

**KIKUCHI FUJIMOTO DISEASE
AND DISCOID LUPUS ERYTHEMATOSUS OVERLAP IN A
YOUNG FEMALE – AN UNUSUAL ASSOCIATION**

PATHOLOGY DEPARTMENT

Presented by

NAVYA VINOD

Under guidance of

Dr FASEELA K K



INTRODUCTION

- Kikuchi-Fujimoto disease [KD] is a rare disease with unknown aetiology
- affects women less than 40 years of age
- presentation : persistent low-grade fever and enlargement of the cervical lymph nodes.[1]
- There is a well-established link between systemic lupus erythematosus (SLE) and KD; patients with KD frequently have a previous history of SLE or subsequently develop SLE [2].

INTRODUCTION

- Discoid lupus erythematosus (DLE) : autoimmune connective-tissue disease that represents a subset of conditions on the cutaneous lupus spectrum.
- lesions characterized by disc-shaped plaques on photo-exposed skin with inflammatory hyperpigmentation and adherent scales. [3]
- Less commonly, DLE has also been reported to occur both before and after a KD diagnosis. [4,5,6].
- Here, I am presenting a case of a patient with such an association.

CASE PRESENTATION

HISTORY

- A 23-year-old female presented with multiple patches of scalp hair loss in the past 3 months
- associated with pain and pruritus
- Subsequently she noticed a swelling in left upper part of neck for the past 1 month
- insidious in onset , initially small , gradually progressed to size of 2×1 cm
- Associated with pain

- Evening rise of temperature for past 1 month.
- Associated with loss of appetite
- No h/o chronic cough, haemoptysis , weight loss, fatigue
- No h/o any pressure effects

PAST HISTORY

- No h/o DM, HTN, BA, drug allergy , dyslipidaemia, thyroid dysfunction
- No history of previous surgeries

SCALP

- Multiple ill defined areas of hair loss , size ranging from 1x 1cm to 2x 2 cm.
- hyperpigmented plaque with surrounding dusky erythema
- surface shows scaling
- Carpet tack sign positive
- Regrowing coiled hair present



EXAMINATION FINDINGS

- Firm , tender , mobile lymph nodes palpable in left level II cervical region, bilateral axilla and right inguinal region.
- NECK: measuring 2×1cm
- AXILLA: measuring 1×1 cm
- INGUINAL REGION: measuring 1×1 cm

WORK UP

- 4 mm Scalp punch biopsy taken

MICROSCOPY

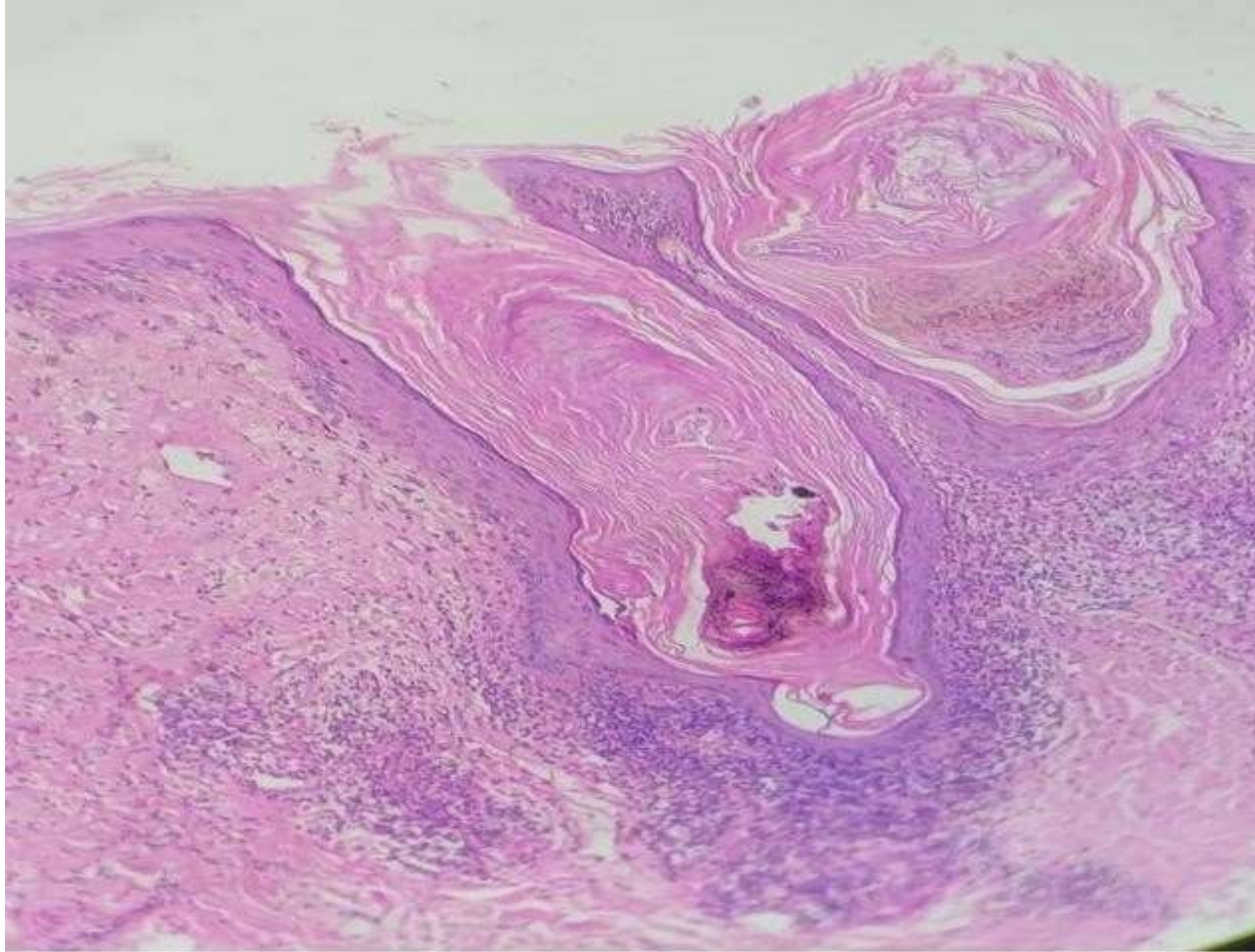


Figure 1 : Scanner power view of H & E stained sample of skin :

- Orthokeratotic hyperkeratosis with follicular plugging
- Epidermal thinning, flattened rete ridges
- Focal basal cell vacuolar degeneration
- Basement membrane thickening
- Dense interstitial perivascular and perifollicular lymphocytic infiltrate

MICROSCOPY

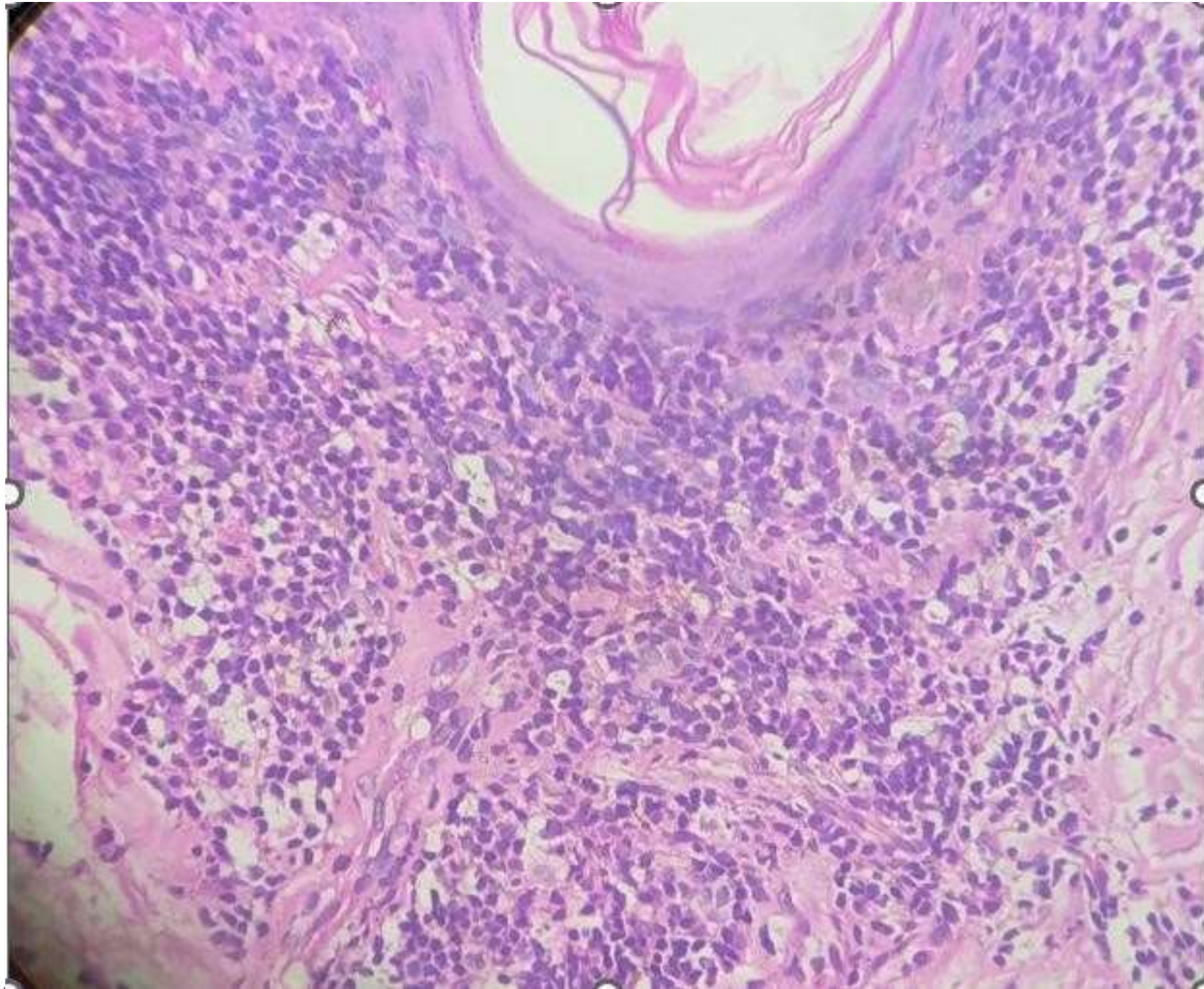


Figure 2: high power view of H & E stained sample of skin

- Squamatization of basal keratinocytes.
- Occasional Civatte bodies
- Dense lichenoid type of inflammatory infiltrate forming interface dermatitis

HISTOPATHOLOGY REPORT

With these histomorphological findings and correlating with clinical features
Diagnosis was consistent with **Discoid Lupus Erythematosus**.

EXCISION BIOPSY OF CERVICAL LYMPH NODE

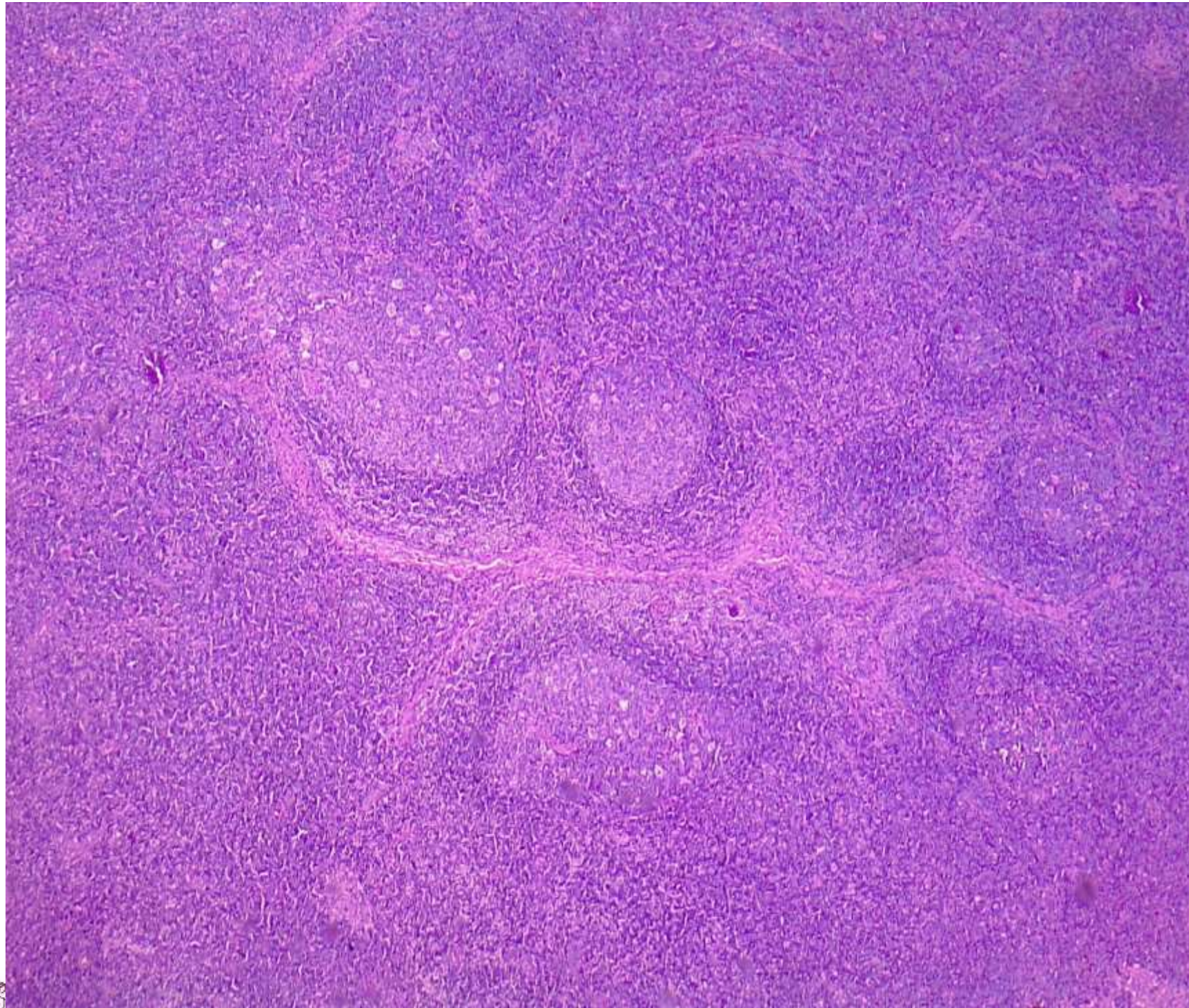


Figure 3: Scanner view of H & E-stained section of cervical lymph node biopsy

- Partial effacement of lymph node architecture.
- few residual follicles having reactive germinal centres

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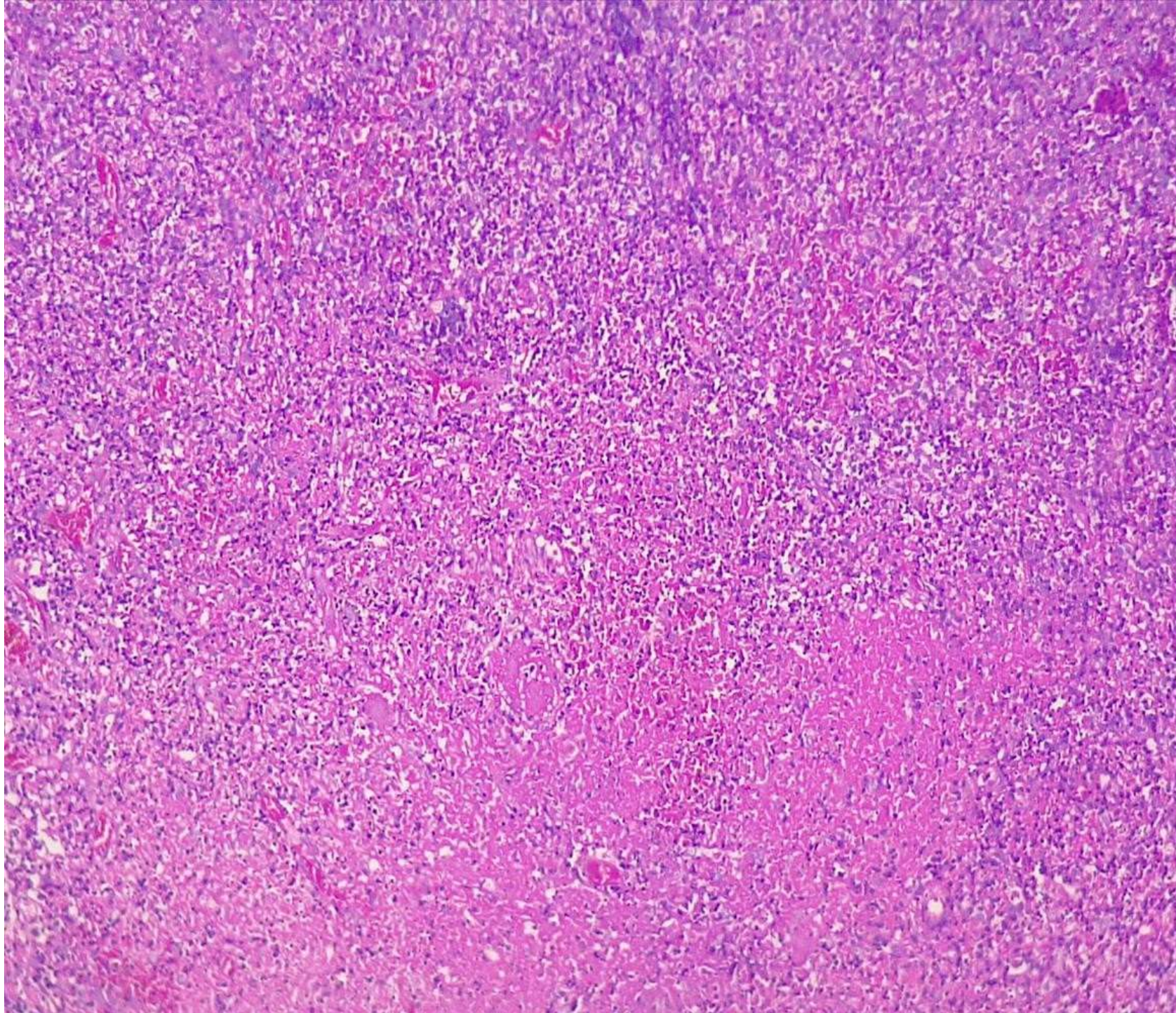


Figure 3: low power view of H & E-stained section of cervical lymph node biopsy

- Patchy areas of necrosis
- consisting of brightly eosinophilic fibrinoid deposits
- Areas of haemorrhage

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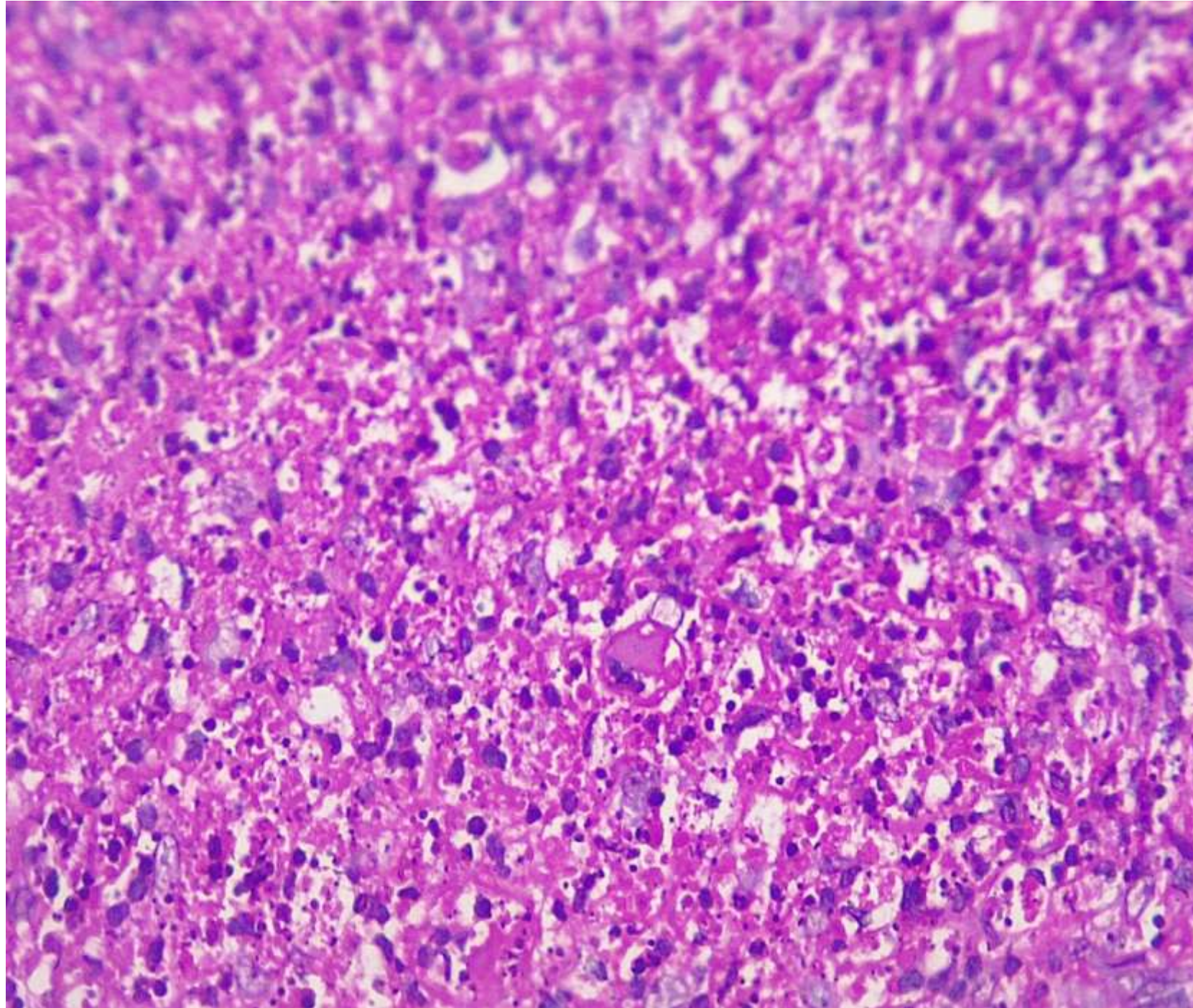


Figure 5 : High power view of H & E stained section of cervical lymph node biopsy

- large aggregates of pale staining histiocytes
- Numerous karyorrhectic debris
- Fibrin deposits

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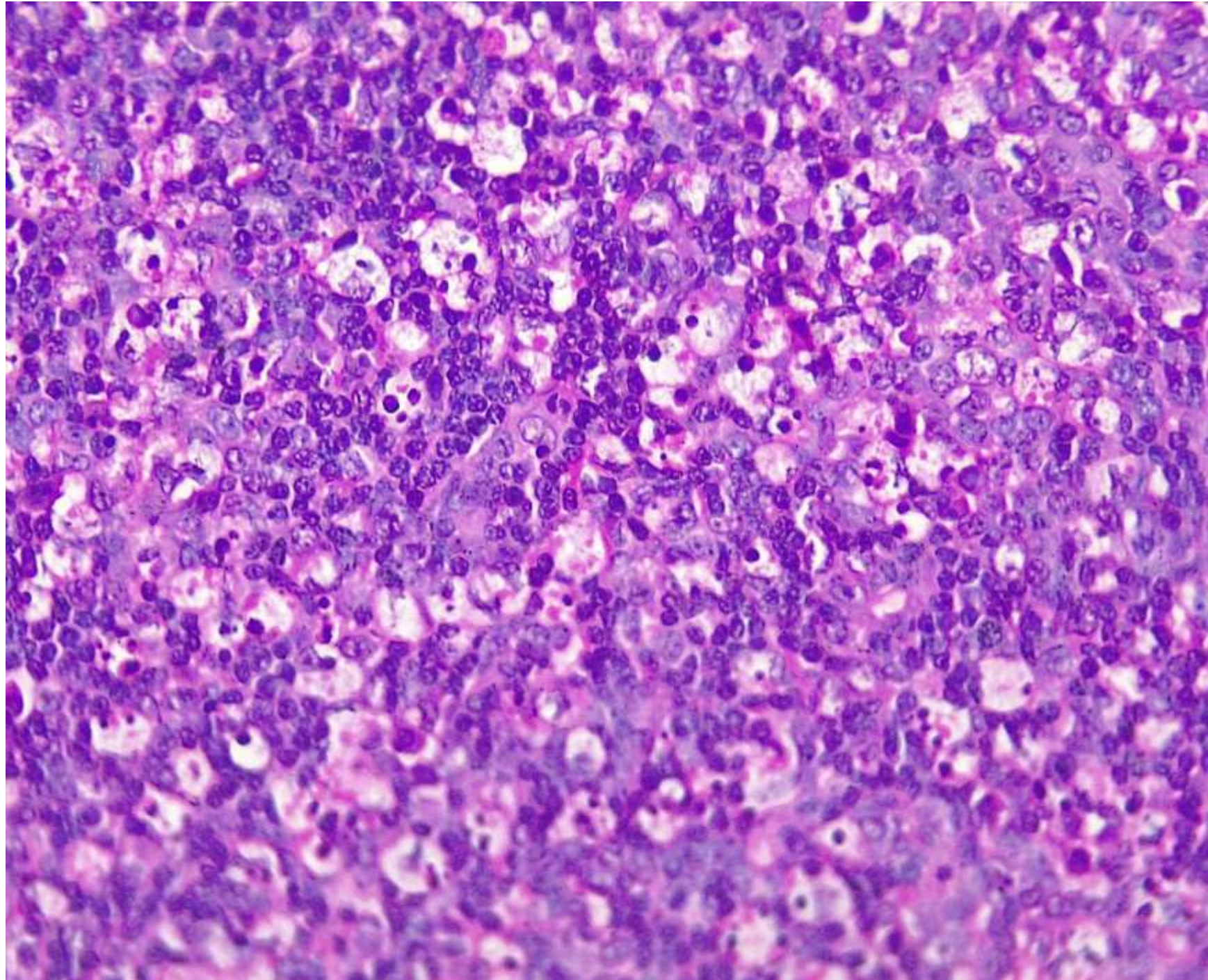


Figure 6: High power view of H & E stained section of cervical lymph node biopsy

- Pale staining histiocytic infiltrates
- Absence of neutrophils and eosinophils.
- No evidence of granuloma or malignancy.

Histopathology of cervical lymph node biopsy :

- suggestive of **Kikuchi Fujimoto Lymphadenitis**

WORK UP

- Tissue AFB staining : No acid fast bacilli seen
- Tissue culture and sensitivity : No bacterial growth in culture after 48 hrs of aerobic incubation at 37° C
- GENEXPERT MTB : Mycobacterium tuberculosis complex not detected

DISCUSSION

DISCUSSION

- Association between KD and DLE is seen here
- The reason for this association is still unknown.
- ? due to exposure of necrosis forming Anti Nuclear Antibodies .
- ? many cases of KD may represent lupus lymphadenitis, thus KD may represent a *forme fruste* of SLE.
- Till date there are only 3 reports of DLE in the absence of SLE in association with KD. [2,7]

DISCUSSION

- Effective management of the condition requires patient education and long-term follow-up.
- Early treatment with hydroxychloroquine has been associated with delayed progression to SLE.
- Notably, hydroxychloroquine has also been used for treatment of KD, which highlights the close association between KD and the various forms of lupus [8].

SUMMARY

- Although KD is a rare condition, clinicians should be aware of its existence and its association with lupus erythematosus.
- Female sex, positive serologies, and cytopenia - risk factors for progression to SLE in patients with KD.
- Patients should receive guidance on avoiding triggers that exacerbate SLE, including:
 - Sun exposure
 - Photosensitizing medications
 - Tobacco

REFERENCES

1. Kikuchi M. Lymphadenitis showing focal reticulum cervical hyperplasia with nuclear debris and phagocytosis. *Acta Hematol Jpn.* 1972; 35:379–380.
2. James JF, Liles JE, Thakore MW. Lupus profundus and discoid lupus following a diagnosis of Kikuchi-Fujimoto disease. *JAAD Case Rep.* 2021 Nov 6; 33:45-47. Doi: 10.1016/j.jdcr.2021.10.028.
3. Zhou AE, Shaikh G, Driscoll MS. Discoid lupus erythematosus of the palms: A case report. *Clin Case Rep.* 2022 Apr 22;10(4): e05048. doi:10.1002/ccr3.5048.
4. Lopez C., Oliver M., Olavarria R., Sarabia M.A., Chopite M. Kikuchi-Fujimoto necrotizing associated with cutaneous lupus erythematosus: a case report. *Am J Dermatopathol.* 2000;22(4):328–333.

REFERENCES

5. Lecoules S., Michel M., Zarrouk V., Gaulard P., Schaeffer A., Godeau B. Recurrent Kikuchi's disease in a patient with discoid lupus. *Rev Med Interne*. 2003;24(9):613–616.
6. Silver S.G., Hong H.C., Ting P.T., Ball N.J. Kikuchi- necrotizing lymphadenitis in association with discoid lupus erythematosus: a case report. *J Cutan Med Surg*. 2004;8(6):442–445.
7. Hurt MA, Weedon D. Weedon D. Weedon's Skin Pathology. 3rd ed. London: Churchill Livingstone Elsevier, 2010. *Dermatol Pract Concept*. 2012 Jan 31;2(1):79–82. doi: 10.5826/dpc.0201a15.. PMID: PMC3997252.
8. Jamali M Ioachim's lymph node pathology, 4th edition *Journal of Clinical Pathology* 2010;63:284.

THANK YOU