xanthogranuloma**

  Right parotid gland, Incision

No. KMUOP-09-1597

Gross Findings

- **Size**

  R't parotid gland

  - 7.0x5.0x3.0 cm

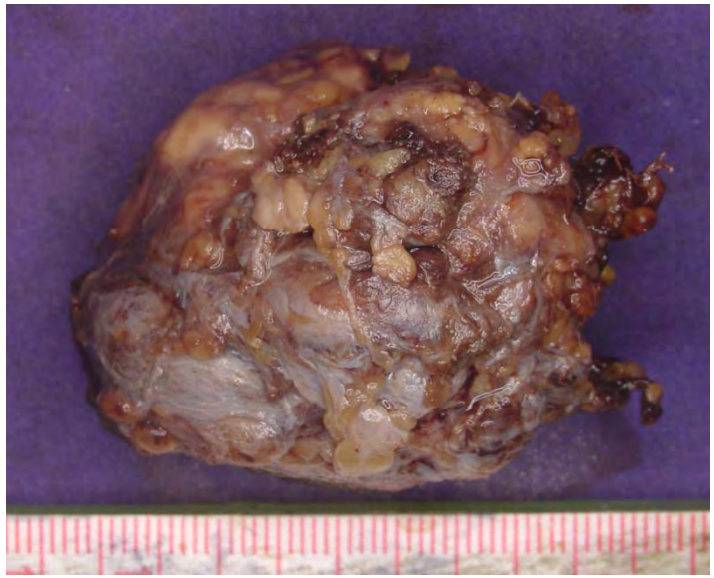
  R't SMD gland

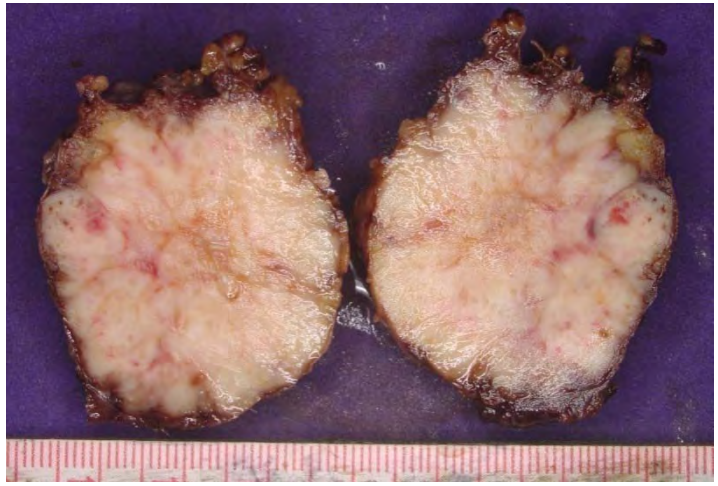
  -

  R't buccal tumor

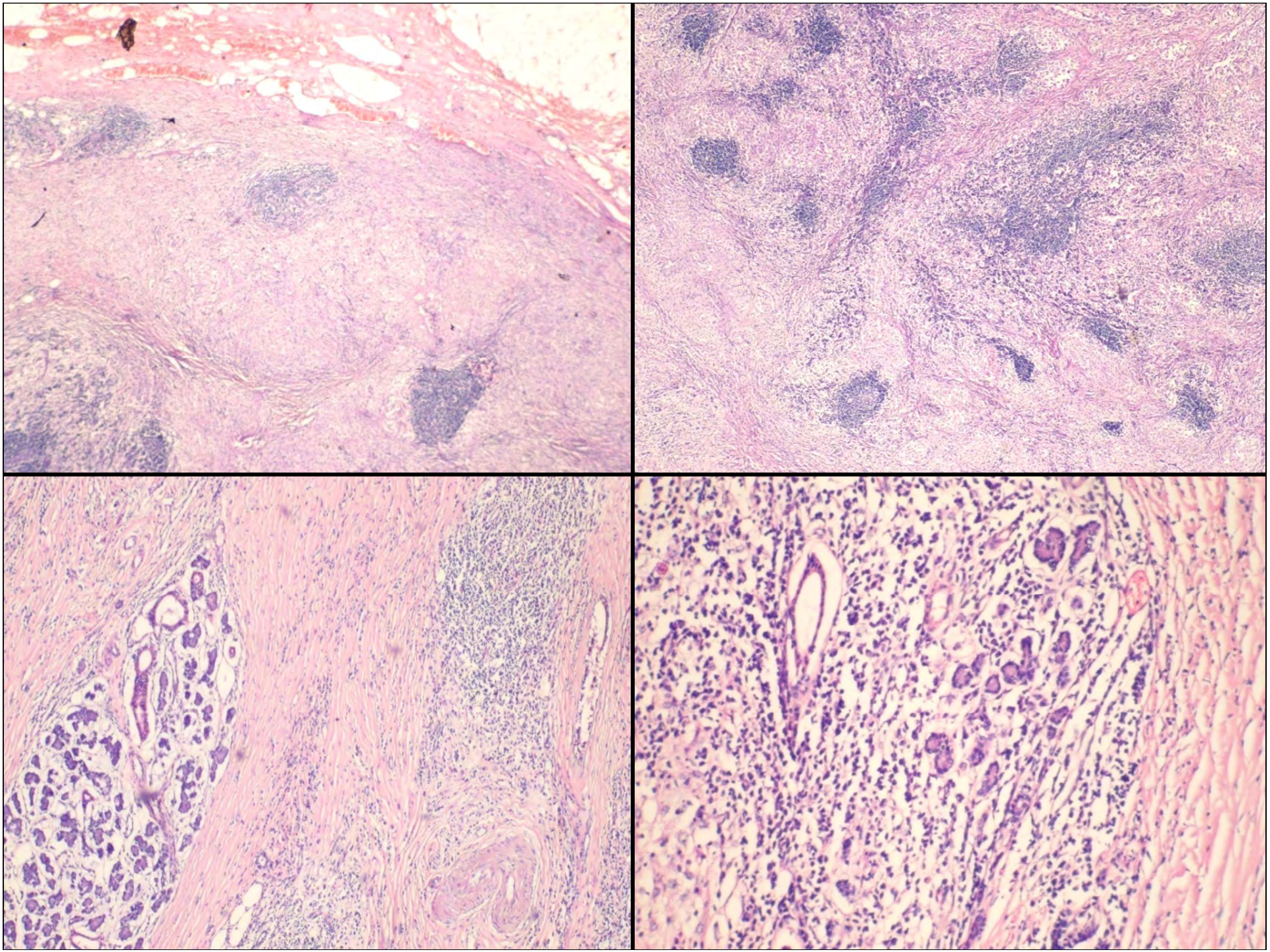
  -

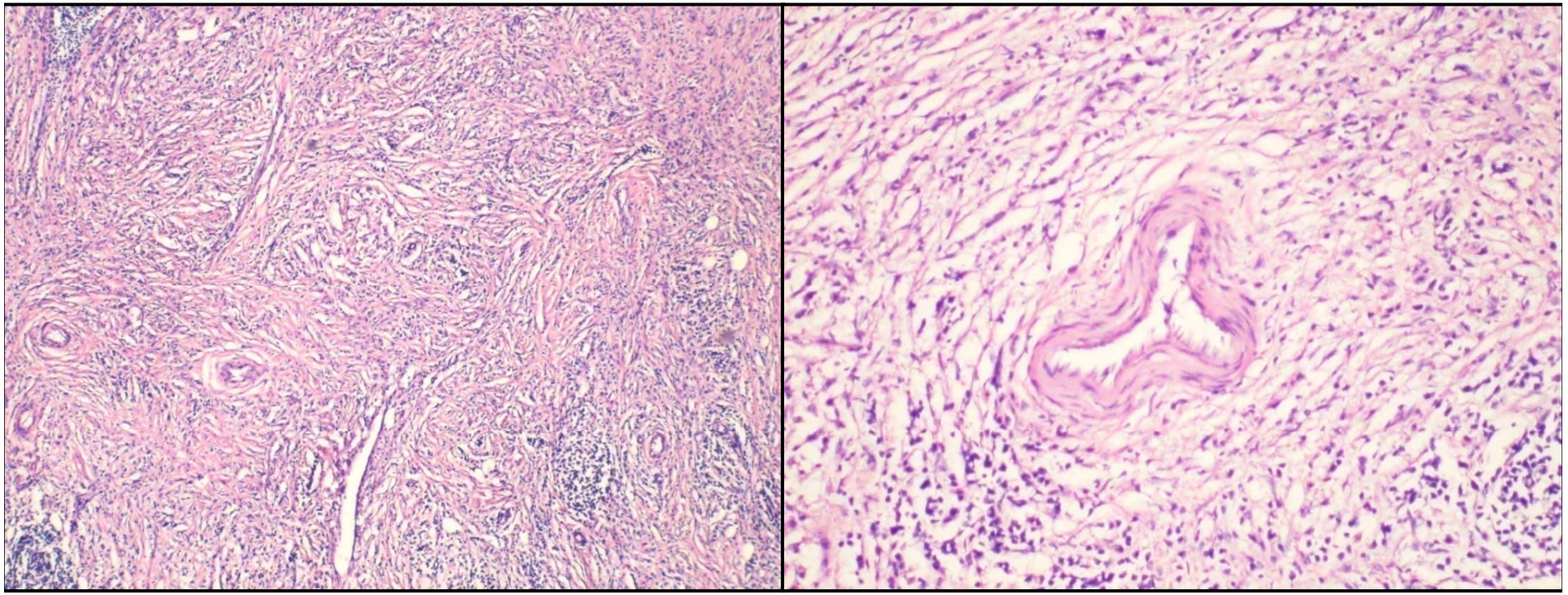
- **Consistency: firm**
- **Cut surface: a well-circumscribed, solid mass**
- **Color: whitish**

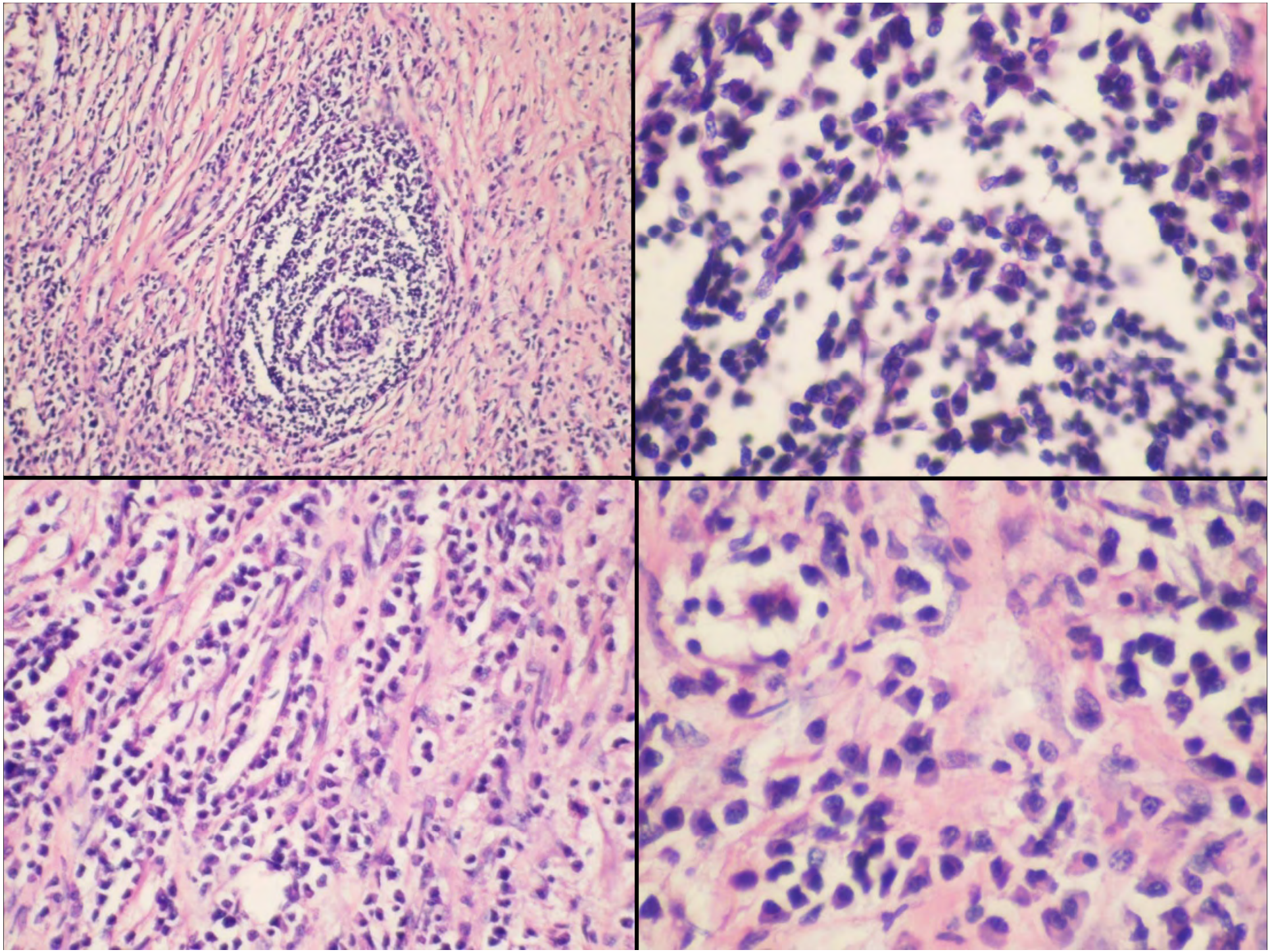


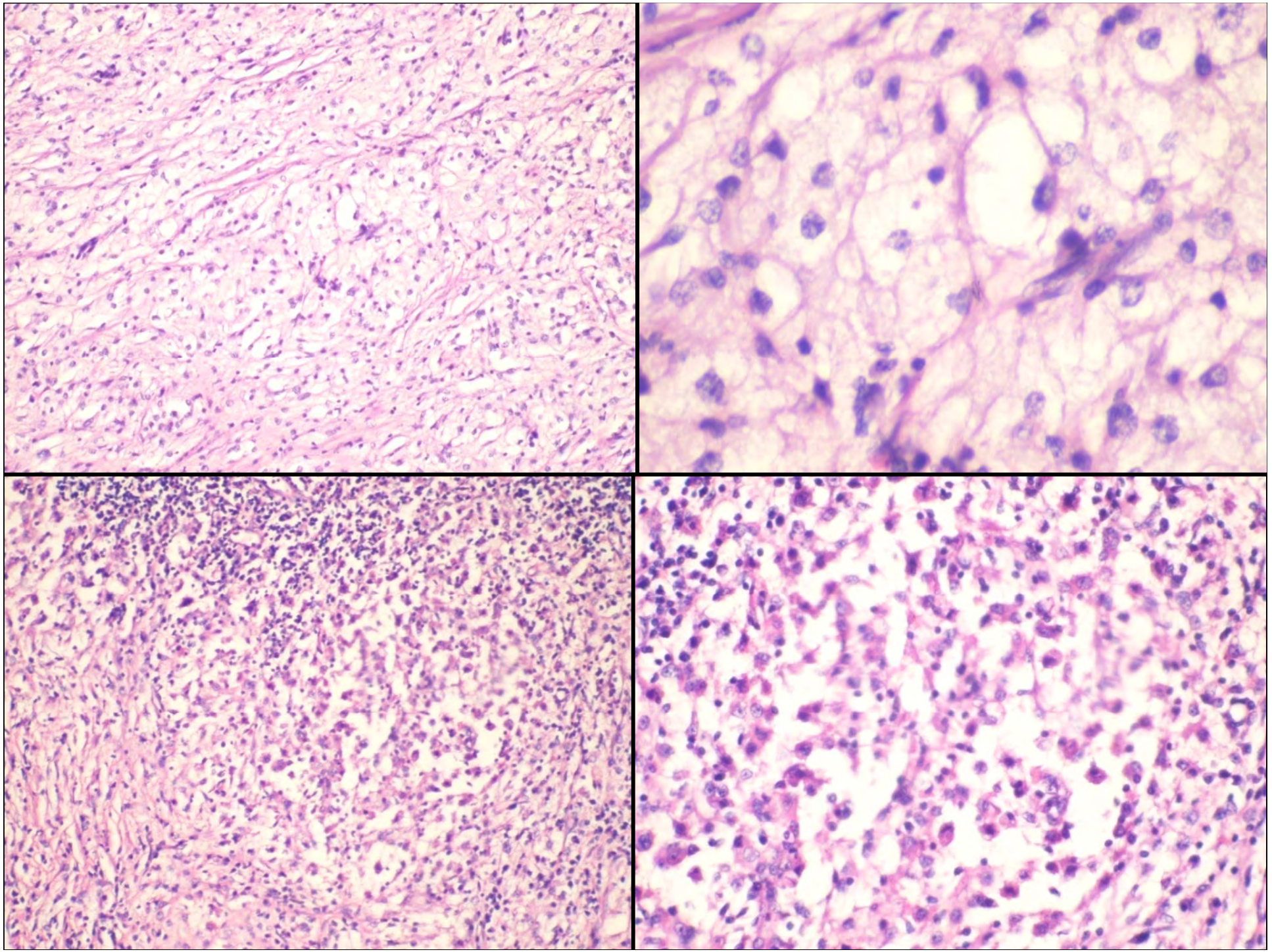


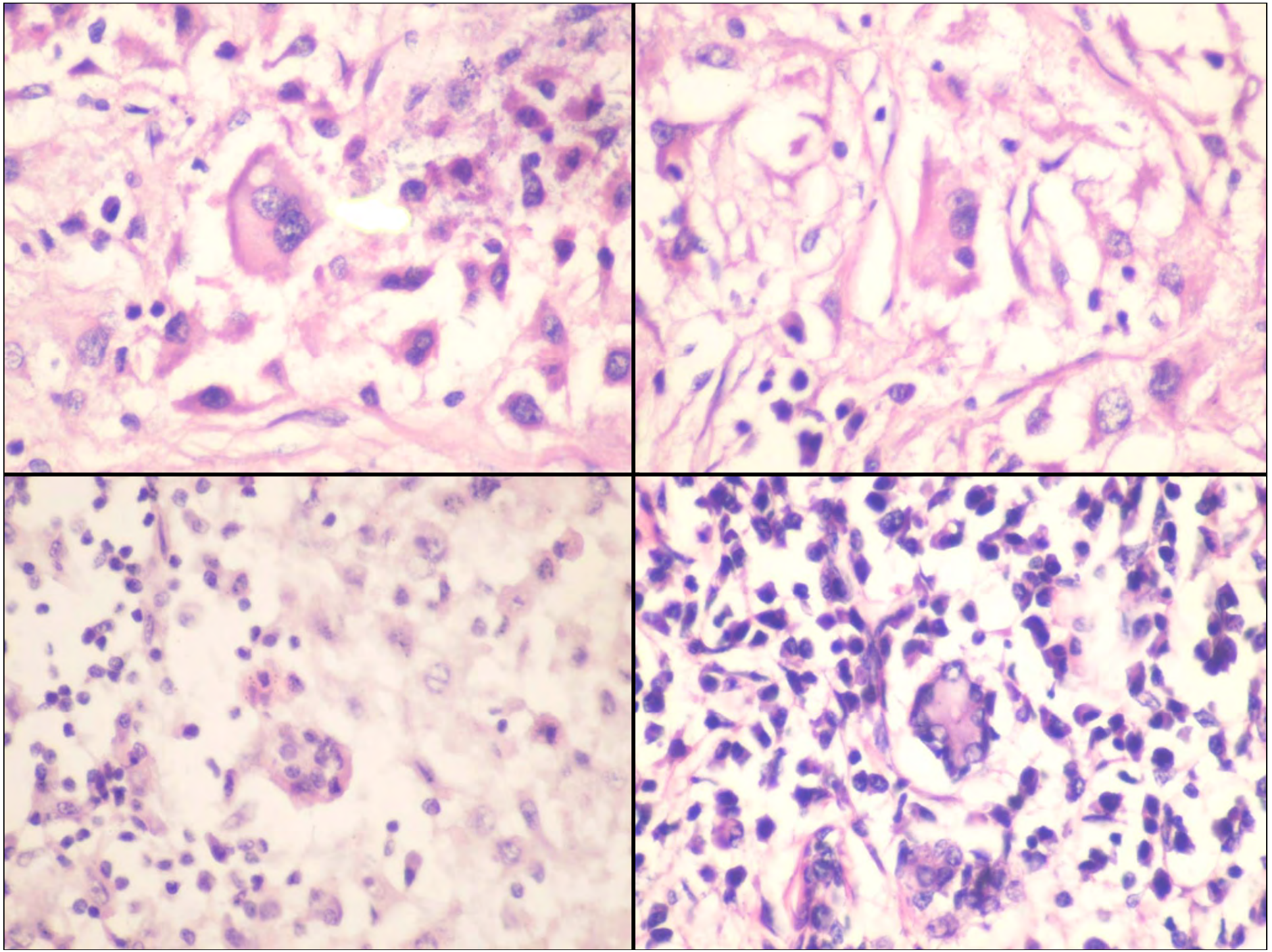
Right Submandibular Gland And Lymph Nodes

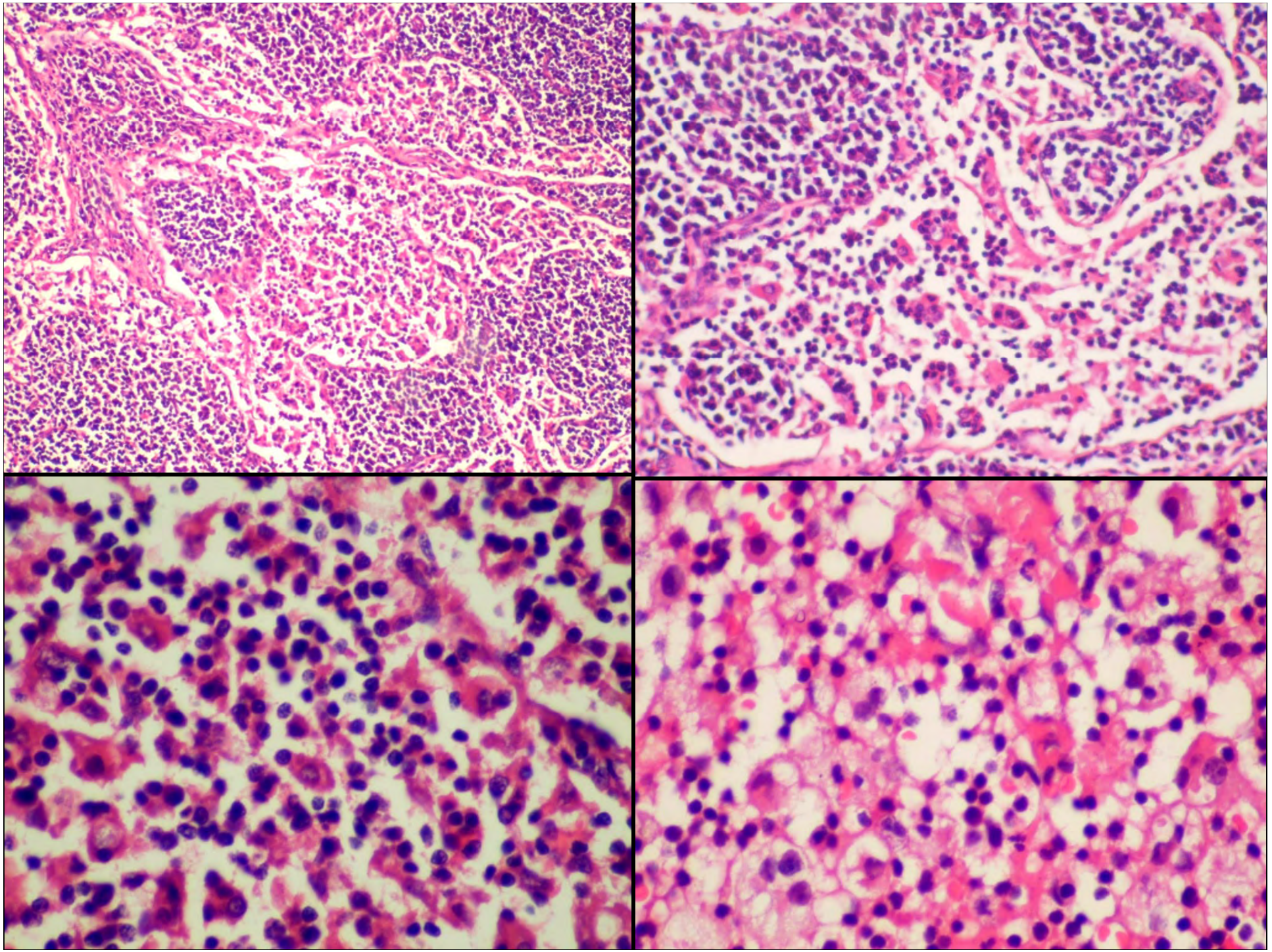




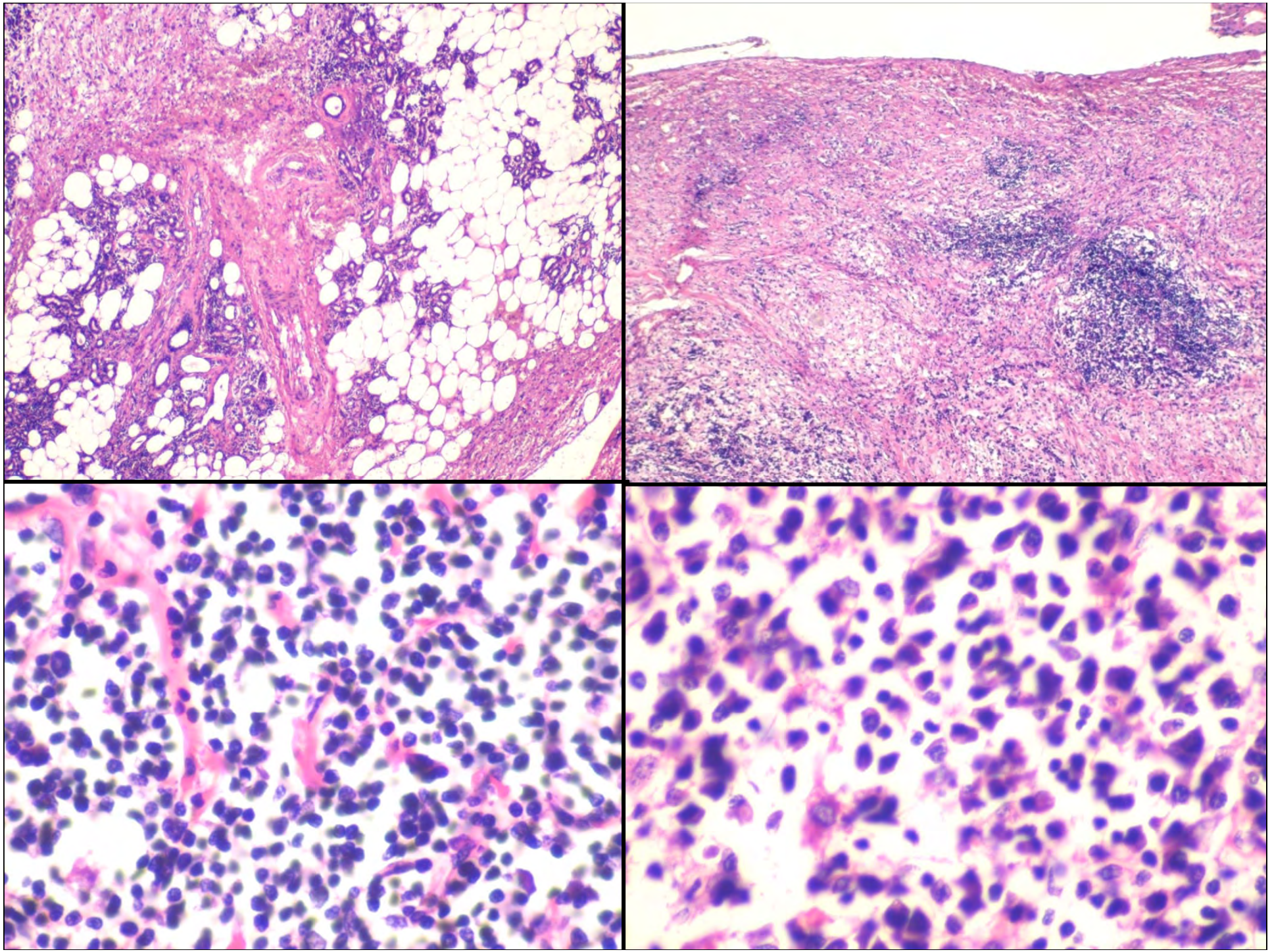


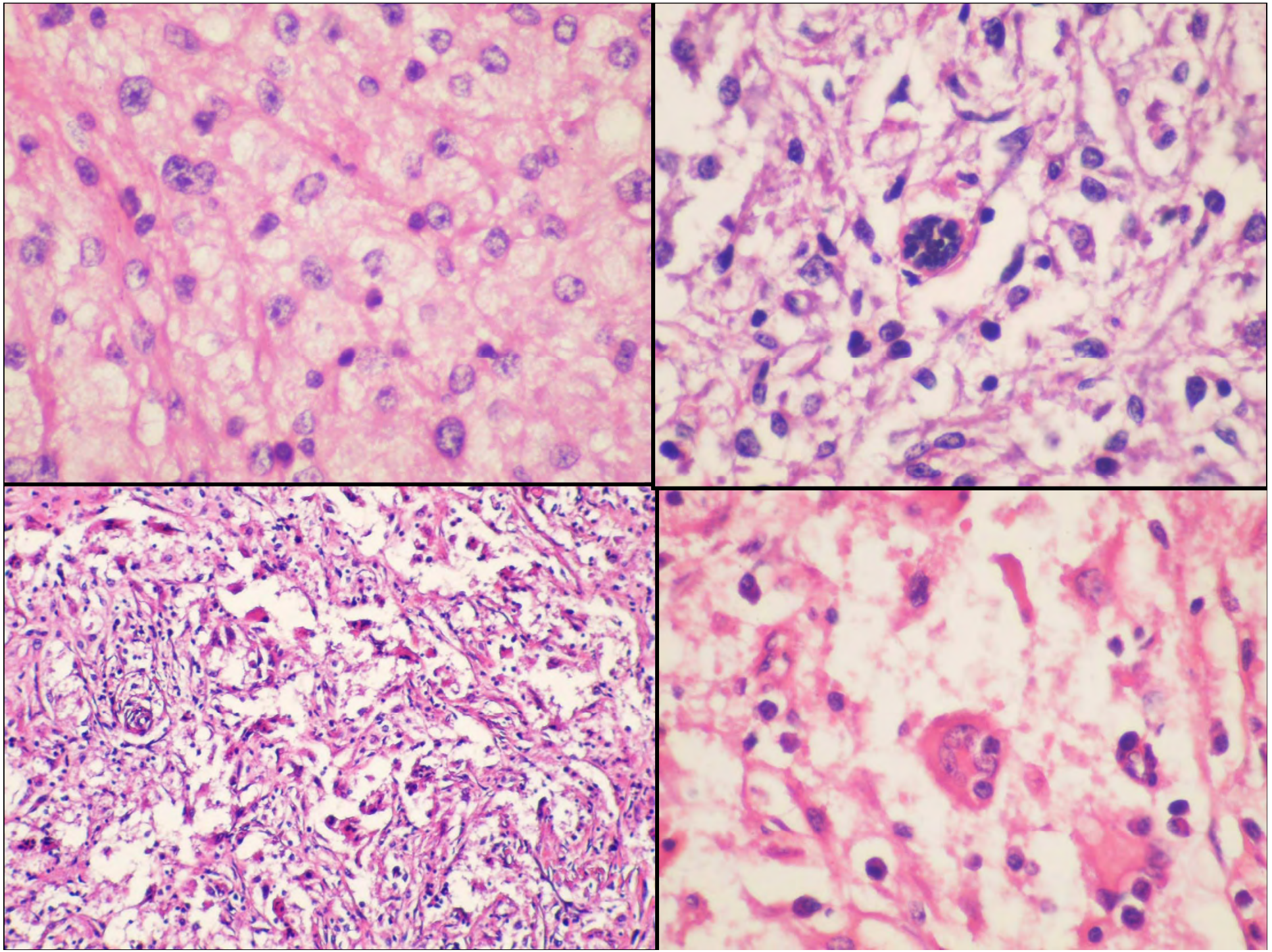




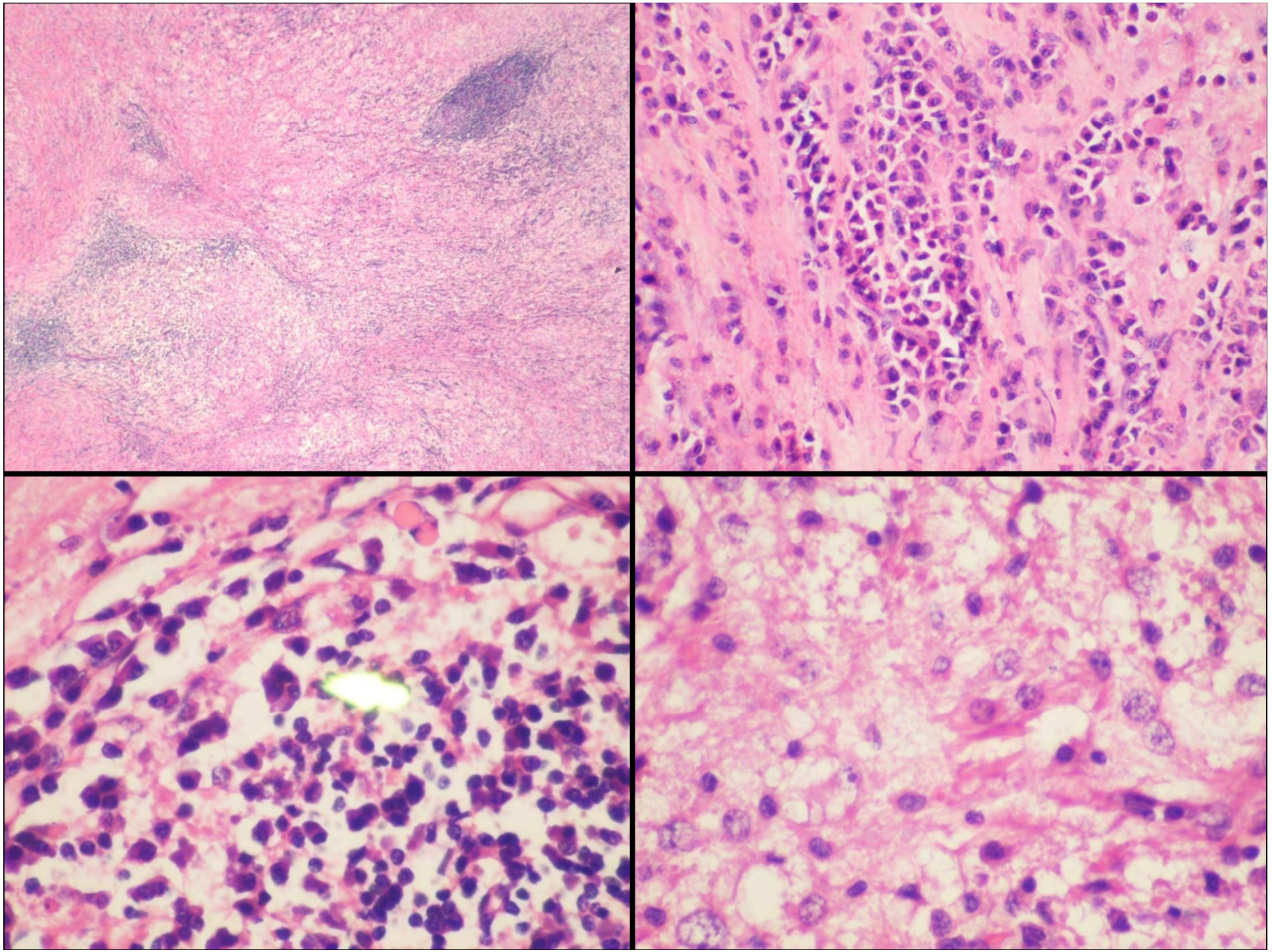


Right Parotid Gland





Right Buccal Tumor



Special Stains

- **Histochemical stain**

  PAS – mucin, polysaccharide
- **Immunohistochemical stains**

  CD68 – histiocyte

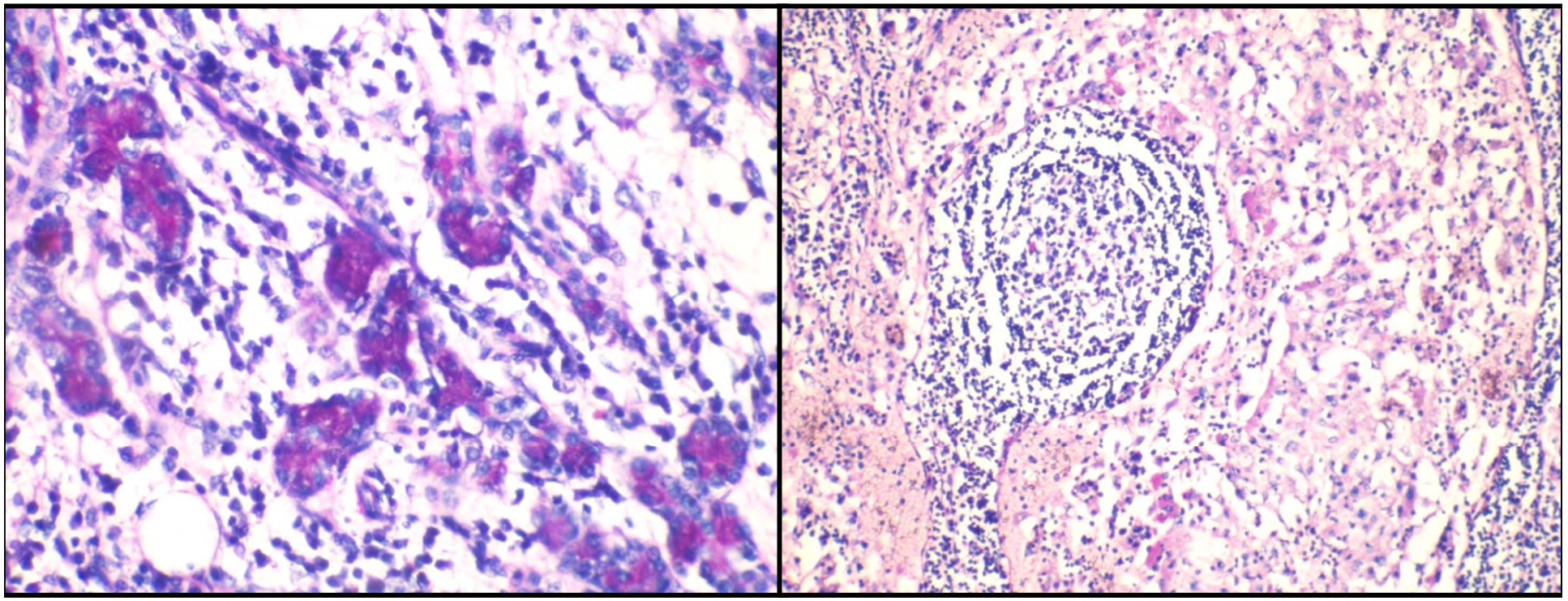
  CD1a – Langerhan cell

  S-100 – Langerhan cell, myoepithelial cell

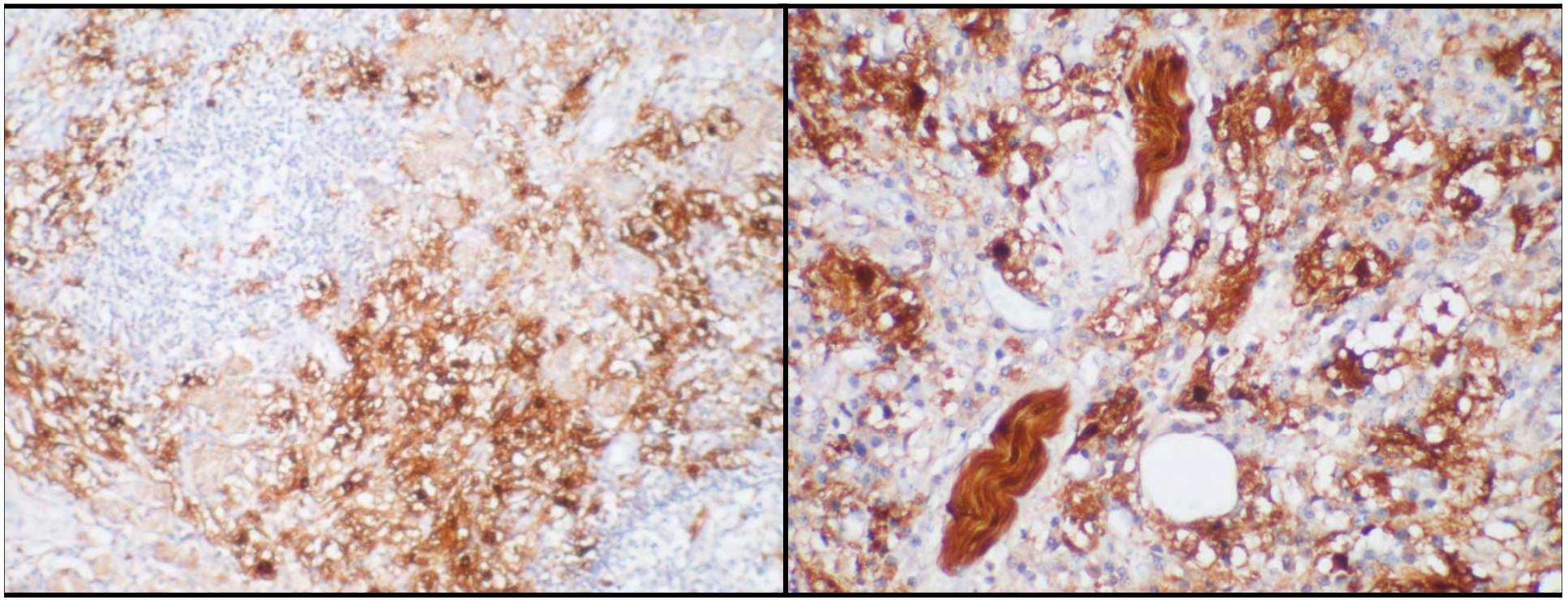
  T-cell – T lymphocyte

  B-cell – B lymphocyte

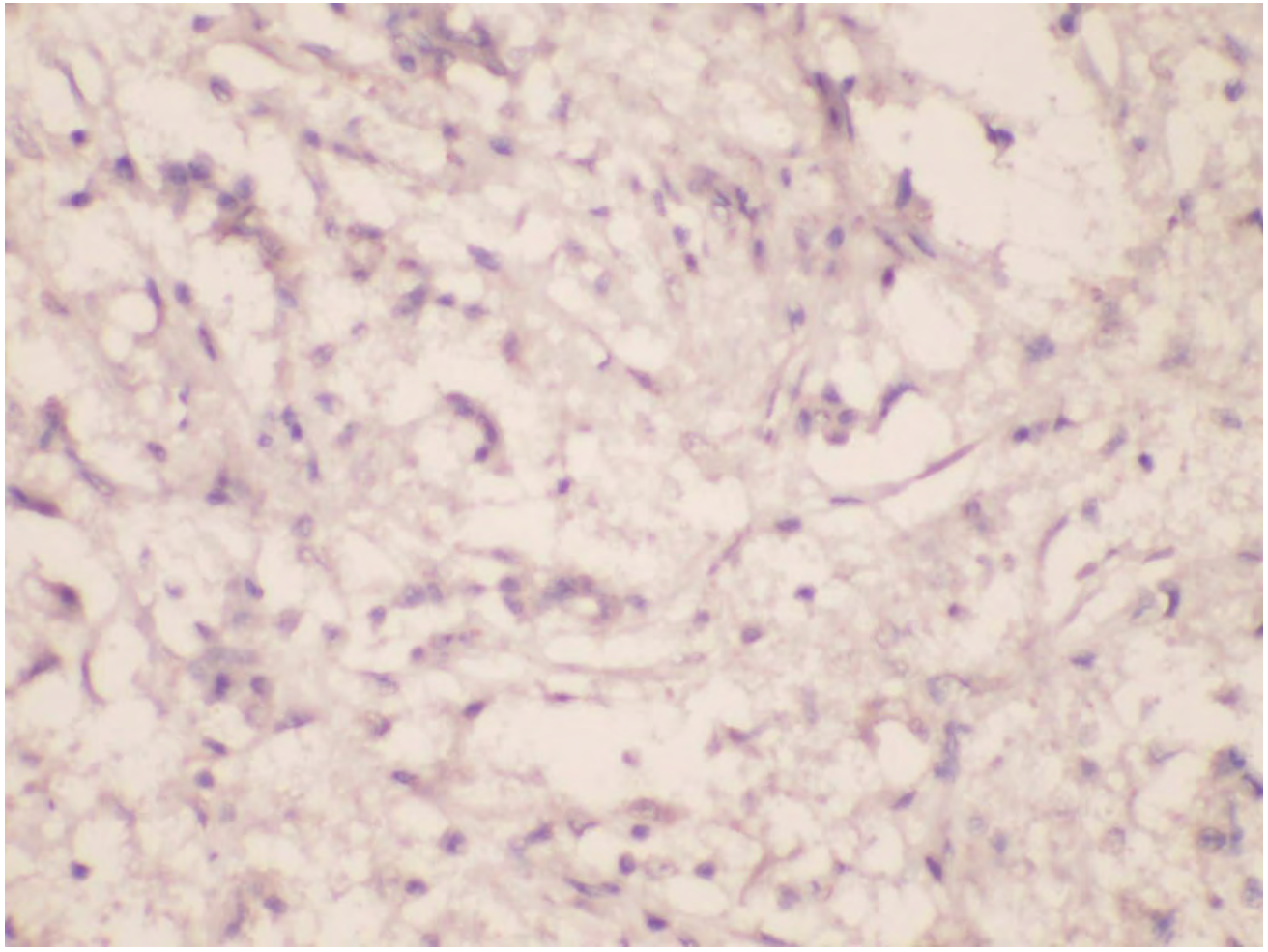
PAS(-)



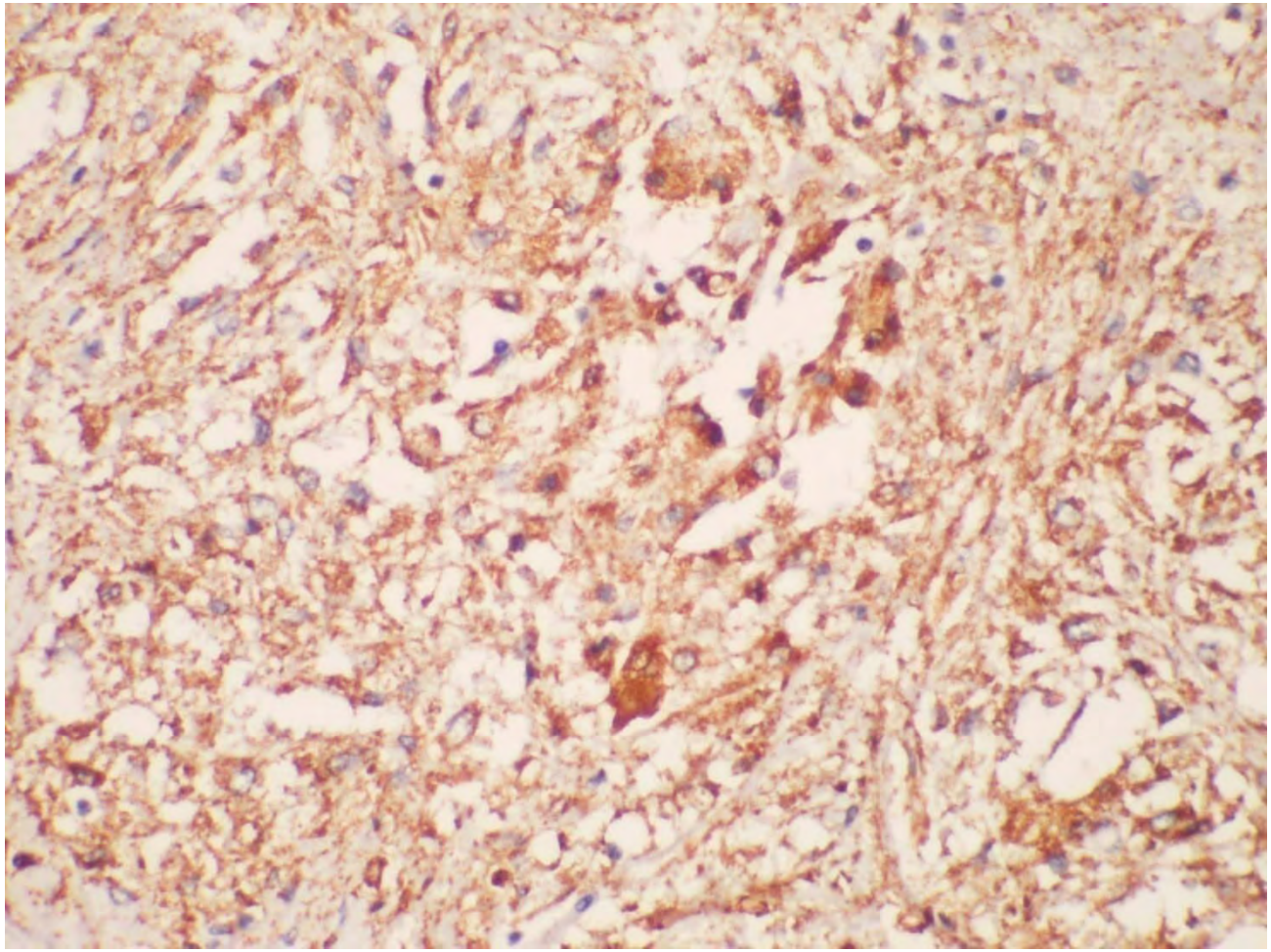
S-100 (+)



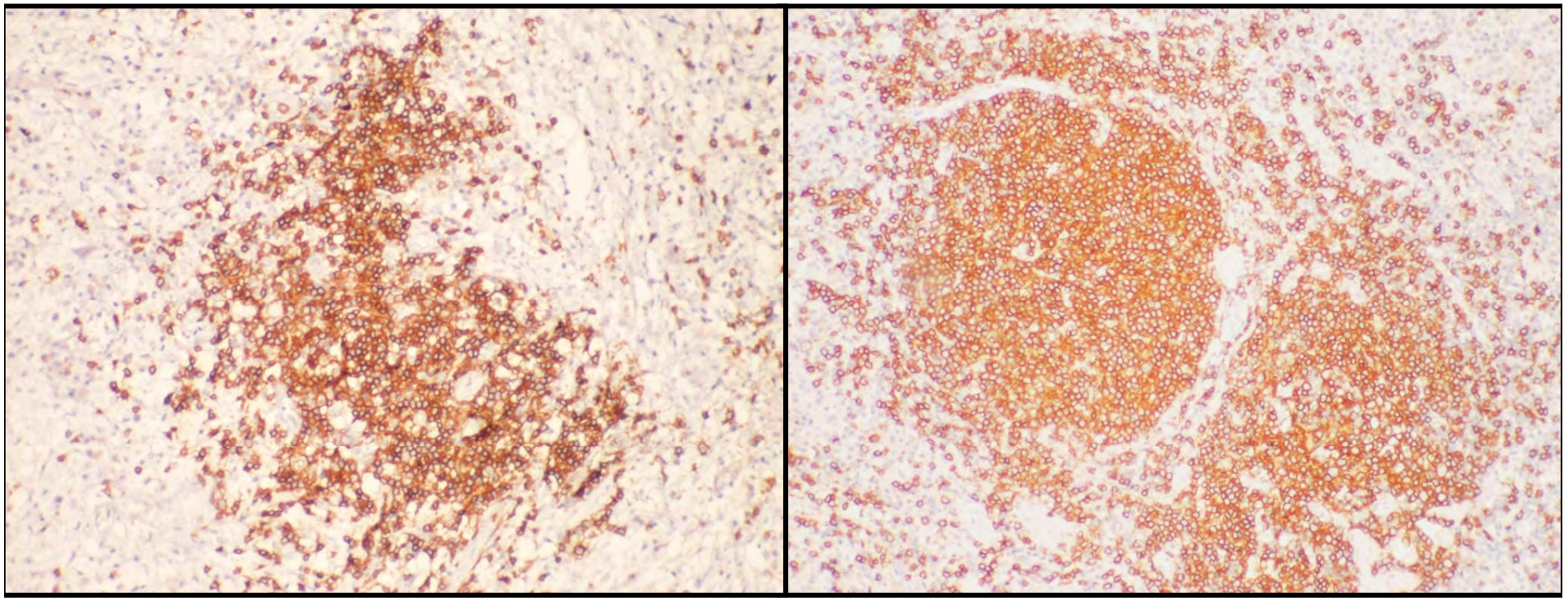
CD1a(-)



CD68(+)



T-cell (+) And B-cell (+)



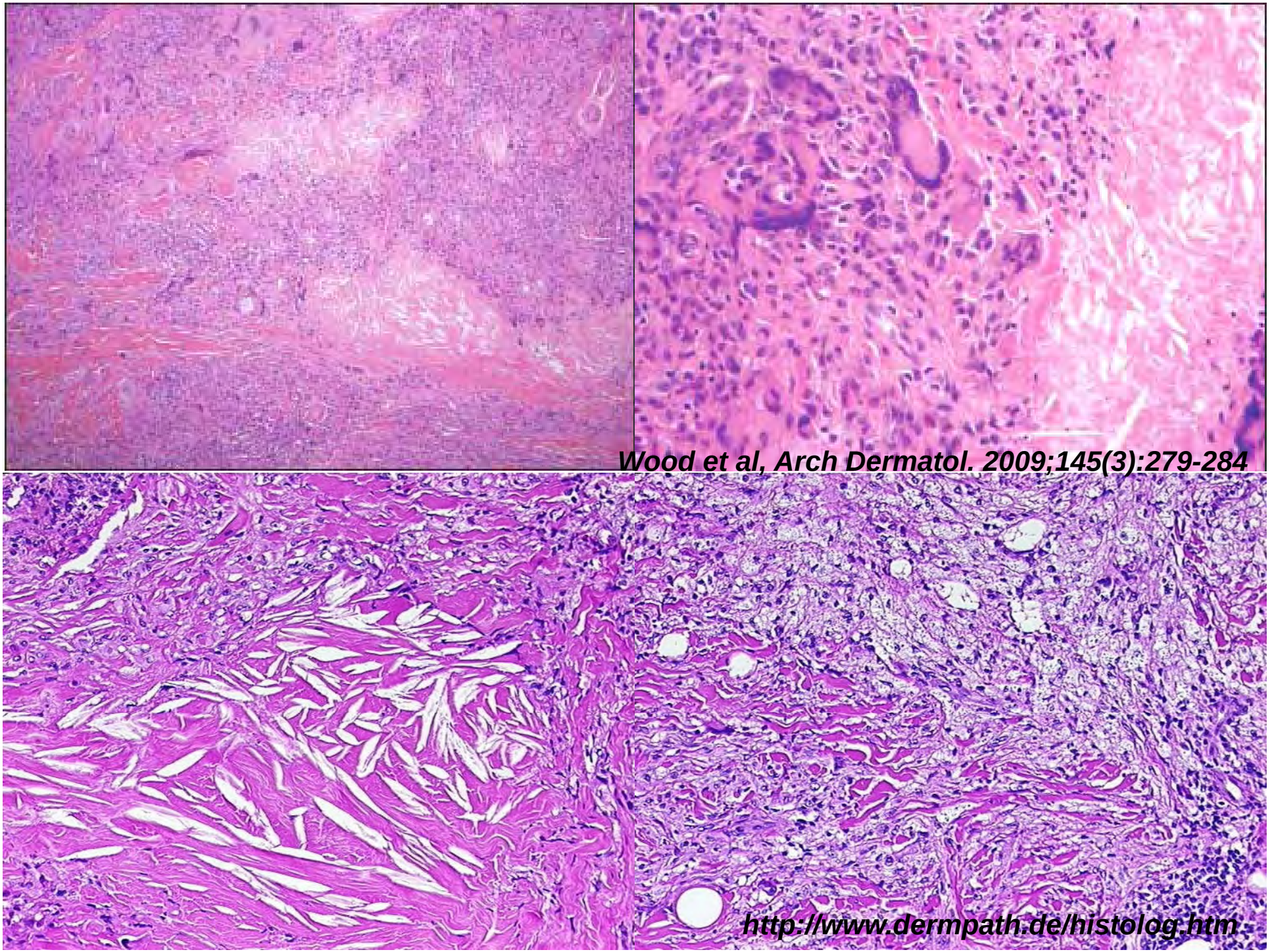
Results Of Special stains

- PAS (-) – mucin (X), polysaccharide(X)
  lipid (?)
- S-100 (+) but CD1a (-) – Langerhan cell (X)
- CD68 (+) – Histiocyte (O)
- T-cell (+) and B-cell (+) with even distribution
  - admixture

Histopathological Features of NXG in Literatures

	NXG	Our Case
Necrobiosis	+, 100%	+
Granulomatous inflammation	+, 83%	+
Lymphoid infiltration	+, 100%	+
Xanthomatized histiocytes	+, 100%	+
Touton and foreign body giant cells	+, 83%	+
Cholesterol clefts	+, 33%	-
Fibrosis, slcerosis	-	+
Sinus histiocytosis in LN	-	+
S-100	-	+

Wood et al, Arch Dermatol. 2009;145(3):279-284; Fernandez-Herrera et al, Semin Cutan Med Surtg. 2007;26:108-113


Wood et al, Arch Dermatol. 2009;145(3):279-284
http://www.dermpath.de/histolog.htm

Histopathological Report

- **Necrobiotic xanthogranuloma**

  Right parotid gland, Excision

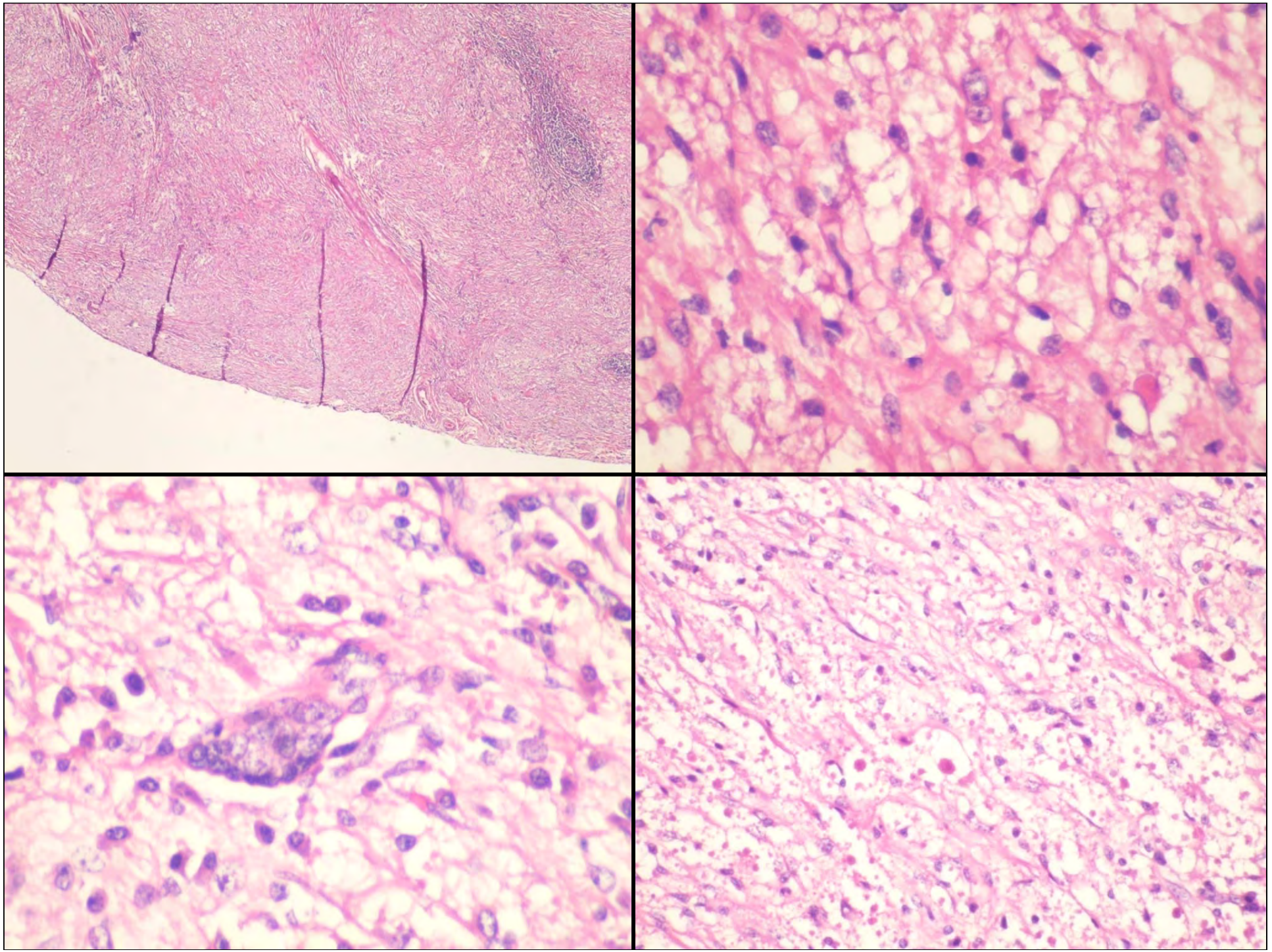
  Right submandibular gland, Excision

  Right buccal space, Excision

No. KMUOP-09-1899

Left Parotid Gland

Size: 4.5x3.8x2.9cm



Histopathological Report

- **Necrobiotic xanthogranuloma**

  Left parotid gland, Excision

No. KMUOP-09-2439

Slides Consultation

- **Dr. 陳授業 , Temple University, Philadelphia**

  - Chronic sclerosing sialadenitis
- **Dr. 鄭懿興 and Dr. Wright, Baylor University, Texas**

  - Rosai-Dorfman disease

  - Chronic Sclerosing sialadenitis
- **Dr. Solt and Dr. Reza , Northwestern University, Illinois**

  - Rosai-Dorfman disease

Summary

- **The nature of lesion**

  - Inflammatory /reactive, not neoplastic

- **Diagnoses**

  1. Rosai-Dorfman Disease

  (Sinus Histiocytosis with Massive Lymphadenitis, SHML)

  2. Chronic sclerosing sialadenitis

  3. Other histiocytic or granulomatous lesions

  eg: Langerhan cell histiocytosis

- **Neither of these diagnoses fit very well.**

Discussion

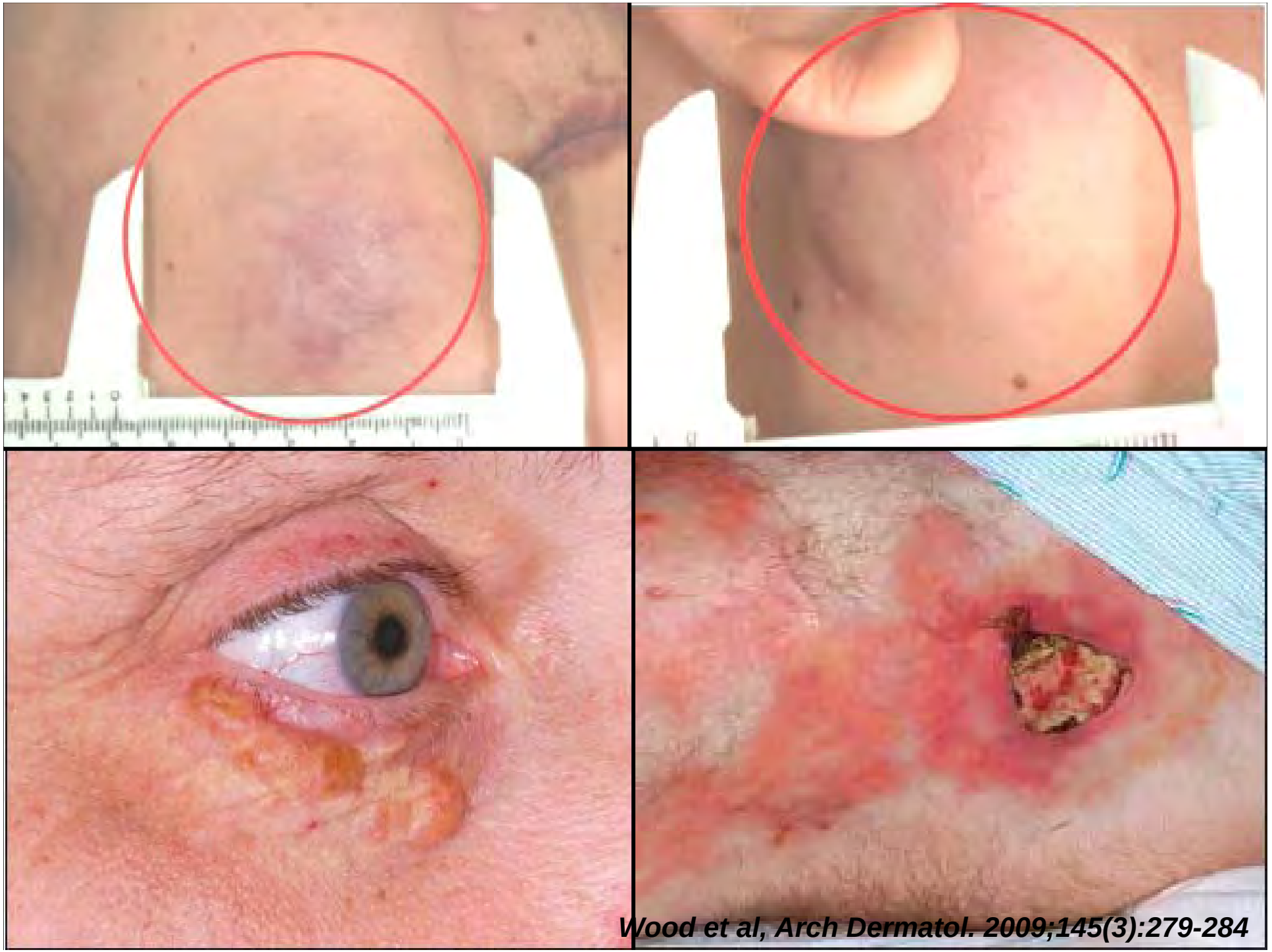
- **Introduction of necrobiotic xanthogranuloma(NXG), Rosai-Dorfman disease(RDD), and chronic sclerosing sialadentitis(CSS)**
- **Comparison between our case and RDD, CSS**

Necrobiotic Xanthogranuloma

- A rare chronic granulomatous lesion, about 100 cases in literatures, class II histiocytic disorder
- Multiple involvement of skin, most common site – periorbital area (80-90%), may involve extracutaneous sites, parotid gland: 1 case
- Skin lesion – multiple indurate yellow-red plaques or nodules, slowly growing, telangiectasia, ulceration, scarring
- Associated with lymphoproliferative disorder

  - Paraproteinemia (80%), multiple myeloma (10%),

Wood et al., Arch Dermatol. 2009;145(3):279-284; Fernandez-Herrera et al., Semin Cutan Med Surtg. 2007;26:108-113; Zainal et al., J Laryngol & Otol. 2009

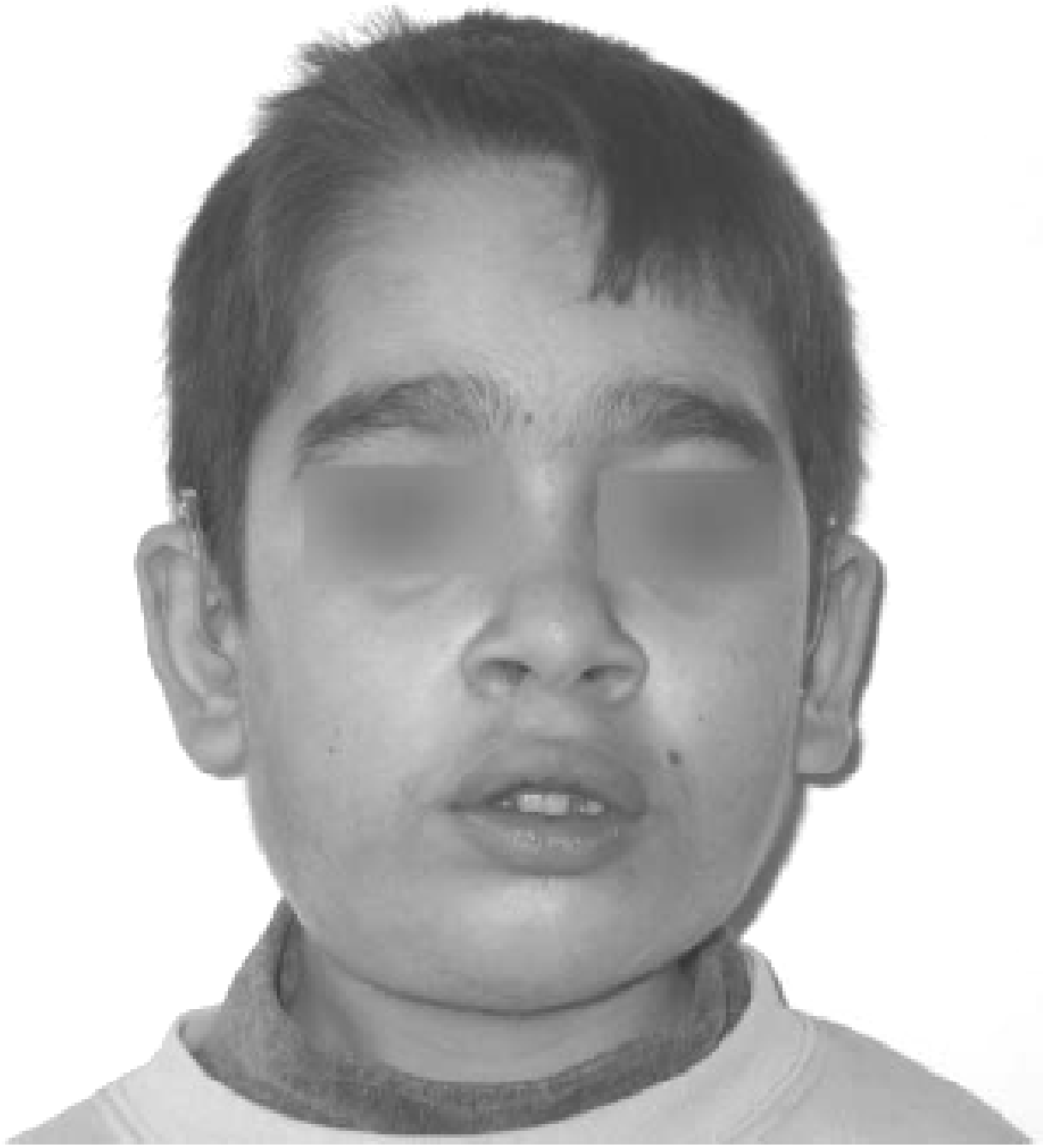


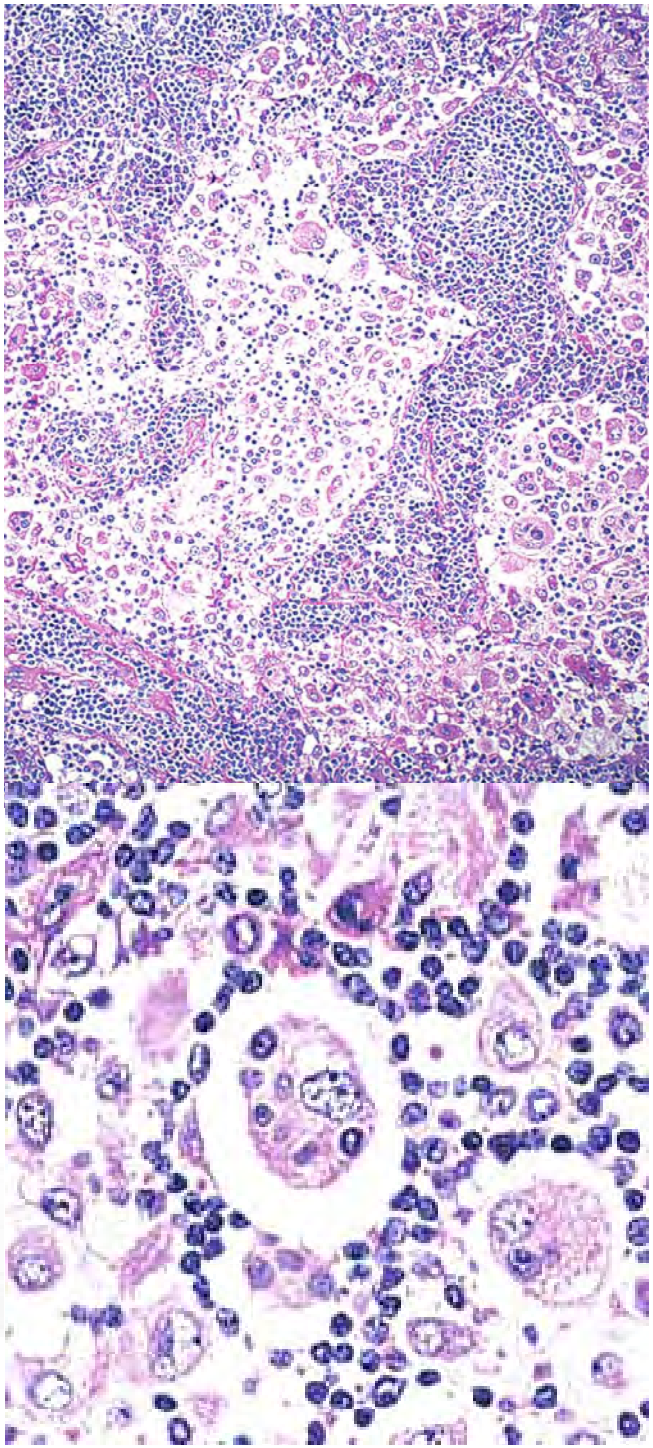
Wood et al, Arch Dermatol. 2009;145(3):279-284

Rosai-Dorfman Disease

- An uncommon benign systemic histioproliferative disorder
- Multiple, bilateral enlarged lymph nodes, most common – cervical, extranodal sites (25-43%) – head and neck, major salivary gland: 22 cases
- Age – 80% < 20 y/o
- May be accompanied by fever, malaise, weight loss, leukocytosis and hyperglobulinemia
- Proliferative histiocytes surrounded by lymphocytes and plasma cells, S-100(+), emperipolesis

Panikar et al.,Diag Cytopathol. 2005;33:187-190; Huang et al., Ann Acad Med Singapore 1998;27:589-93





Guven et al., Dentomaxillofacial Radiology 2007;36:428-433;
http://www.pathconsultddx.com/pathCon/

Chronic Sclerosing Sialadenitis

- **An uncommon chronic inflammantory disorder in salivary gland, esp. submandibular gland → Kuttner's tumor**
- **Systemic – sclerosing pancreatitis, colangitis, retroperitoneal fibrosis → IgG4-related sclerosing disease, multifocal fibrosis**

  Localized – salivary gland → CSS
- **Serum immunoglobulin elevated, ANA elevated**
- **Lymphoplasmacytic infiltration, lymphoid follicles, eosinophilia, sclerosis, fibrosis**

Kitagawa et al, Am J Surg Pathol. 2005;29:783-791

Laboratory Studies

	NXG	RDD	CSS	Our Case
Serum Ig	⇧ > 80%	⇧	⇧	⇧
Gamma-pathy	Mono-clonal IgGκ 60% IgGλ 26% IgA 14%	Polyclonal	Mono-clonal IgG4	IgG
ESR		⇧		
ANA			⇧	

	NXG	RDD	CSS	Our Case
Necrobiosis	+	-	-	+
Granulomatous inflammation	+	-	-	+
Lymphoid infiltration	+	+	+	+
Xanthomatized histiocytes	+	+	-	+
Multinucleated giant cells	+ Touton	+	-	+
Cholesterol clefts	+	-	-	-
Fibrosis, slcerosis	-	-	+	+
Sinus histiocytosis	-	+	-	+
Emperipolesis	-	+	-	-
S-100	-	+	-	+
Eosinophilia	-	-	+	-

Diagnosis of Our Cases

Necrobiotic Xanthogranuloma

Conclusion

- 因爲NXG 的稀有性, 對診斷與治療形成很大的挑戰

  診斷 – 有一部份非典型的病理特徵

  治療 – 沒有正式或已形成共識的療法可參考

  手術 – facial nerve trauma
- 追蹤照護

  Xerostomia – rampant caries, 加強口腔衛生照護

  Lesions in left submandibular gland and posterior neck – 不影響外觀, 是否考慮以藥物治療

  Associated malignancy – lifelong follow-up

Thank you for your attention !