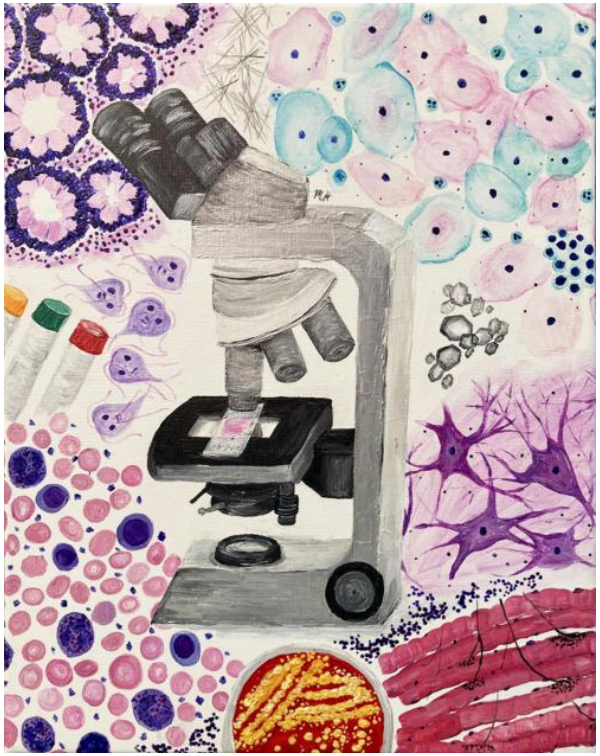
Case Presentation

Dr.Salar Nasr

Resident Of Pathology

MUI

Head & Neck Pathology



Attending Pathologist

Dr.Maryam Derakhshan

Patient History

30 years old

Left Sub-Mandibular mass

No Pain and any Symtoms

Report of Sonography

در محل لمس برجستگی در ناحیه ساب مندیبولار چپ تصویر یک توده هایپواکو با حدود مشخص به ابعاد ۳۶*۲۵ میلی متر مشاهده می شود که در تشخیص افتراقی آن آدنوم پلئومورفیک قرار می گیرد.

Figure.1 (X4)

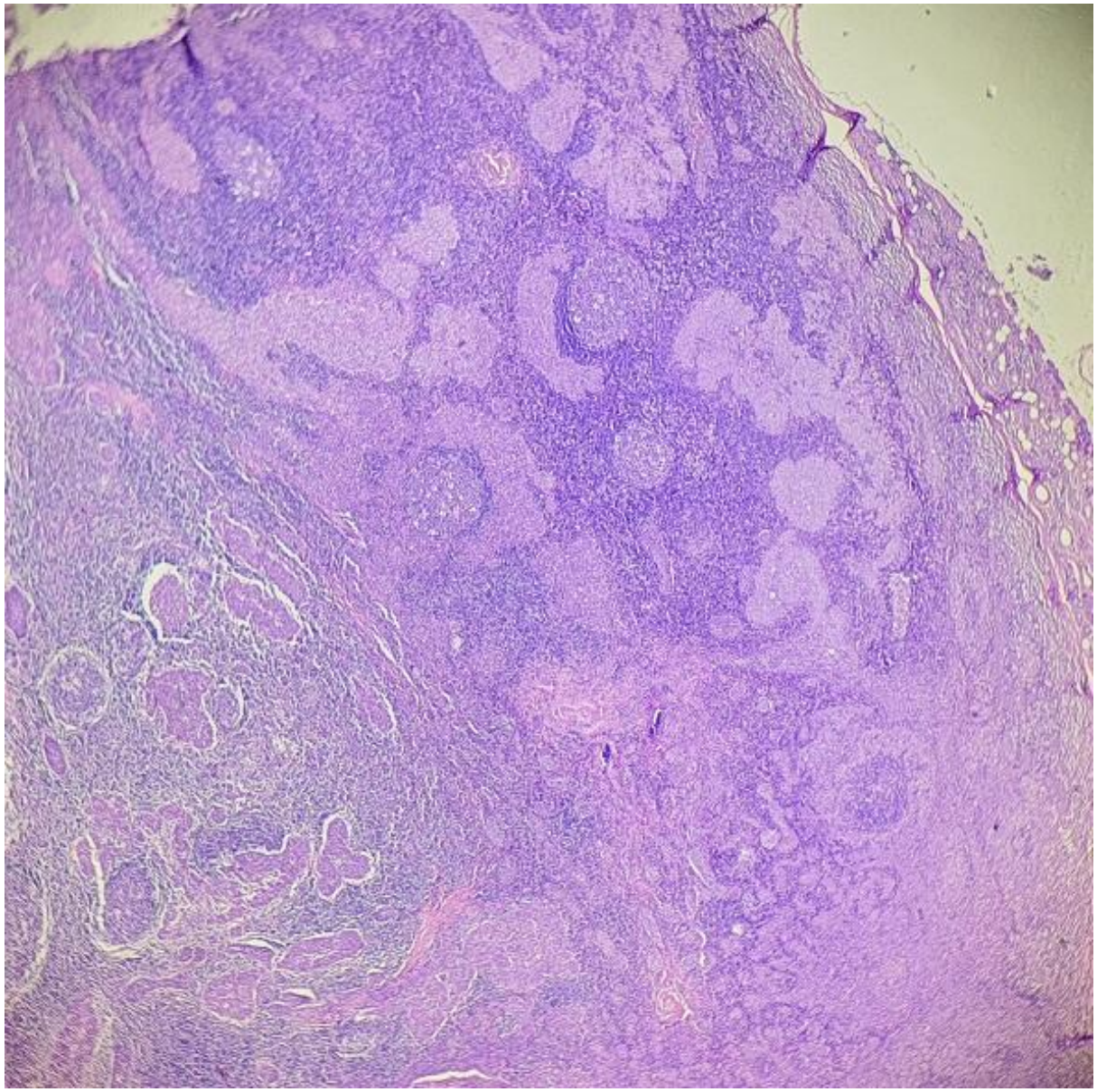
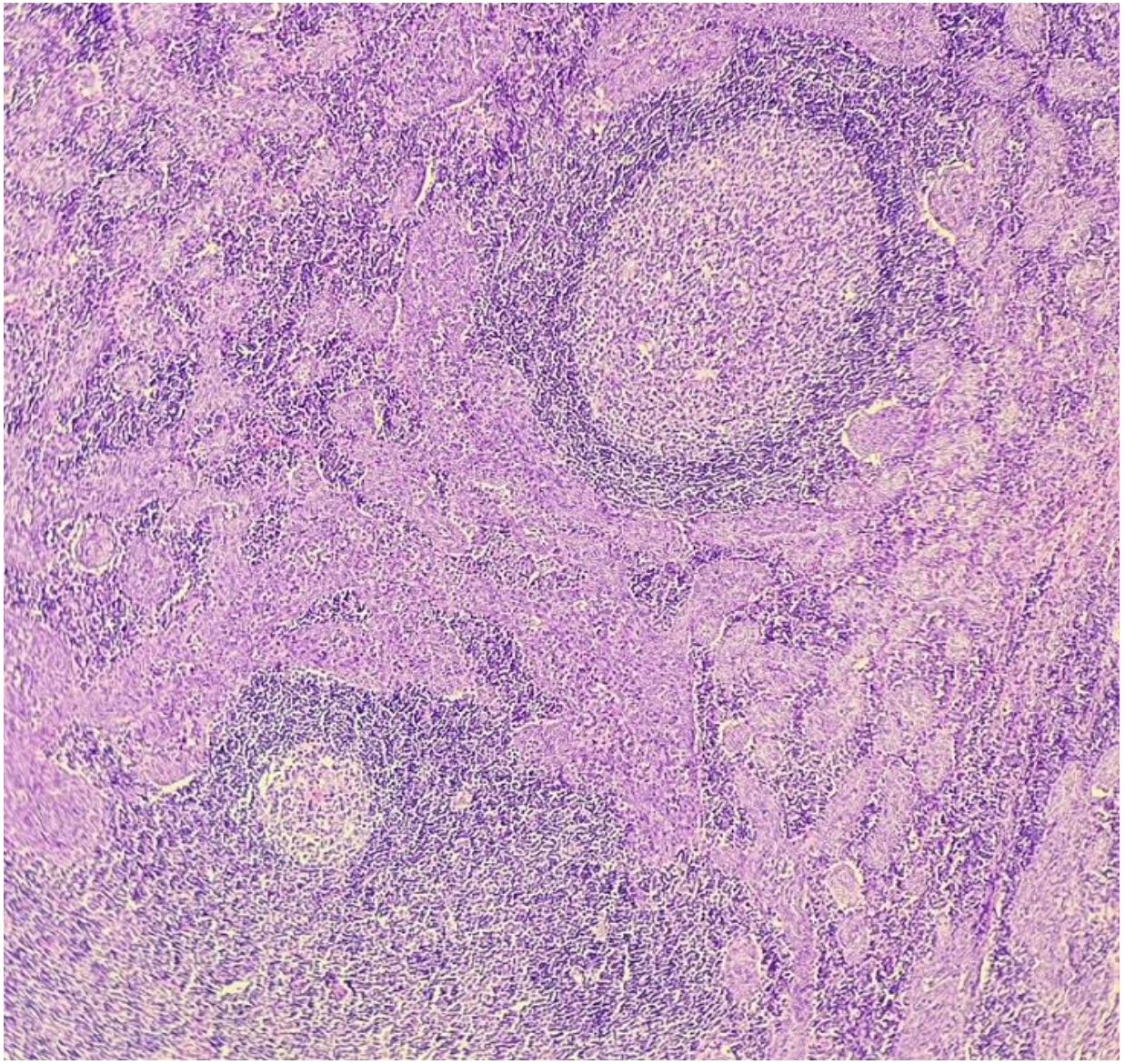


Figure.2 (X10)



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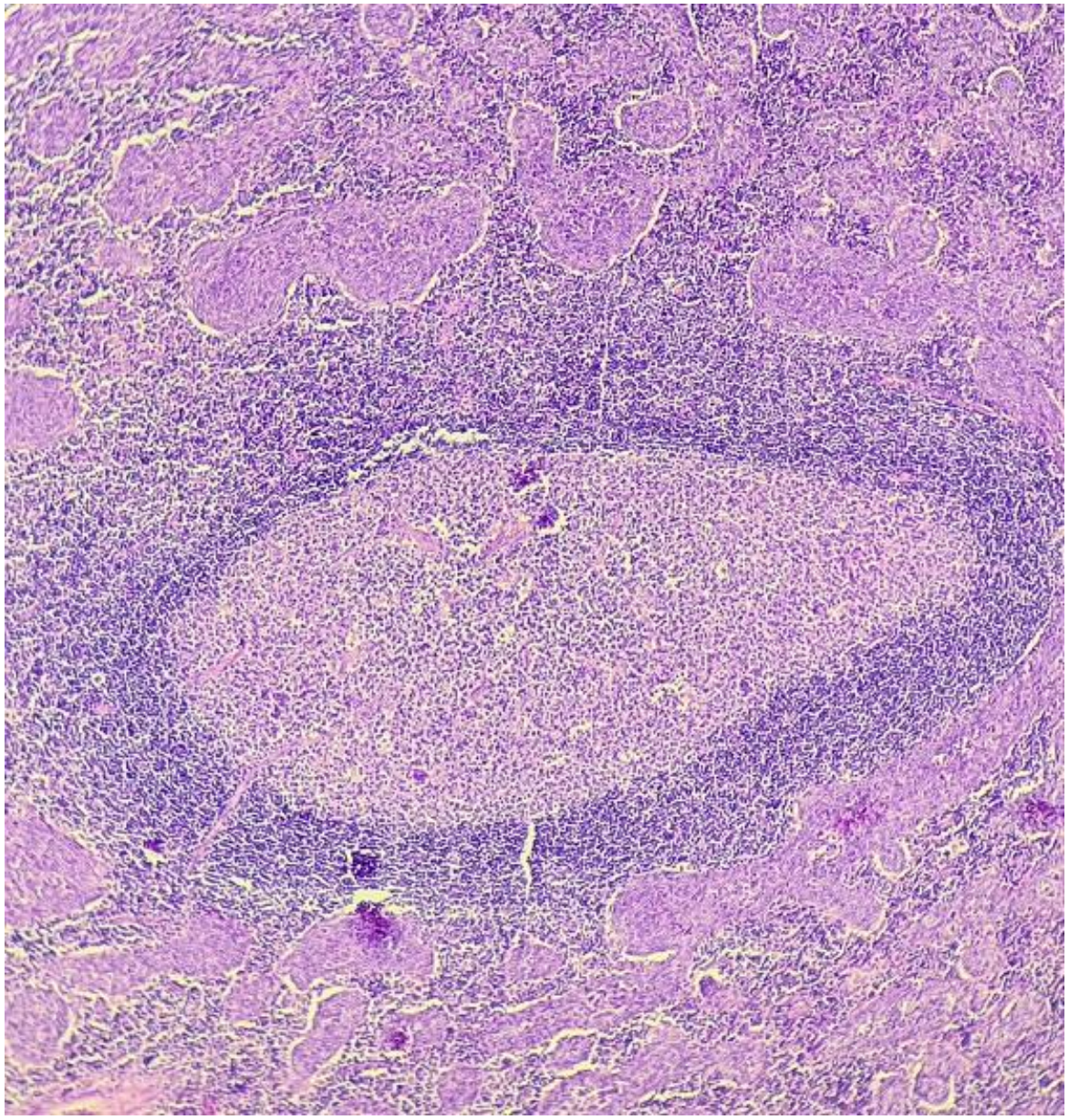

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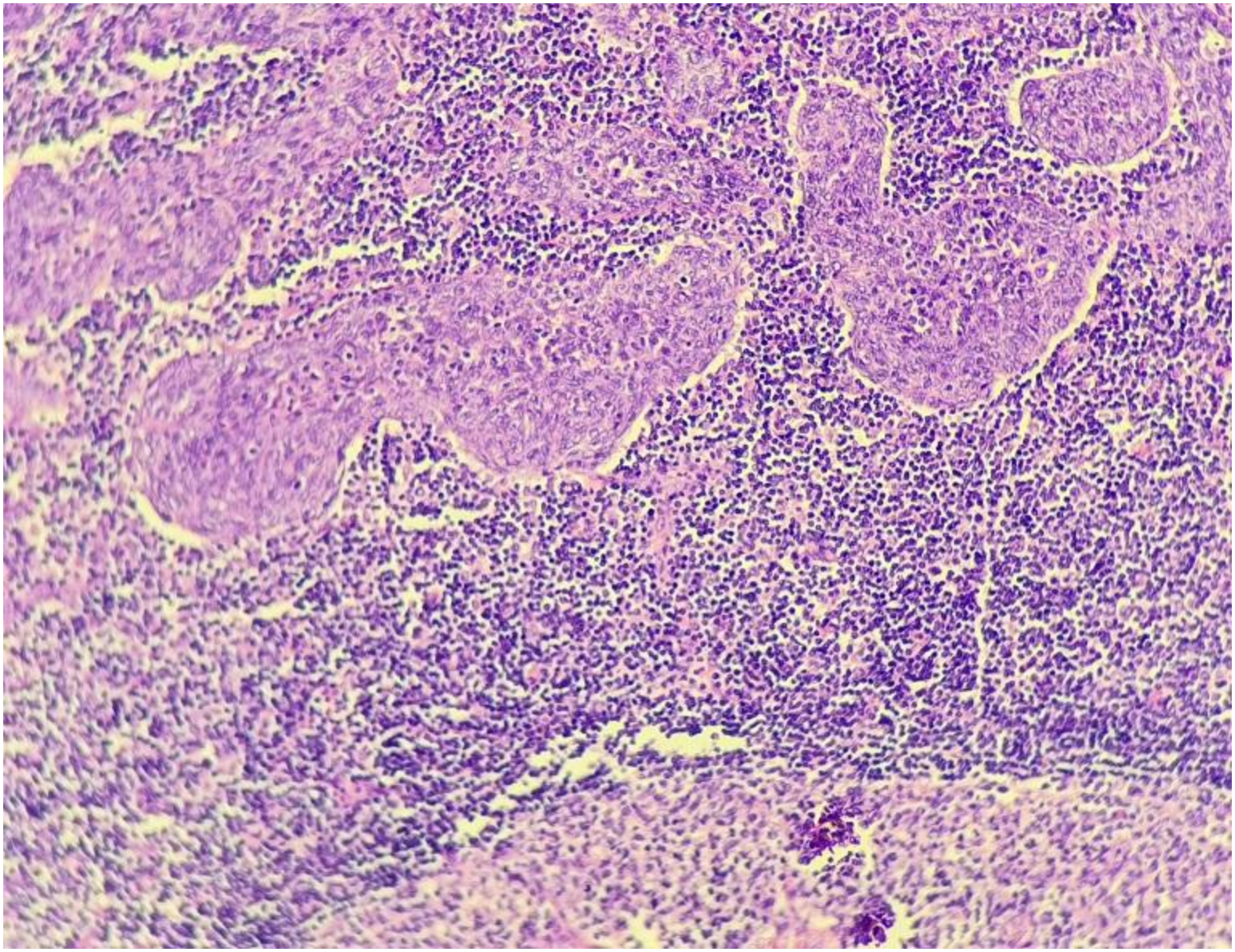

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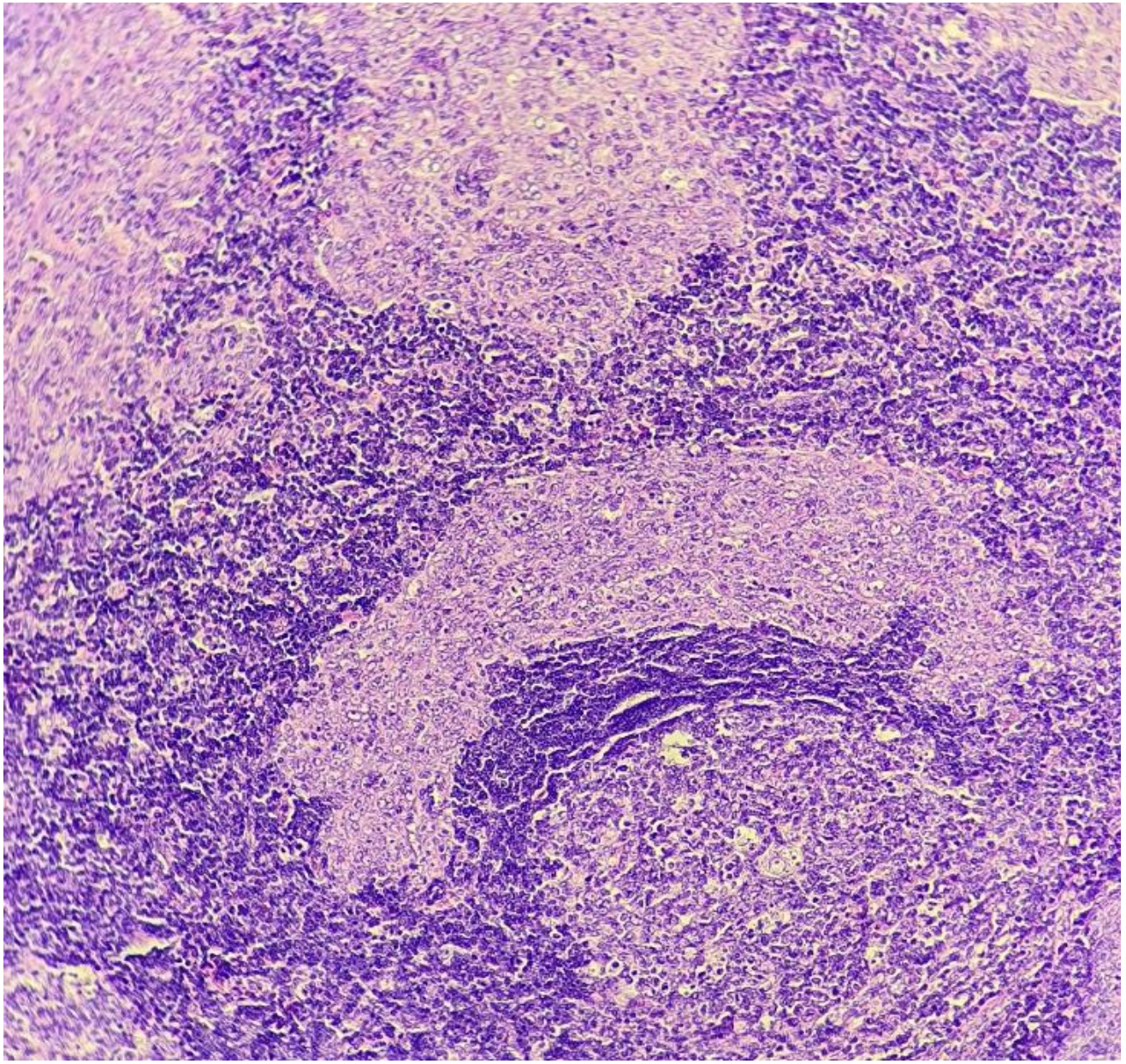

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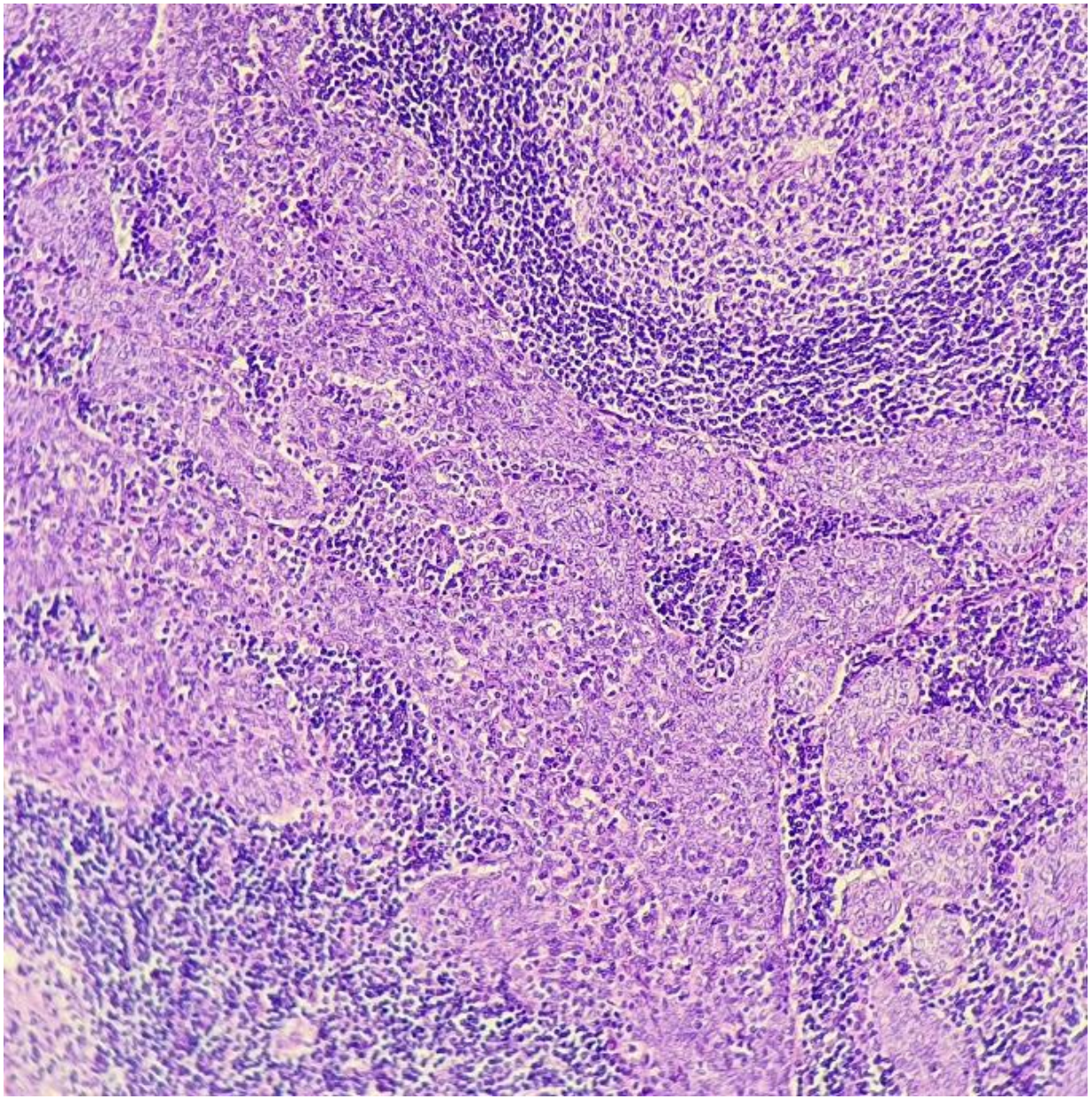


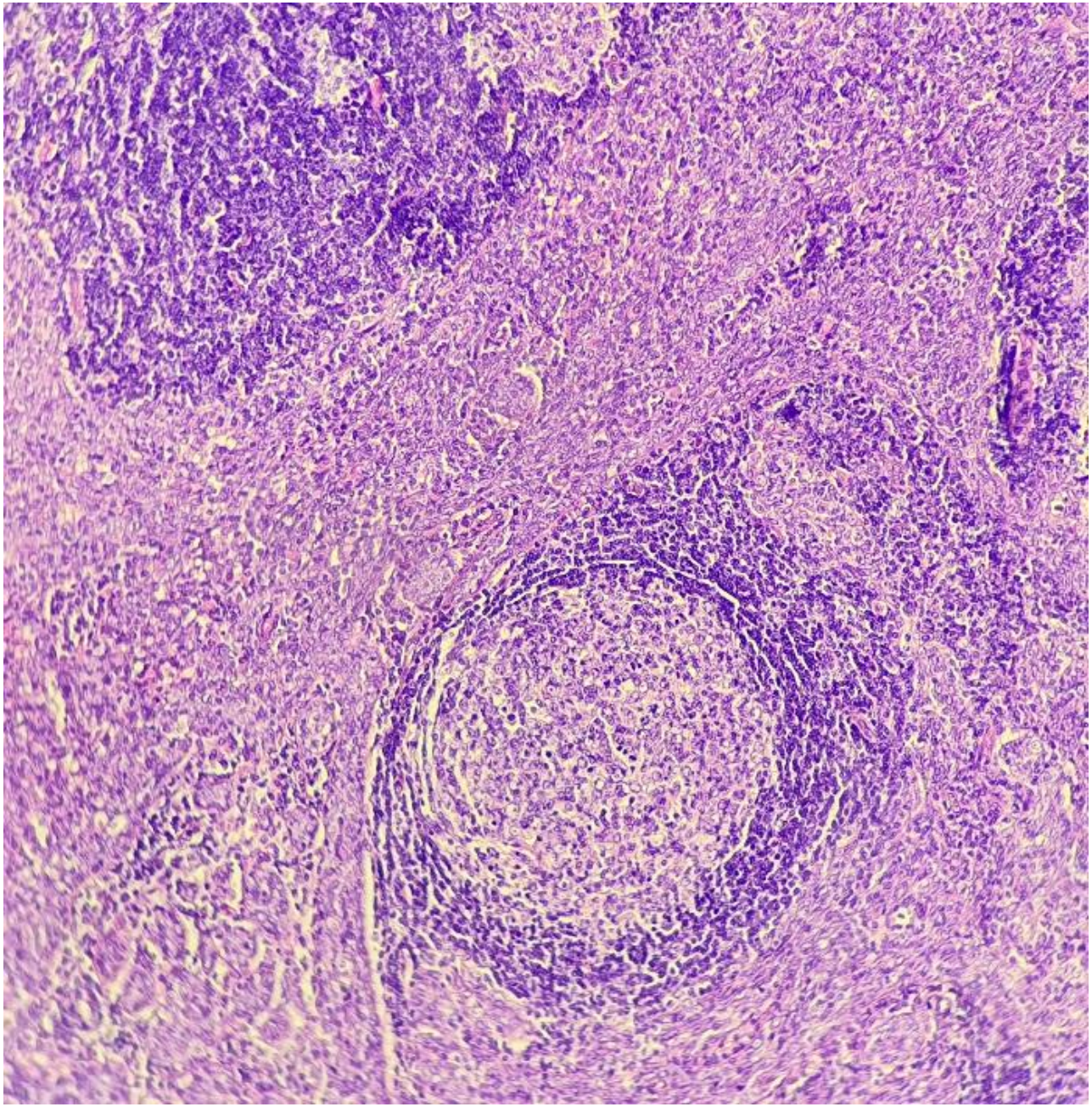
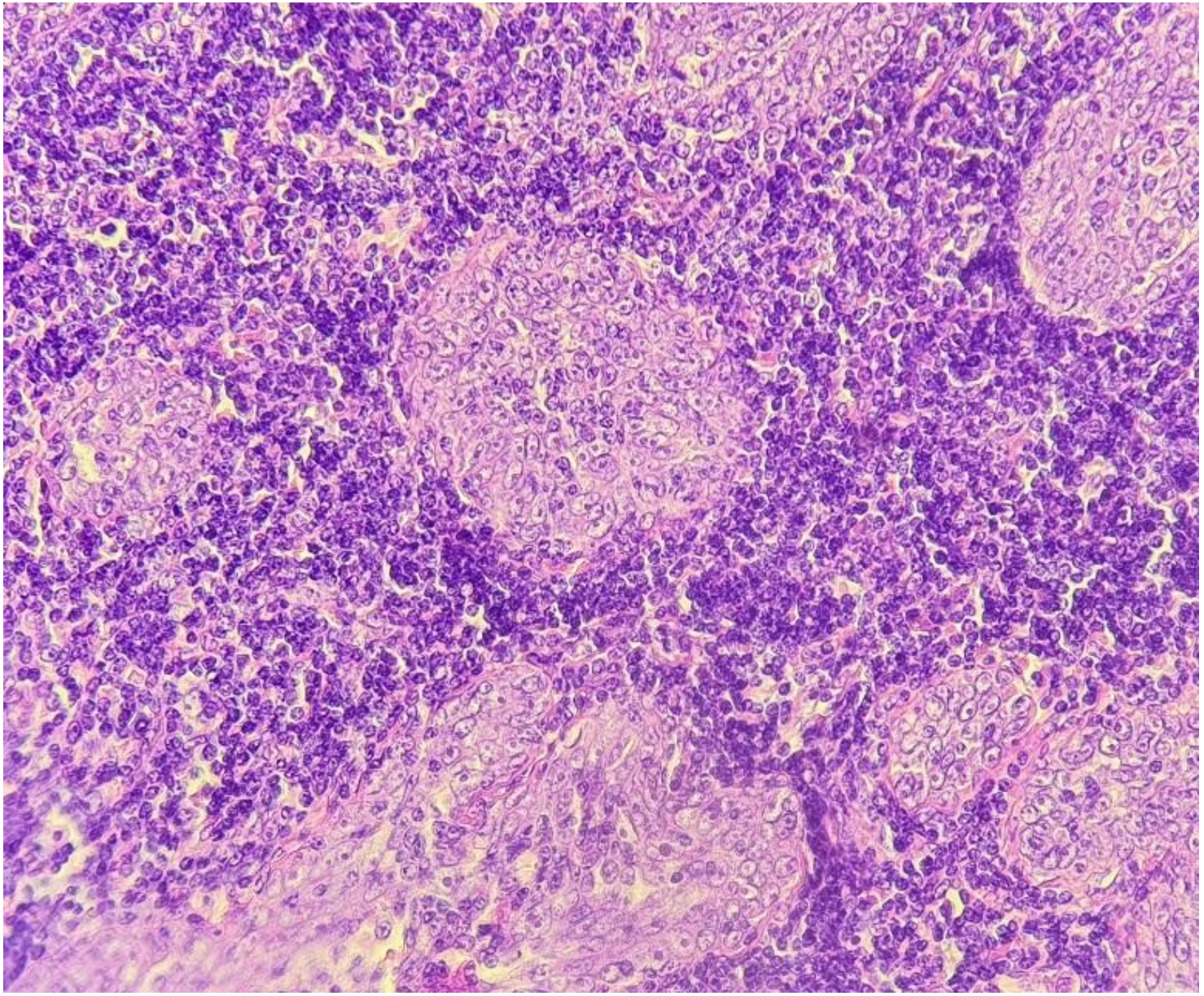


Figure.7 (X20)

Figure.8 (X40)



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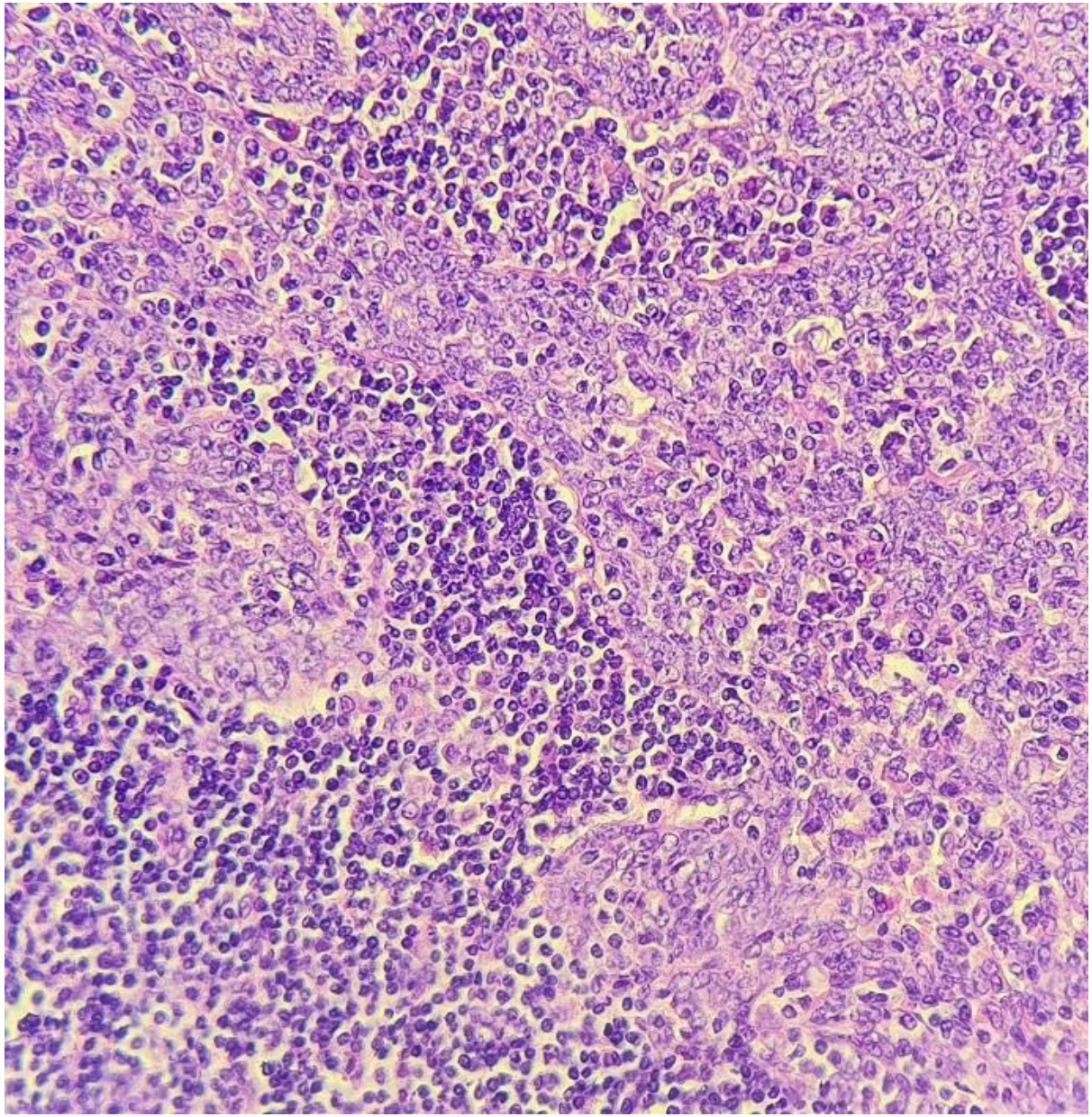

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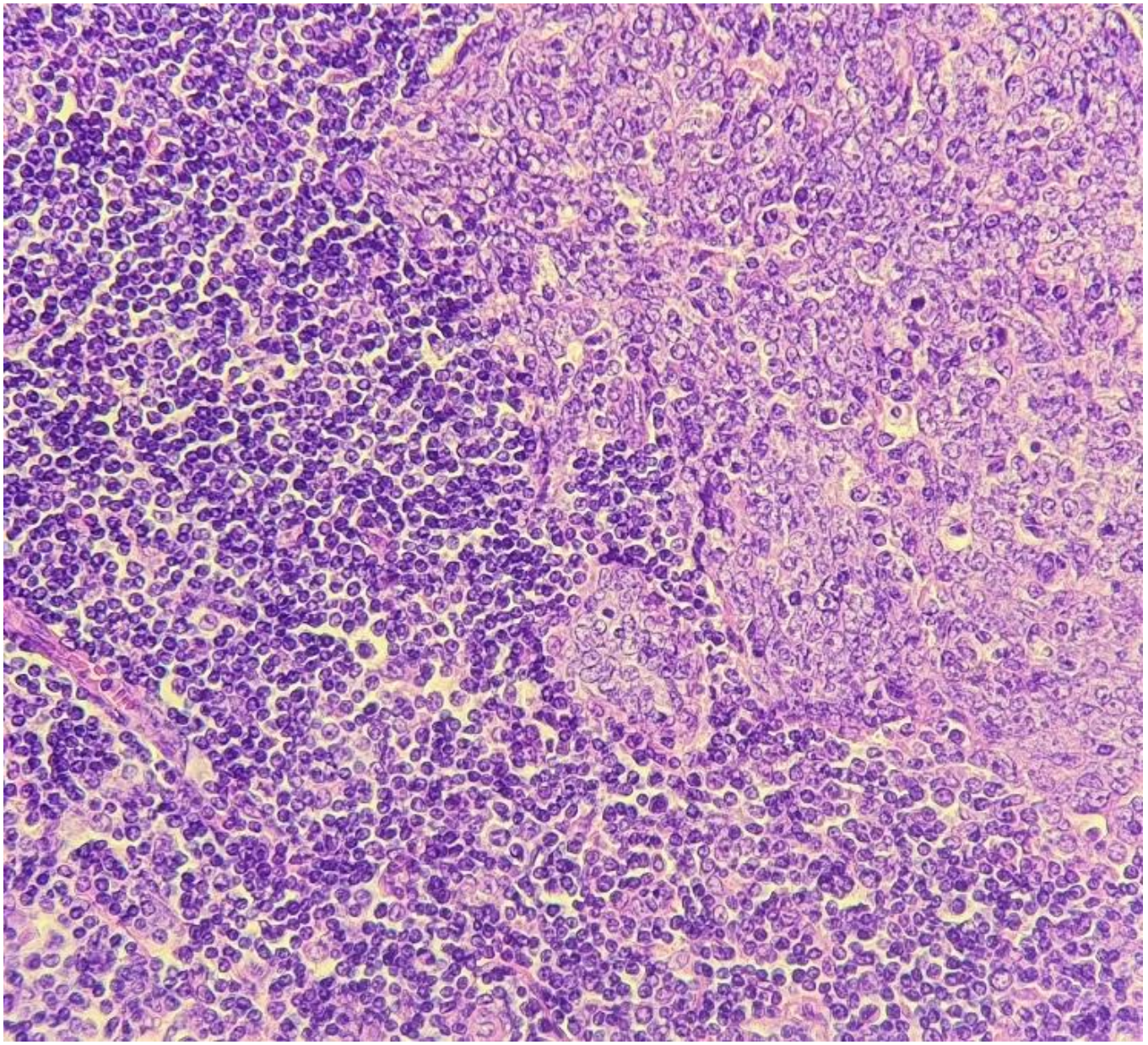
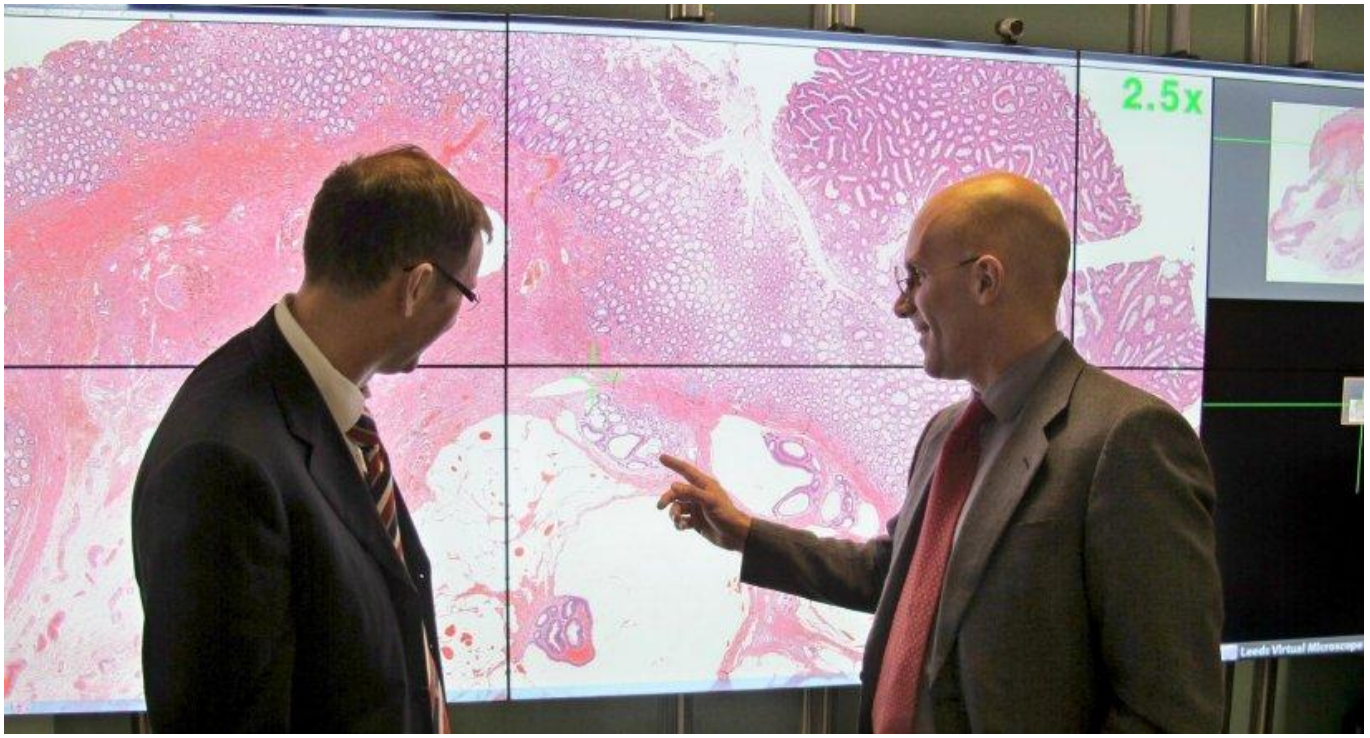




What is your Dx and DDx ?

DDx

1. Lymphoepithelial Cyst
2. Lymphoepithelial Sialadenitis (LESA)
3. Lymphoepithelial Carcinoma
4. Nasopharyngial Carcinoma
5. Metastasis
6. Lymphoma

Figure.11 (X40)

IHC Staining – **CK**

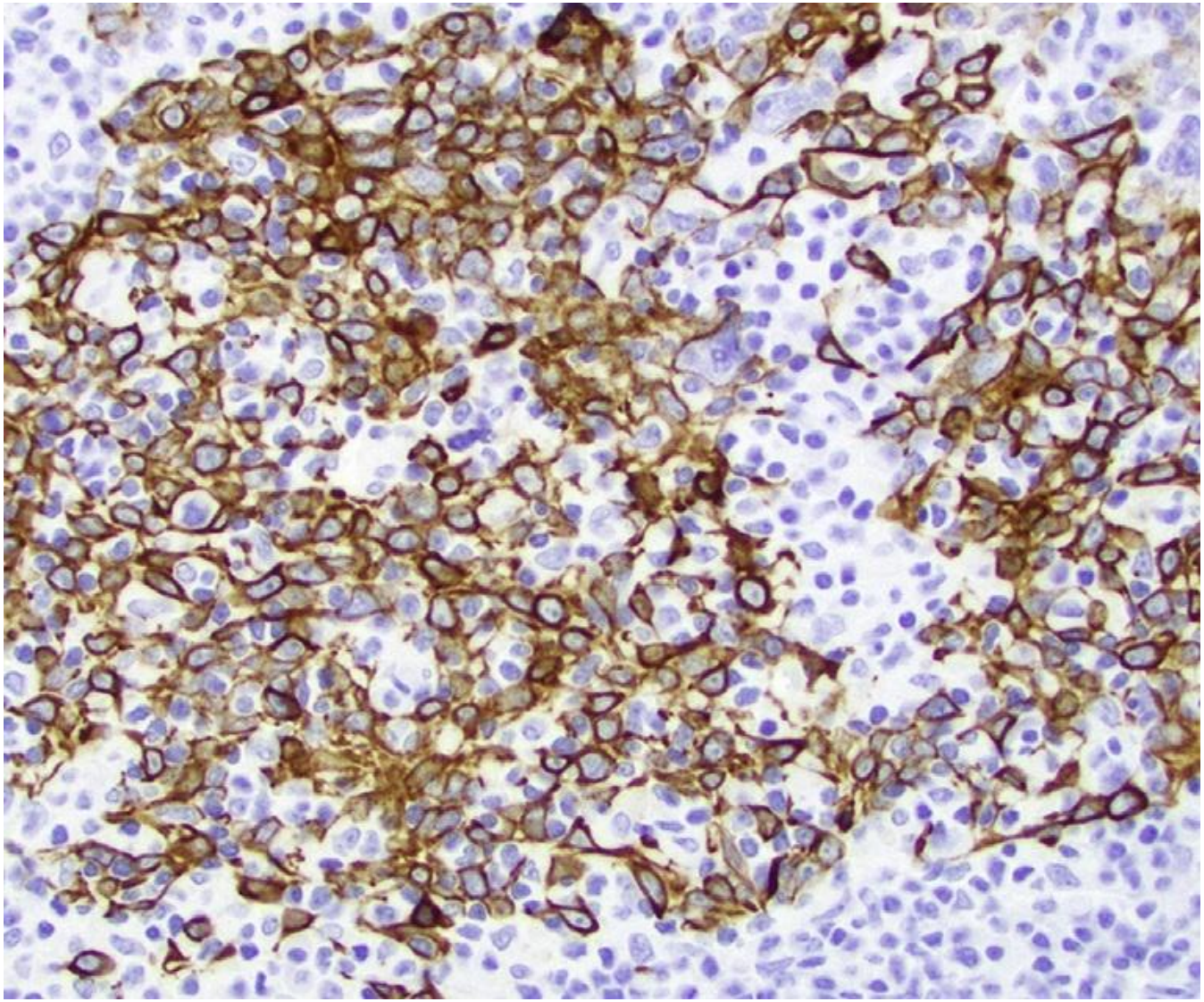


Figure.12 (X40)

IHC Staining – **CK 5/6**

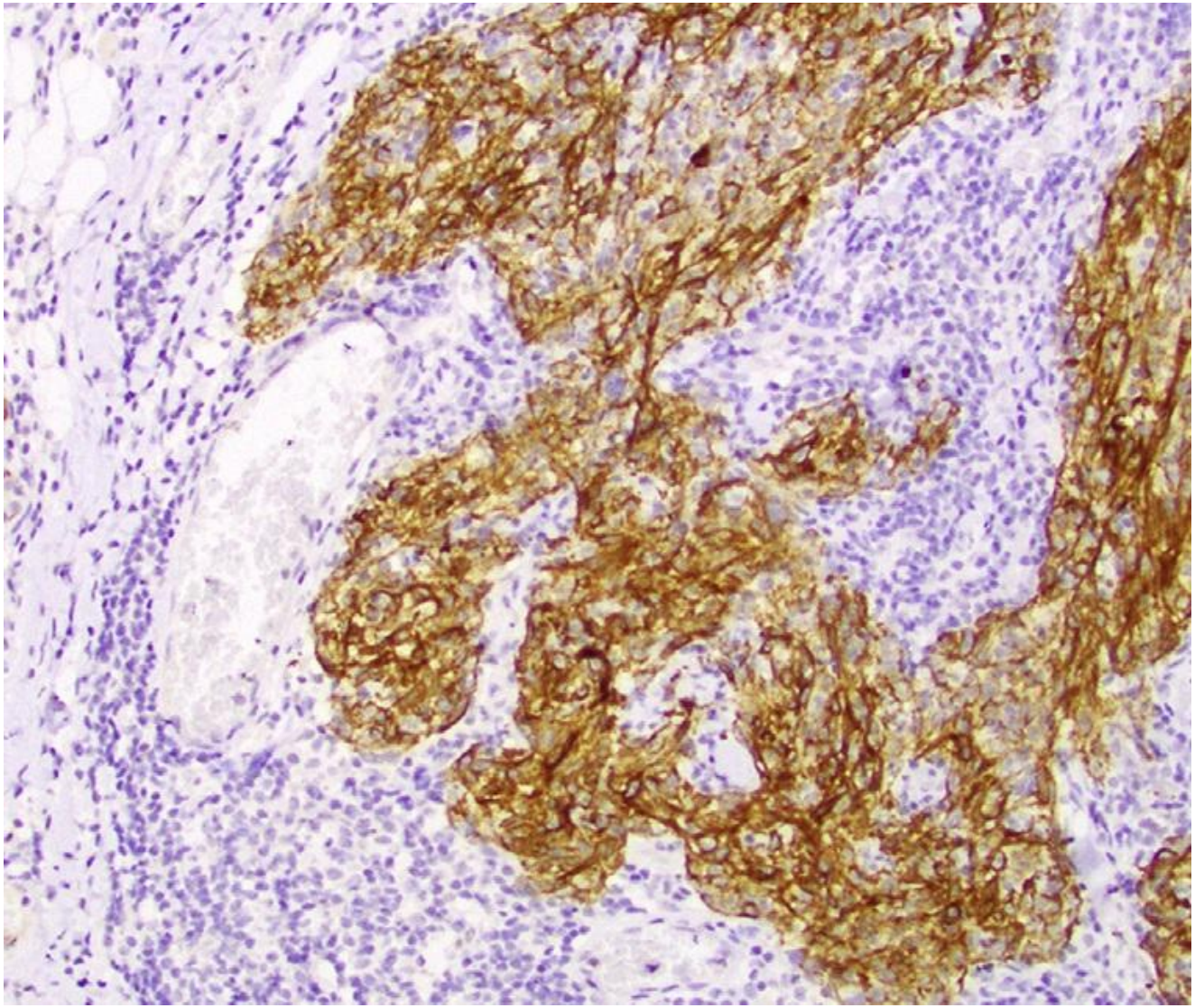
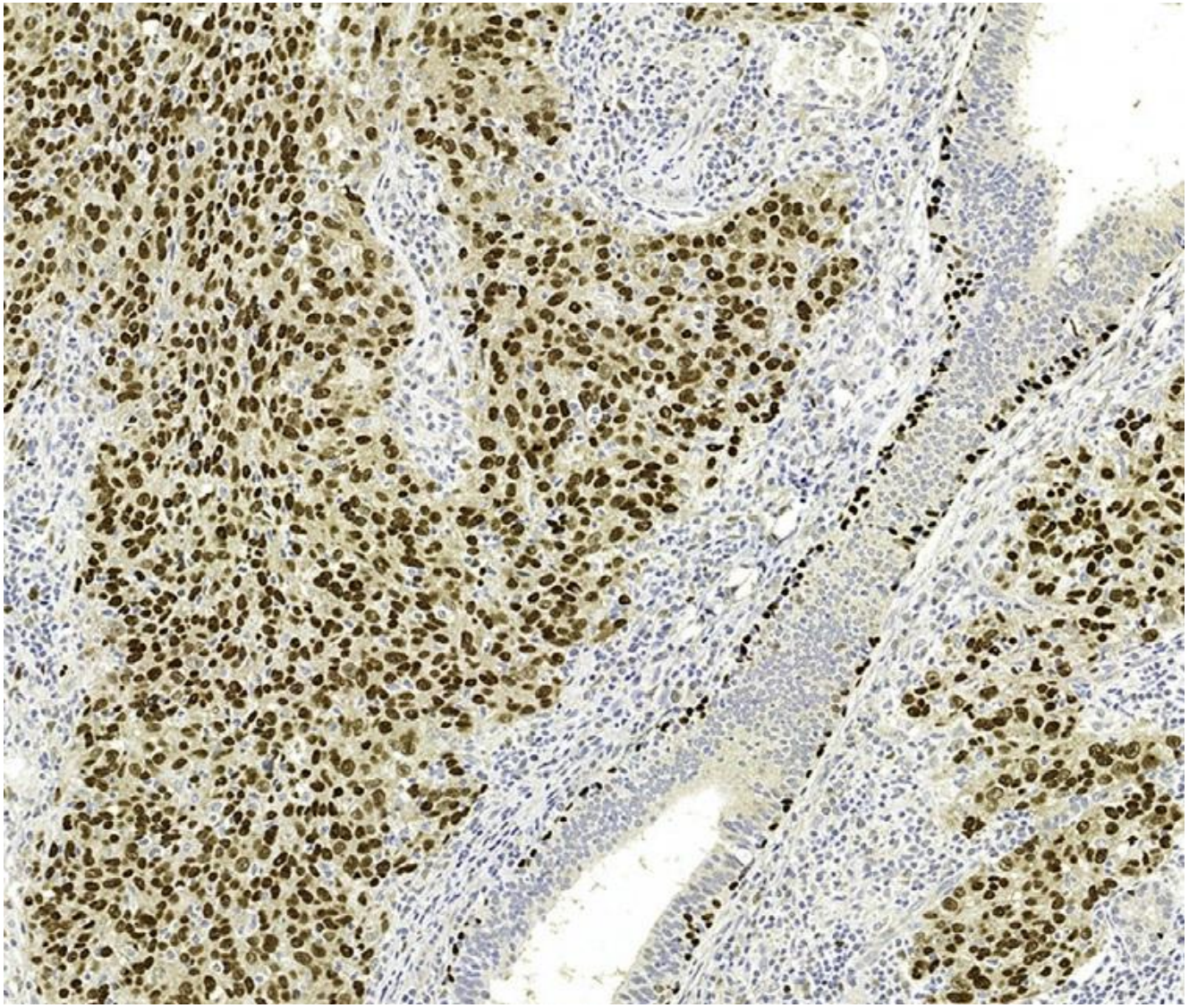


Figure.13 (X40)

IHC Staining – **P63**



What Is Your Dx ?

Dx

Lymphoepithelial Carcinoma of Salivary Glands

Lymphoepithelial Carcinoma of Salivary Glands

Review Article - 2021

Lymphoepithelial Carcinoma of Salivary Glands

Lester D.R. Thompson, MD[a,*], Rumeal D. Whaley, MD[b]

KEYWORDS

• Lymphoepithelial carcinoma • Salivary gland • Immunohistochemistry • EBER

Key points

- Lymphoepithelial carcinoma is a rare undifferentiated primary salivary carcinoma that is associated with a florid lymphoid background.
- Epstein-Barr virus association is near 100% in endemic populations (e.g., Eskimo/Inuit) while the association in nonendemic populations is not as reproducible.
- Morphology, immunohistochemical stains, and Epstein-Barr virus encoded small RNA (EBER) in situ hybridization (ISH) are insufficient to distinguish lymphoepithelial carcinoma from nasopharyngeal carcinoma, which requires clinical evaluation.
- These tumors, irrespective of race or ethnicity, may express EBER, but a negative EBER-ISH does not exclude the diagnosis.

ABSTRACT

Lymphoepithelial carcinoma of salivary glands (LECSG) is an uncommon neoplasm. This article summarizes the findings of 438 cases in a review of the literature. Concurrent lymphoepithelial lesions may suggest a primary tumor. The tumor shows a nonkeratinizing carcinoma intimately associated with a rich lymphohistiocytic infiltrate, destroying adjacent salivary gland tissue. Irrespective of race or ethnicity, the tumors usually express Epstein-Barr virus, with Epstein-Barr virus encoded small RNA (EBER) and/or latent membrane protein-1 (LMP-1), although a subset does not. There is an overall good prognosis of about 80% at 5 years.

OVERVIEW

Undifferentiated carcinoma with an associated prominent, nonneoplastic, lymphoplasmacytic cell infiltrate is now called lymphoepithelial carcinoma (LEC).[1] It was originally described by Hilderman and colleagues[2] in 1962, as the possible malignant transformation of benign lymphoepithelial lesions (BLEL). LEC has gone by a diverse nomenclature, including but not limited to undifferentiated carcinoma, anaplastic parotid carcinoma, lymphoepitheliomalike carcinoma, lymphoepithelial-like carcinoma, undifferentiated carcinoma with lymphoid stroma, malignant lymphoepithelial lesion, nonnasopharyngeal undifferentiated carcinoma, and carcinoma ex lymphoepithelial lesion.[1,3–13] The historical names are not inherently inaccurate, but LEC most accurately reflects the intimate relationship between the epithelial and the lymphoid components of this neoplasm. Undifferentiated nasopharyngeal carcinoma (NPC) is the prototypical LEC.[8,14–18] It is most commonly reported in patients from Southeast Asian and Arctic Inuit populations, as well as descendants of these ethnic groups who migrate to nonendemic

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❖ a rare undifferentiated salivary carcinoma that is associated with a florid lymphoid background

❖ uncommon primary Neoplasm

❖ The tumor shows a non-keratinizing carcinoma intimately associated with a rich lymphohistiocytic infiltrate, destroying adjacent salivary gland tissue

❖ the tumors usually express Epstein-Barr virus, with Epstein-Barr virus encoded small RNA (EBER) and/or latent membrane protein-1 (LMP-1), although a subset does not

❖ Undifferentiated carcinoma with an associated prominent, non-neoplastic lymphoplasmacytic cell infiltrate is now called lymphoepithelial carcinoma (LEC)

❖ LEC most accurately reflects the intimate relationship between the epithelial and the lymphoid components of this neoplasm

❖ Undifferentiated nasopharyngeal carcinoma (NPC) is the prototypical LEC

❖ It is most commonly reported in patients from Southeast Asian and Arctic Inuit populations, as well as descendants of these ethnic groups who migrate to nonendemic countries

❖ Nearly all of the LECs in endemic patients express Epstein-Barr virus (EBV) in the neoplastic cells, which can be confirmed by a variety of different techniques (ISH or LMP-1 reactivity)

❖ LEC of salivary gland

(LECSG) is the most common salivary gland malignancy,

mostly identified in the parotid gland (for up to 90% of all salivary gland malignancies)

❖ It is always prudent to

exclude a Nasopharyngeal primary before making

a definitive diagnosis of a salivary gland primary

given similarly affected ethnic groups

❖ Most of the EBV-associated carcinomas are of the lymphoepithelial type

❖ LMP1 in human epithelial cells has been shown to deregulate epithelial growth and inhibit differentiation, with the cells showing loss of contact inhibition, spindling, and a tendency to proliferate

❖ There is an increased risk of lymphoepithelial carcinomas (salivary or nasopharynx) in patients with human immunodeficiency virus (HIV) infection who progress to acquired immunodeficiency syndrome (AIDS)

❖ However, HIV-associated salivary gland disease usually encompasses lymphoid hyperplasia, follicular involution, lymphoepithelial cysts (usually bilateral), and lymphoepithelial lesions

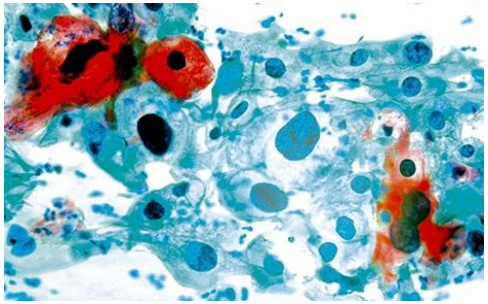
CLINICAL FINDINGS

- nonspecific symptoms, including swelling or a mass in the salivary gland
- Pain (approximately 5%) and nerve findings (approximately 2.5%) uncommonly recognized
- In advanced cases, skin and soft tissue fixation may be seen
- Symptoms are present over an exceptionally broad time frame, although most patients report slightly less than 2 years of symptoms
- Almost all tumors affect the major glands, with the parotid (76%) and submandibular glands (17%)
- Occasionally, intratumoral necrosis or cystic change is noted

LABORATORY STUDIES

Most patients show some serologic evidence of previous infection with EBV

EBV viral capsid antigen (VCA) to immunoglobulin (Ig), IgM, and/or IgG, EBV nuclear antigen (EBNA) IgG, and early antigen (EA)-IgG antibodies are seen in patients with a past infection, they are not always seen in patients with LEC



CYTOLOGY

Smears show single to clustered large polygonal and spindled cells in syncytial sheets

The cells have limited to moderate amounts of cytoplasm and have a high-grade, undifferentiated appearance

The nuclei are vesicular with prominent nucleoli

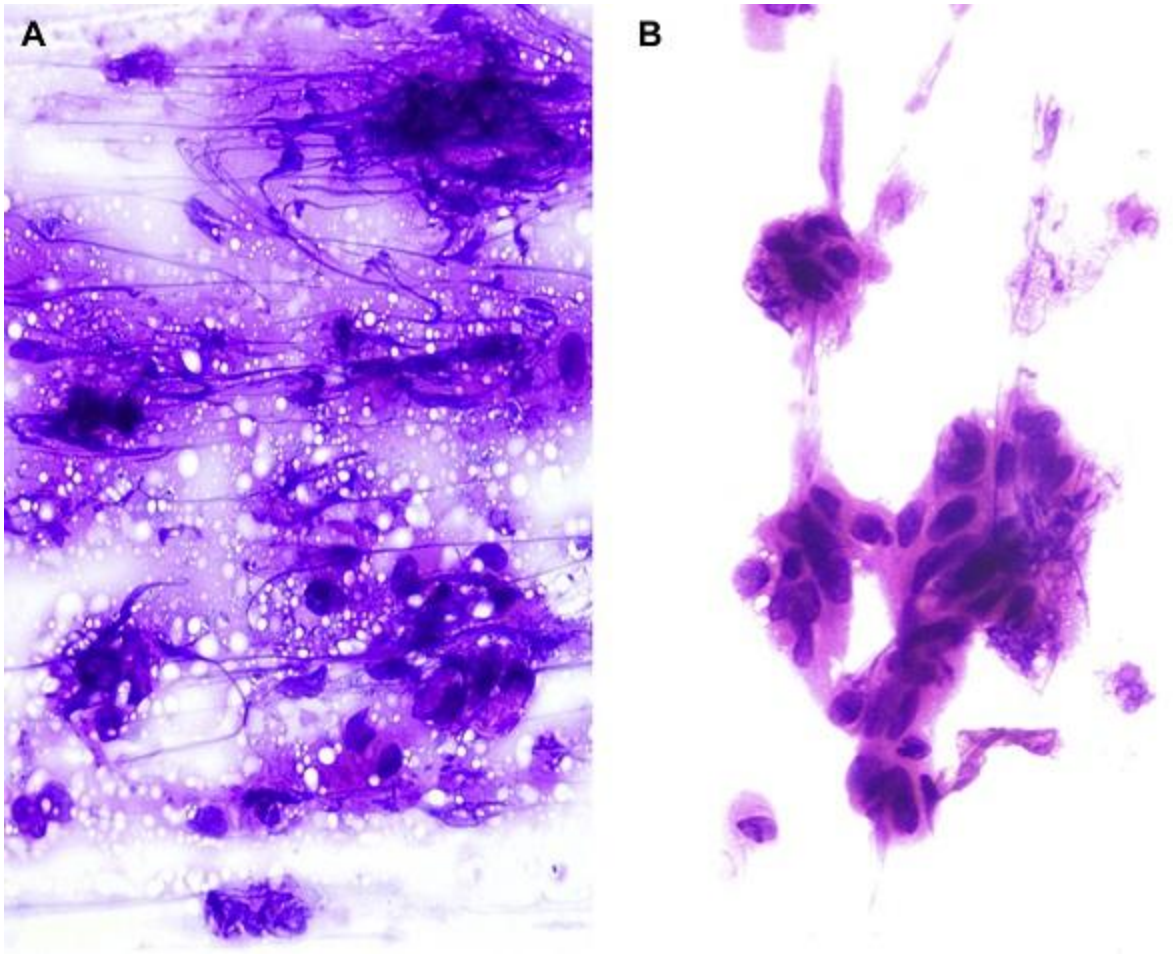
Mitotic activity is brisk

Most have a prominent mixed lymphoid population that may mask the isolated epithelial elements, resulting in a misdiagnosis

Metastatic nasopharyngeal carcinoma to an intra–salivary gland lymph node may also be a consideration

a dense lymphoplasmacytic infiltrate may mimic an intra–salivary gland lymph node, and, depending on the type of population present, may suggest a lymphoma

If the epithelial cells are not atypical, lymphoepithelial sialadenitis or lymphoepithelial cyst may also be considered



(A) The smears are cellular, showing prominent lymphoid matrix tangles in the background with large, polygonal, syncytial epithelial cells

(B) There is a cluster of atypicalepithelial cells with a high nuclear to cytoplasmic ratio

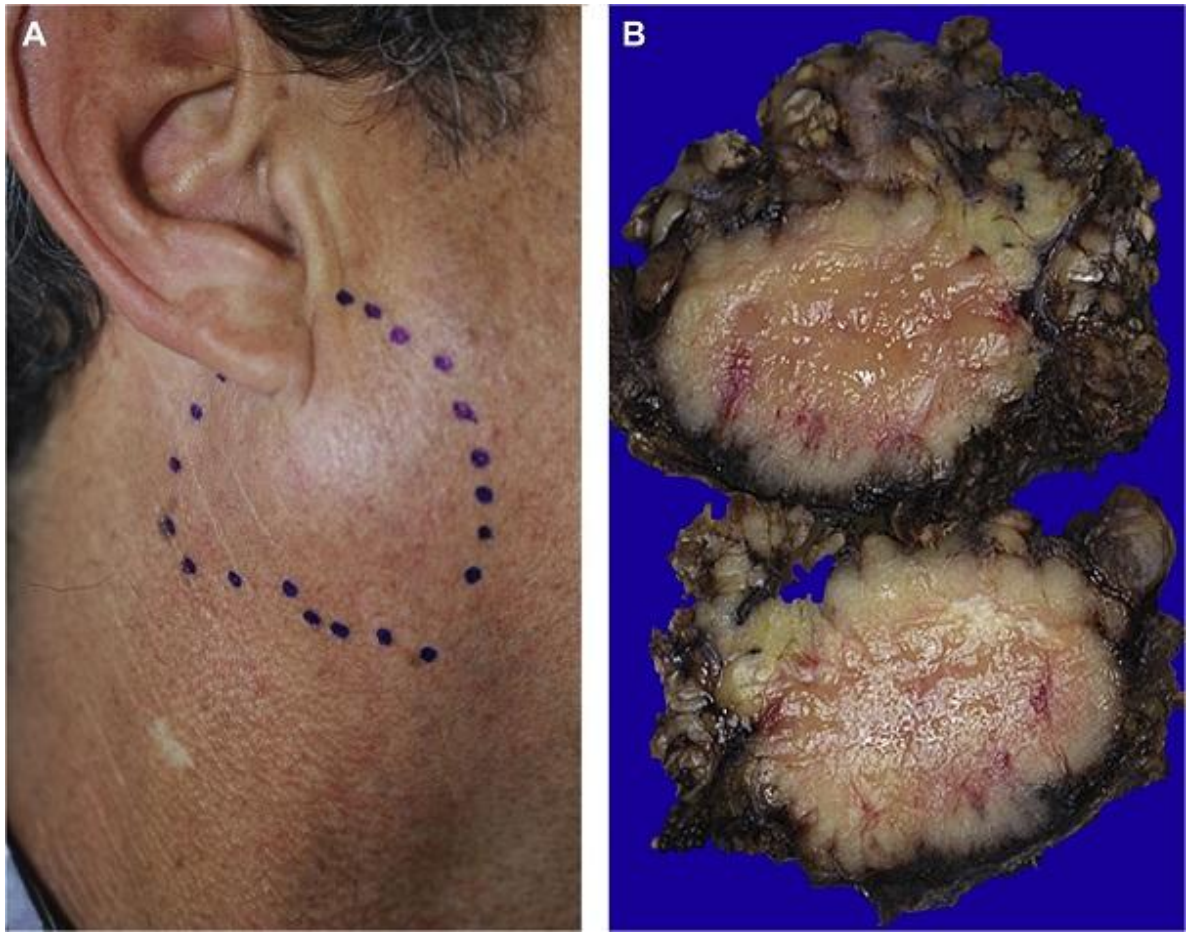
Many primary salivary gland neoplasms have a prominent lymphoid stroma associated with them, such as mucoepidermoid carcinoma and acinic cell carcinoma, and these tumors should not be confused with LEC

MACROSCOPIC FEATURES

LEC grossly are typically circumscribed but not encapsulated, with a lobulated, firm, tanwhite cut appearance

Other cases show an infiltrative appearance into the adjacent salivary glands, fat, skeletal muscle, and skin.

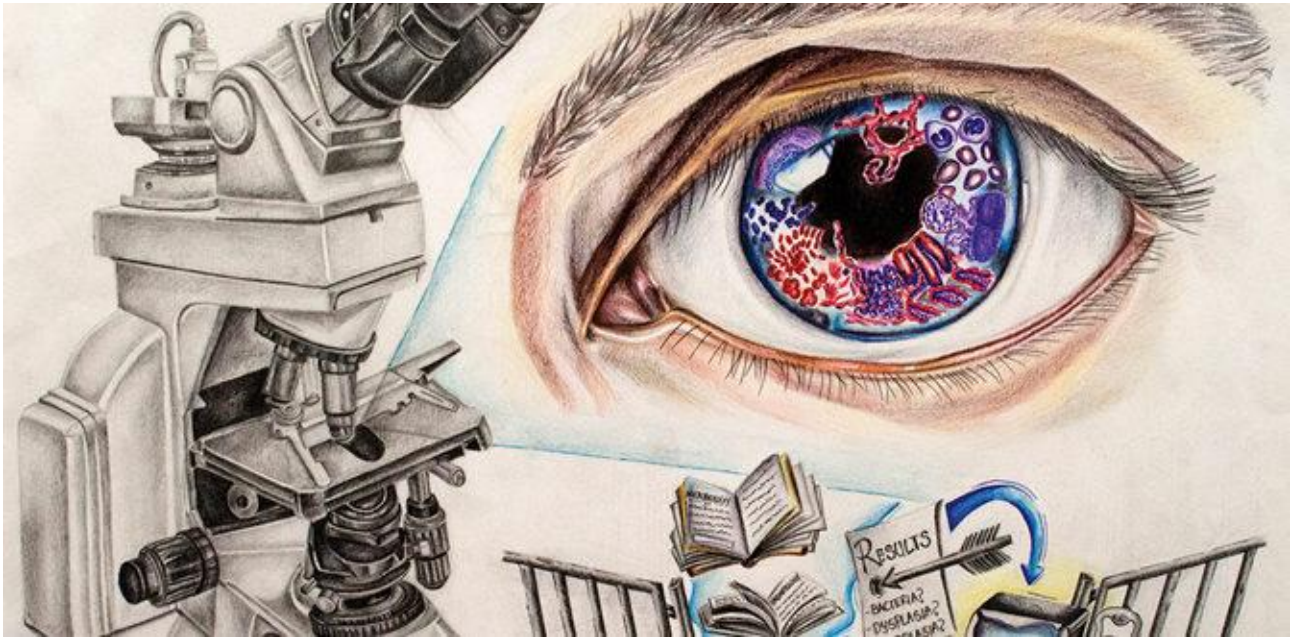
The tumors range up to 15 cm with most greater than 2 cm.



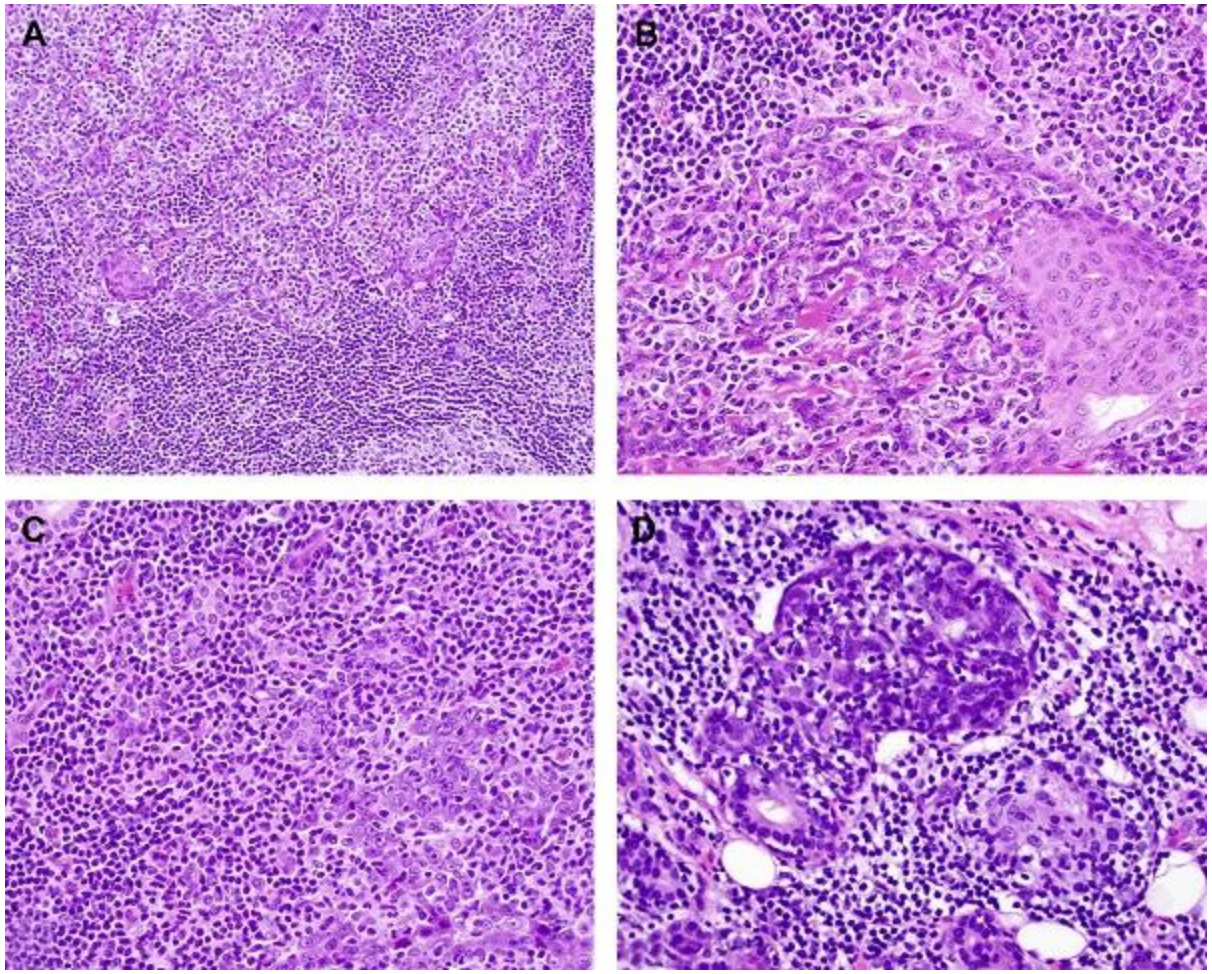
(A) Clinical examination showed a parotid gland swelling without skin erythema in a patient without a nasopharyngeal mass.

(B) There is a fish-flesh pale cut surface of the parotid gland, showing a mass replacing the entire gland

Microscopic (histologic) description



malignant transformation from lymphoepithelial sialadenitis (LESA) or myoepithelial sialadenitis (MESA).



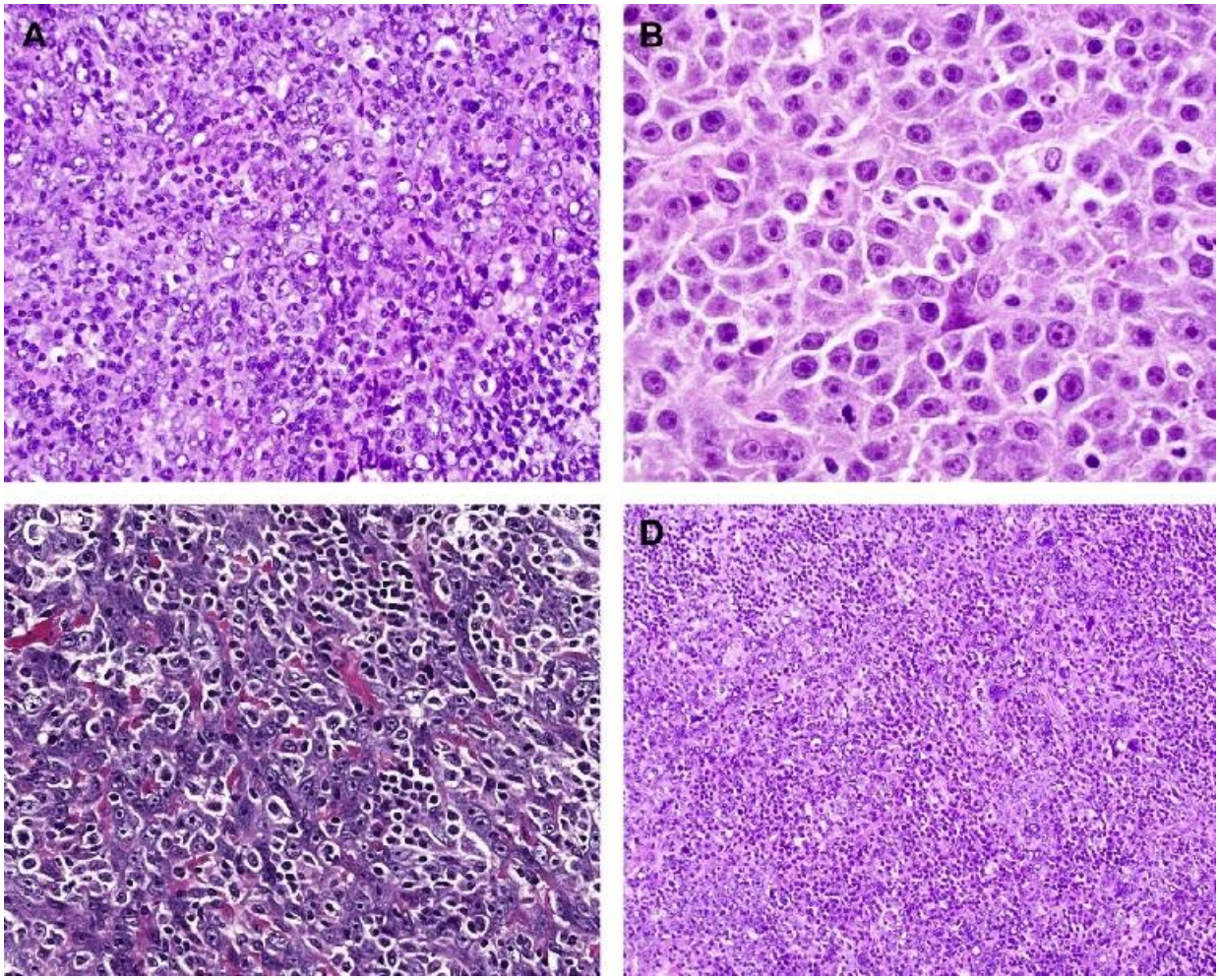
LESA adjacent to LEC

(A) Monocytoid lymphocytes are seen associated with a terminal acinar duct lobule.

(B) Squamous metaplasia intimately associated with lymphoid elements.

(C) Epithelial proliferation associatedwith lymphocytes, but lacking cytologic atypia.

(D) A terminal duct with inflammatory cells. Histiocytes are noted in the lower right corner.



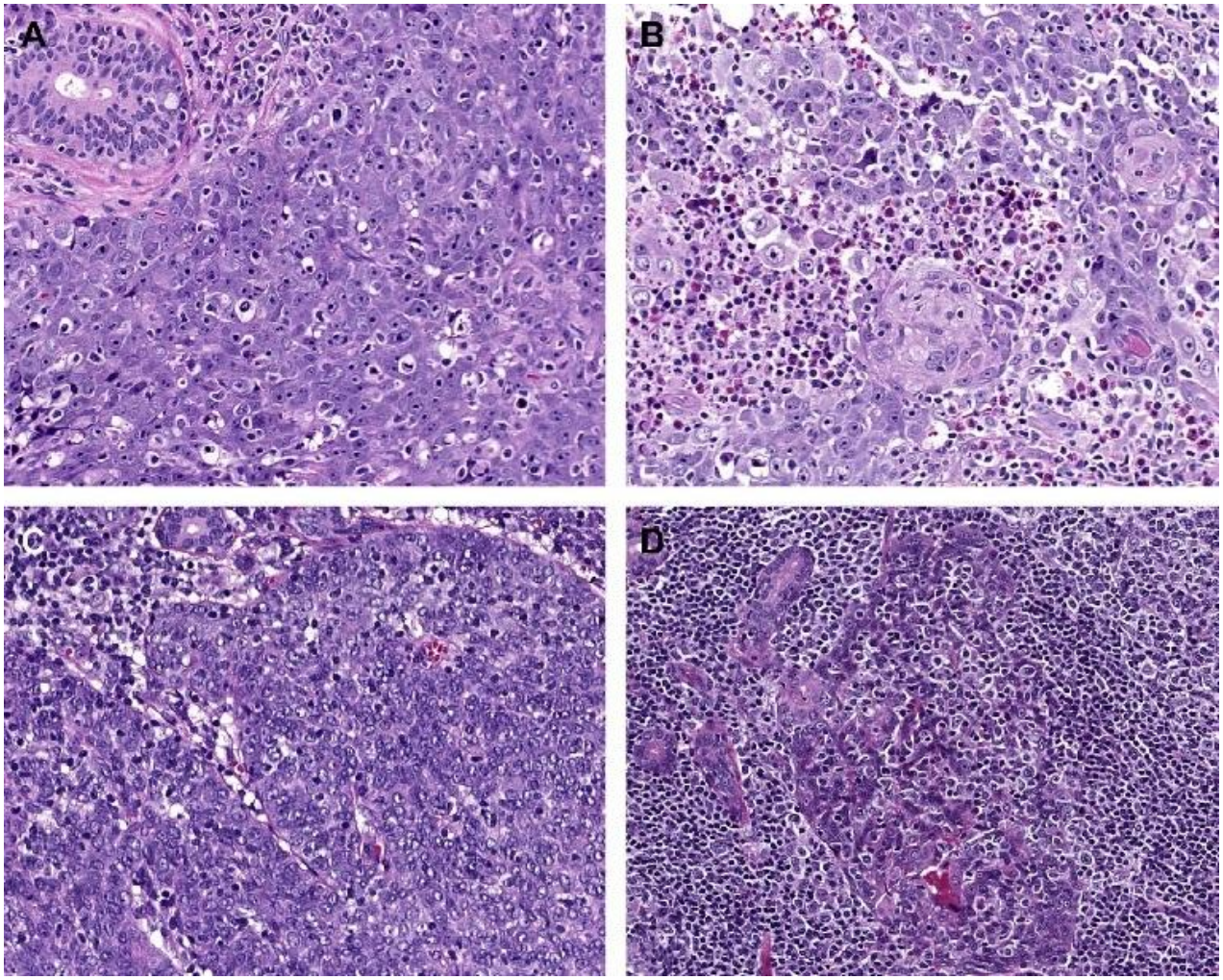
Cellular features of LEC

(A) Syncytium of large, polygonal neoplastic cells with vesicular chromatin

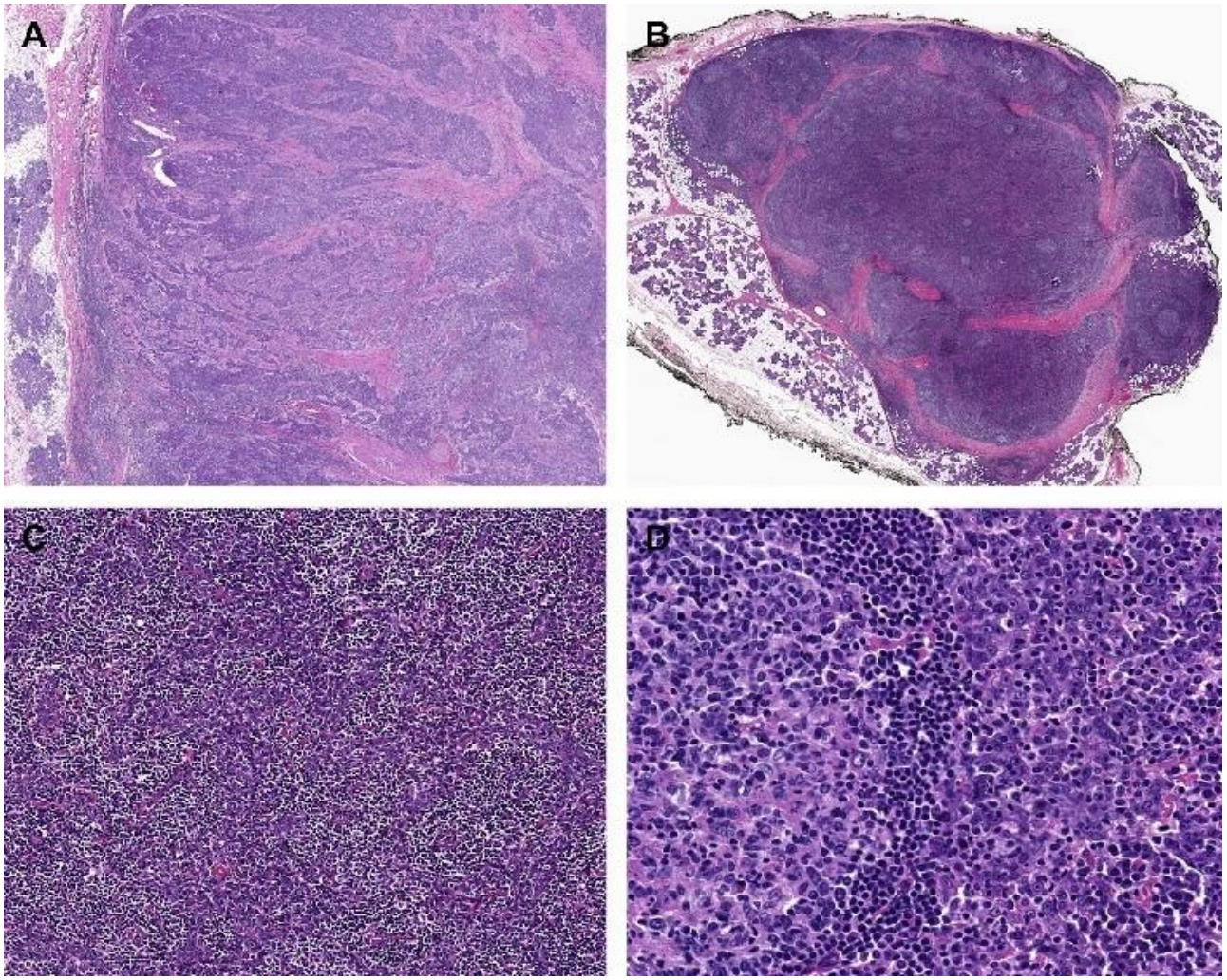
(B)Large, prominent, hypereosinophilic nucleoli

(C) Cytoplasmic extensions surround vesicular nuclei

(D) Smudgeand focal tumor giant cells within a sea of neoplastic cells



(A) Cellular pleomorphism is easily noted, with a benign duct for comparison
(B) Keratin pearl formation is seen, along with an acute inflammatory infiltrate
(C) Adjacent ducts are unremarkable, with tumor arranged in a sheet
(D) There is subtle atypia in this central island of tumor, adjacent to a duct and a germinal center



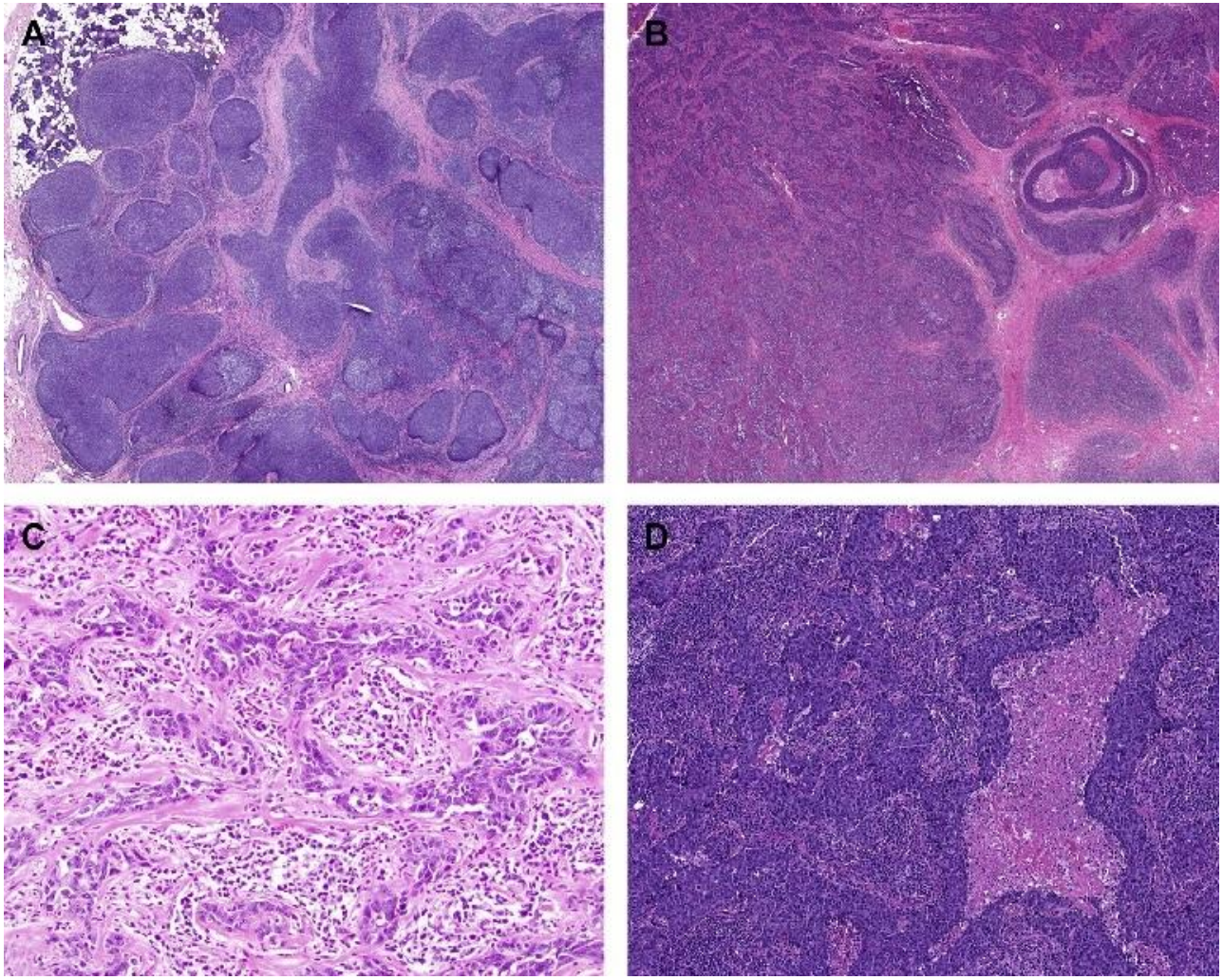
Classic Schmincke pattern

(A) Circumscribed tumor, arranged in sheets with separating fibrosis

(B) Multinodulartumor with prominent lymphoid component, showing numerous germinal centers

(C) Subtle epithelial component intermixed with the lymphoid cells

(D) A germinal center (left) as a point of comparison for the neoplastic cells (right)



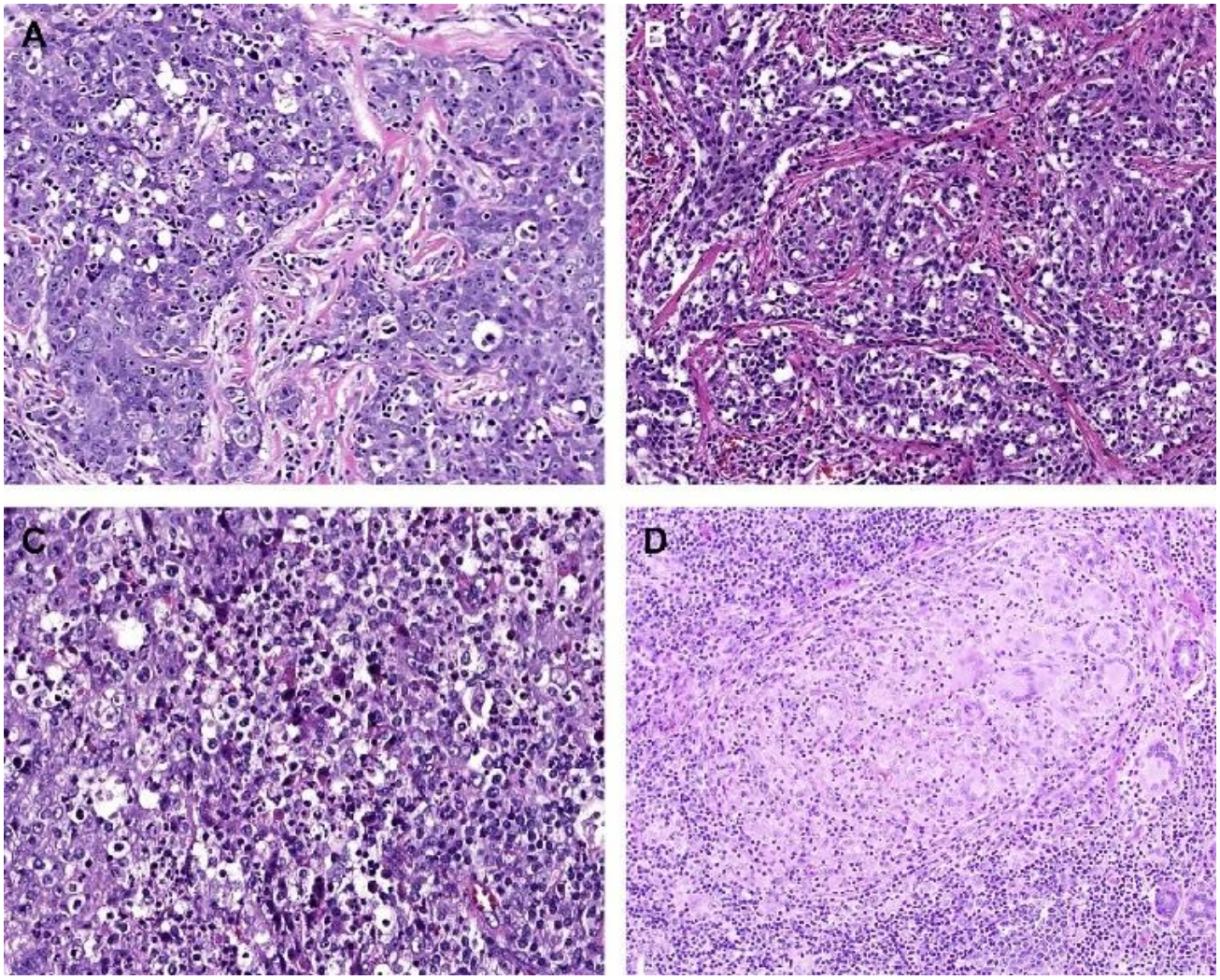
Classic Regaud pattern

(A) There are large nests and lobules of tumor

(B) Large cell nests separated by heavy fibrosis, creating a jigsaw appearance

(C) High-power view of a jigsaw pattern with a more prominentstroma

(D) Central area of comedonecrosis surrounded by ribbons of neoplastic cells



(A) Starry-sky appearance is composed of both lymphocytes and epithelial cells
(B) Numerous histiocytic cells create a starry-sky appearance
(C) Neoplastic cells adjacent to areas of acute inflammation and histiocytes
(D) Multinucleated giant cells with histiocytes are present adjacent to the tumor

TREATMENT AND PROGNOSIS

The optimal management of LEC of the major salivary glands is complete excision with clear surgical margins followed by adjuvant radiotherapy to the tumor bed and neck

The tumor category has a better prognosis than other undifferentiated carcinomas of the salivary gland (such as neuroendocrine carcinoma, NUT carcinoma, metastatic SCC)


این نکته رمز اگر بدانی، دانی
هر چیز که در جستن آنی، آنی
سعود قادری