Kikuchi's Disease with Inguinal Necrotizing Lymphadenitis: A Case Report

Presenter: Kaushik Rajesh Joshi 2nd year MBBS Student
Guide: Dr. Haricharan K R, Professor Department of Paediatrics
Co- Guides:

Dr. Rahul R, Senior Resident Department of Paediatrics,

Dr. Nirupama M, Professor and Head of the Department of Pathology,

Dr. Asha Benakappa, Professor and Head of the Department of Paediatrics

Departments: Department of Paediatrics
Department of Pathology

Background

- Kikuchi disease is a self-limiting inflammatory disease commonly affecting the paediatric age group.
- It is also known as Kikuchi Fugimoto disease or Kikuchi Fugimoto histiocytic necrotizing lymphadenitis.
- It is characterized by systemic symptoms such as fever, night sweats, weight loss, fatigue, generalized lymphadenopathy, headaches, arthralgia, sore throat, rash, nausea, vomiting, and may even have neurological involvement.
- Kikuchi disease's etiology is unclear, often divided into autoimmune and infectious causes.[1]
- Its diagnosis is based on histopathology of the affected lymph node, and treatment is empirical.
- The histopathology slide characterizes the presence of histiocytes, lymphocytes, multiple necrotic foci, histiocytic karyorrhexis, and the absence of neutrophils and eosinophils.[1]

History

Demographic details:

- Age: 9 years old
- Gender: Male
- Occupation: Student

Chief Complaints:

- Fever for a month
- Pain and swelling in the right inguinal region for a month

History of Presenting Illness

- A previously healthy nine-year-old boy presented in April with complaints of fever for a month along with swelling and pain in the right inguinal region.
- The fever was insidious in onset, on and off in nature, moderate grade, and intermittent without any aggravating factors, but relieved on medications. It was not associated with chills, rigors, diurnal variation, or rashes.
- The patient also noticed a swelling on the upper part of his right groin region 1 month ago. It was spontaneous in onset, gradually progressive, and was associated with non-radiating pain. The swelling was mobile and painful. It was noticed to be a 2x2 cm swelling.
- Pain was insidious in onset, and dull aching type. It was not associated with any aggravating or relieving factors.
- No history of vomiting, bleeding, weight loss, constipation, or evening rise of temperature.

History

- Past history: No similar complaints in the past, and no history of hospitalisation or any surgeries.
- Family history: No similar history in family members.
- Birth history: No significant history.
- Immunization history: Immunized according to the National Immunization Schedule.
- Developmental history: Age-appropriate.

Examination

- The patient was moderately built and nourished, conscious, oriented to time place, and person.
- **Vitals**:
  - Temperature: 100 ºF
  - Heart Rate: 84 bpm
  - Respiratory Rate: 24/ min
  - SpO2: 97% at room air
- **Anthropometry**: Weight – 24 kg

  Height – 129 cm

  BMI – 14.4 kg/m^2
- No signs of pallor, icterus, cyanosis, clubbing, or edema.
- Right inguinal lymphadenopathy was present.

Systemic Examination

- **Cardiovascular System** – Normal S1 and S2 were heard, and no murmurs were heard.
- **Respiratory System** – Bilateral vesicular breath sounds heard. No wheeze or other abnormal breath sounds were heard.
- **Central Nervous System** – Conscious & alert
  - pupils bilateral reactive to light,
  - tone and power normal
  - Reflexes normal

Systemic Examination

•**Per Abdomen** –

INSPECTION :

- The abdomen was flat,
- Umbilicus was central and normal,
- The skin over the abdomen was normal,
- All quadrants of the abdomen moved equally with respiration, no visible mass, peristalsis, sinuses, or dilated veins.

PALPATION :

- Soft, non-tender and no organomegaly was present.
- No local rise of temperature

▪No scars, hernial orifices appear normal.

PERCUSSION: No signs of fluid accumulation on the abdomen.

AUSCULTATION: Bowel sounds heard

Local Examination

- Right inguinal lymphadenopathy was present.
- The swelling was mobile, with smooth skin on its surface.
- It was sized around 3cm x 4cm.
- Swelling was associated with tenderness, but no local rise in temperature.
- Swelling was firm in texture.
- Skin over the swelling was normal with no discoloration.
- The swelling was not associated with any discharge.

Differential Diagnosis

1. **Tubercular lymphadenitis**
2. **Reactive lymphadenitis**
3. **Lupus lymphadenitis**
4. **Cat scratch disease**
5. **Lymphoma**
6. **Infectious mononucleosis**

Investigations Ordered And Interpretation

INVESTIGATIONS	INTERPRETATION
COMPLETE BLOOD COUNT	Elevated neutrophils and reduced lymphocytes
ERYTHROCYTE SEDIMENTATION RATE	ESR levels were elevated
PERIPHERAL SMEAR	Normochromic normocytic blood picture
CBNAAT	Negative
USG - ABDOMEN AND PELVIS	Bilateral necrotic lymphadenopathy

Ultrasound – Abdomen And Pelvis

- Ultrasound scan revealed:
- Normal liver, gall bladder, pancreas, spleen, kidneys, urinary bladder, and prostate.
- <u>Right side</u>:
  - USG revealed 2-3 partially necrotic, enlarged lymph nodes.
  - Largest lymph node measured 30 x 30 mm.

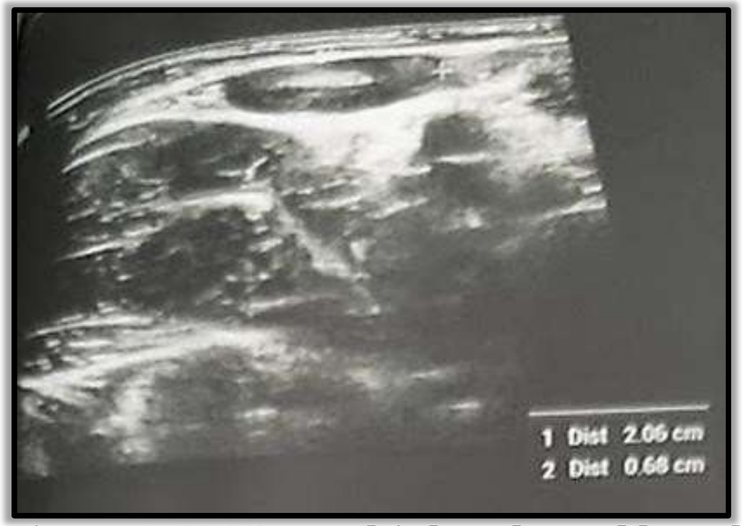

Figure 1: USG- multiple enlarged lymph nodes in the right groin region

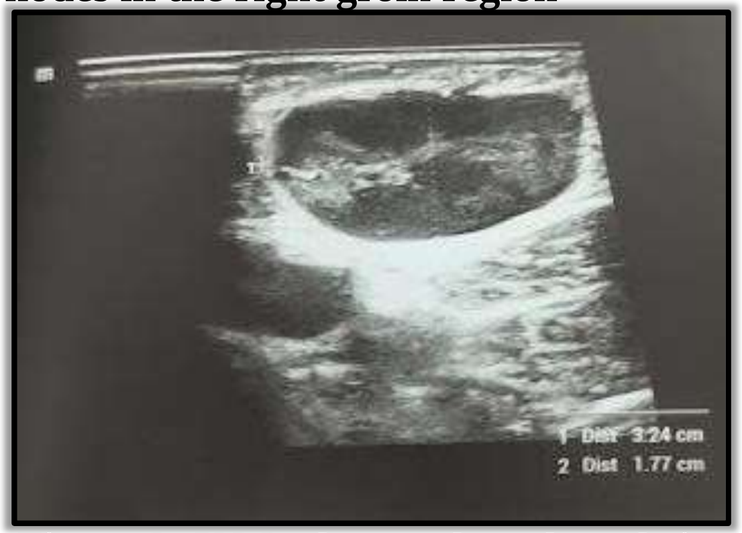

Figure 2: USG - largest lymph node in the right groin region

Ultrasound – Abdomen And Pelvis

- Left side:
  - USG revealed a partially necrotic, enlarged superficial inguinal lymph node measuring 20 x 7 mm.

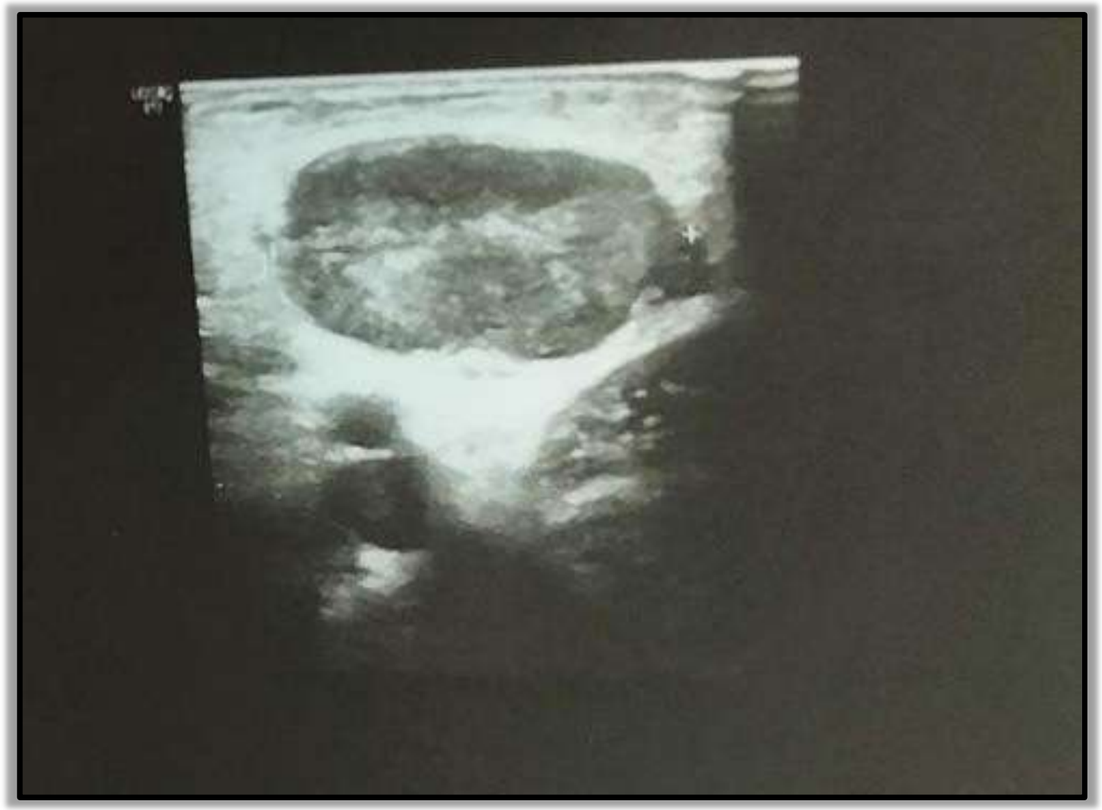

Figure 3 USG - enlarged lymph node in the left groin region

Histopathology

- The patient was scheduled for an excisional biopsy under general anesthesia.
- The biopsy specimen was sent for histopathologic examination.
- Gross: Nodular tissue measuring 3x2.5x0.7 cm.
- Cut surface appeared grey-white.
- The excised right side lymph node was examined under scanner view (10x), and 40x magnification after staining with H and E stain.

Histopathology

- Histopathologic examination revealed:
  - Multiple foci of focal loss of follicular architecture with necrosis of the cortical and paracortical areas.
  - Capsular infiltration
  - Karyorrhectic debris distributed in areas of eosinophilic fibrinoid material.
  - Histiocytes
  - Lymphocytes
  - Absence of neutrophils and eosinophils
- Histopathologic features suggest Kikuchi’s disease.

Histopathology

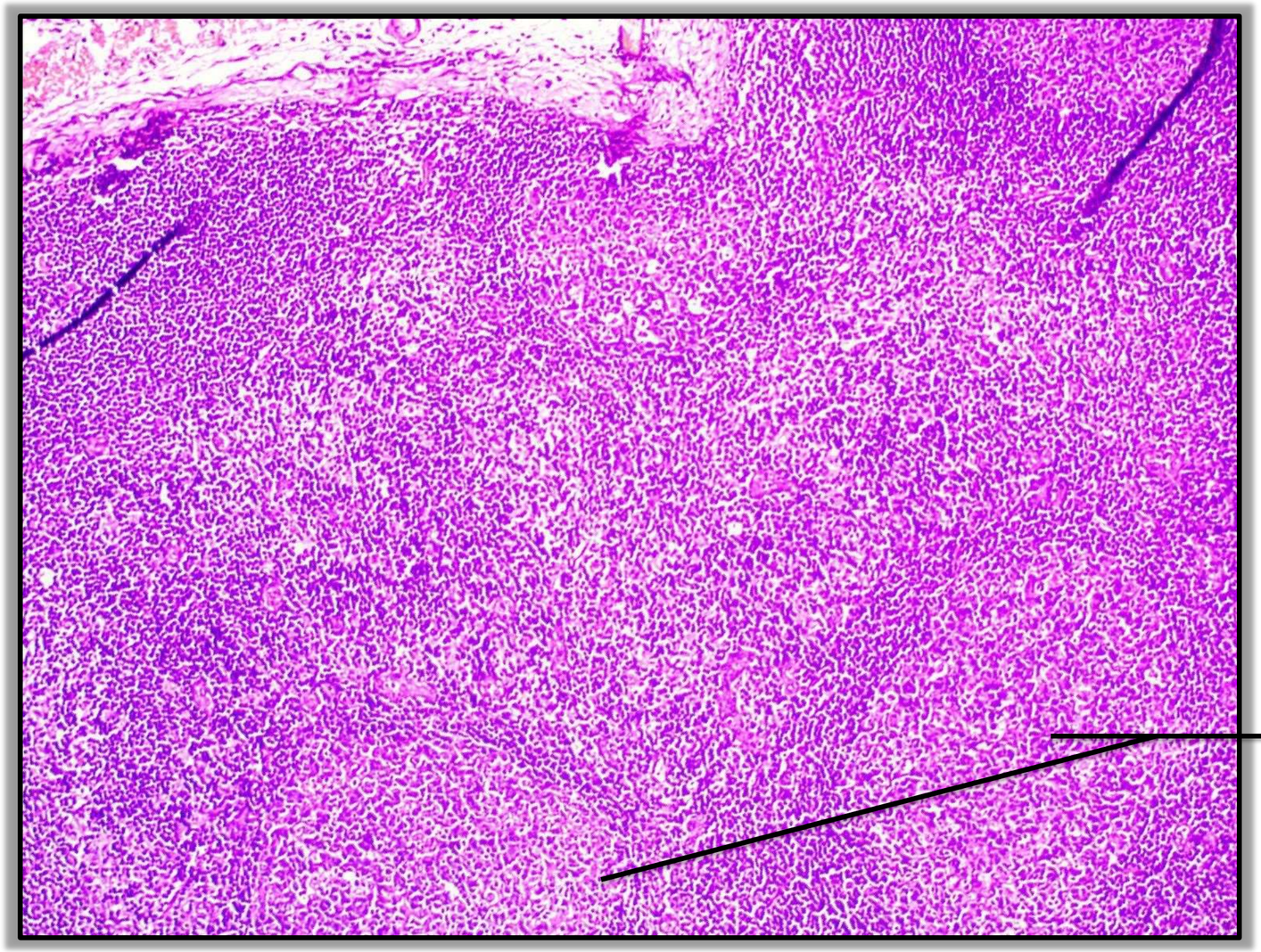

Figure 4: Histopathology of lymph node stained by H and E stain under 10x magnification showing partially retained follicular architecture

Histopathology

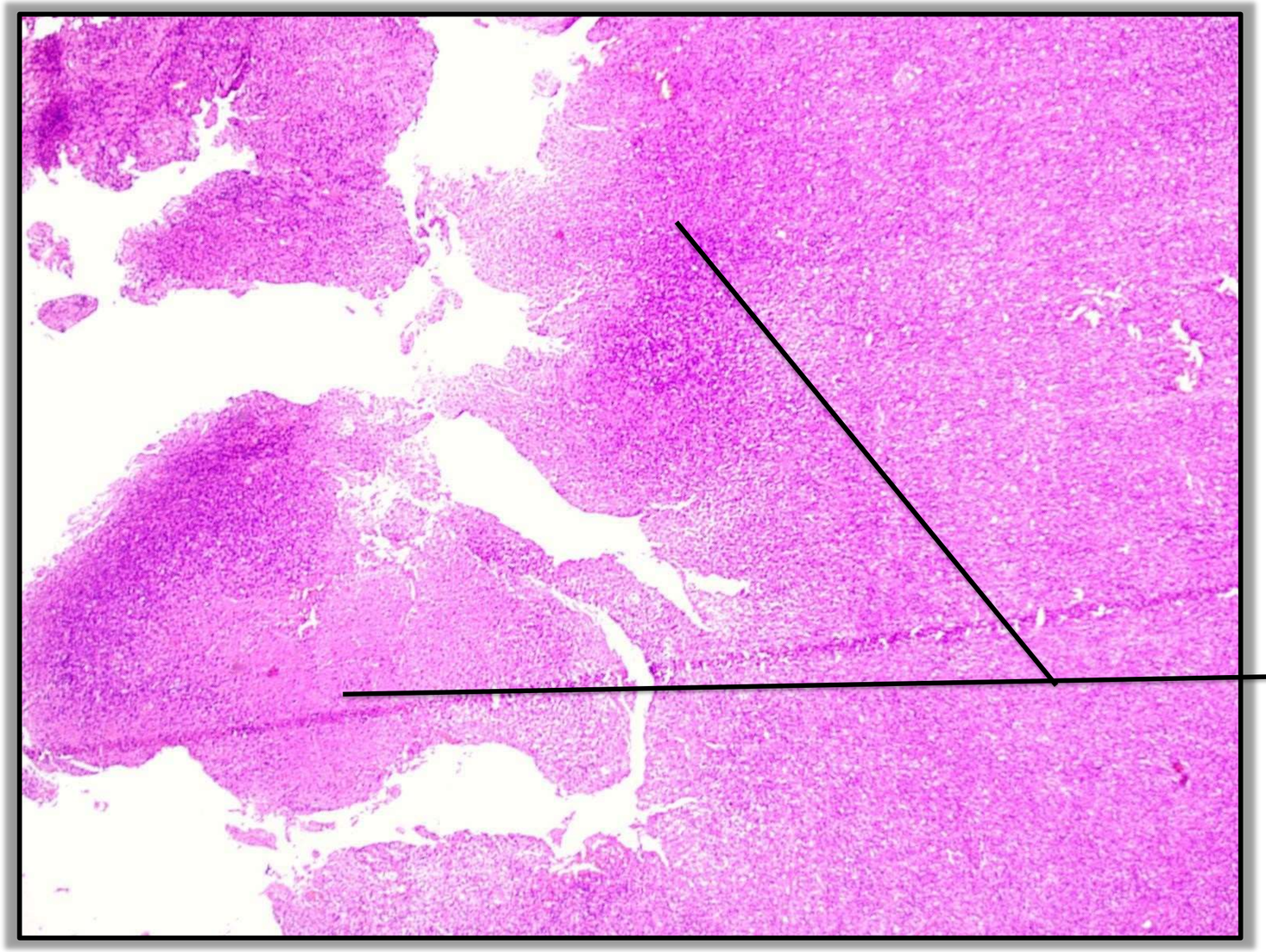

Figure 5: Histopathology of lymph node stained by H and E stain under scanner view showing lymph node architecture with confluent areas of necrosis

Histopathology

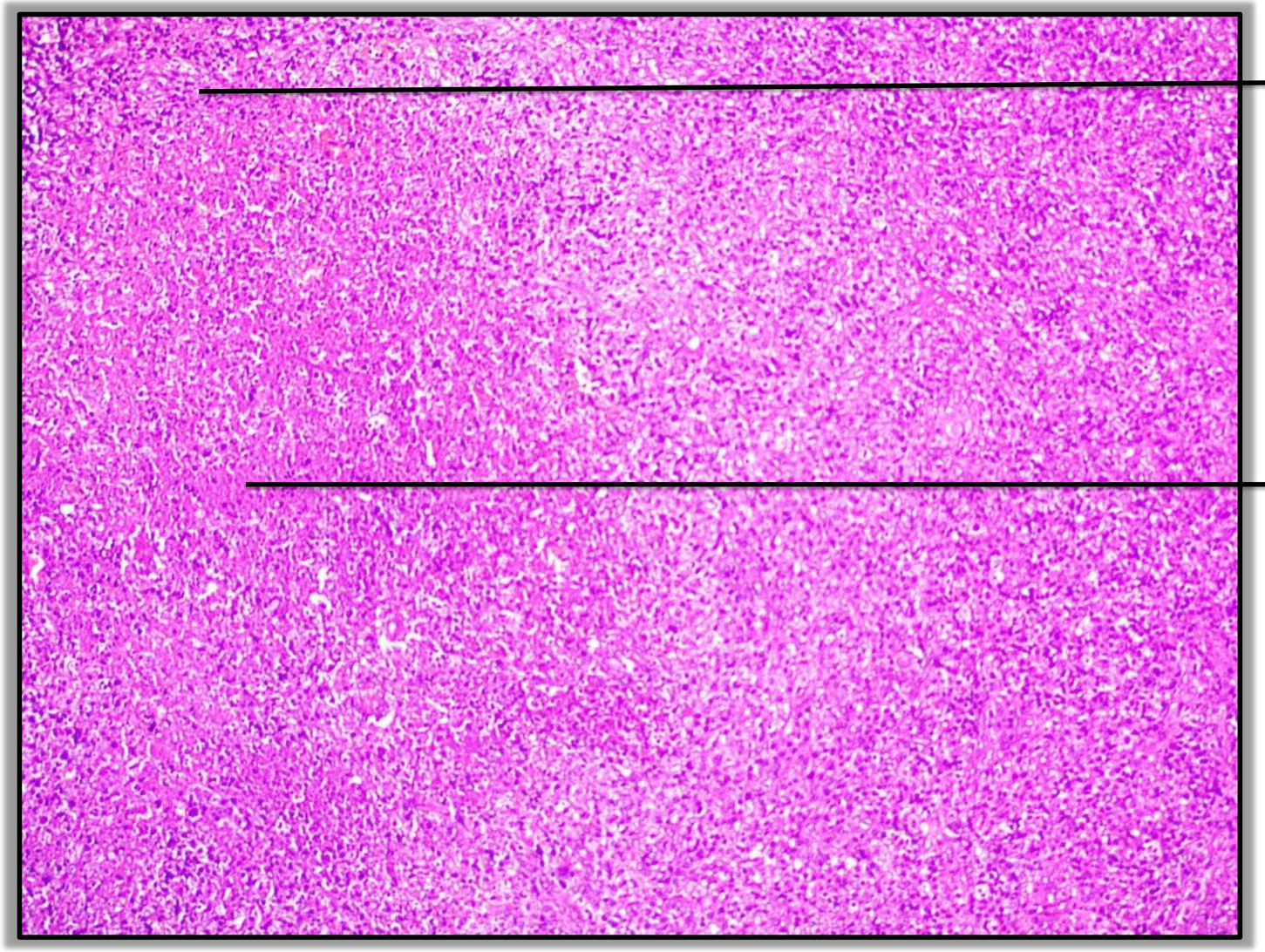

Figure 6: Histopathology of lymph node stained by H and E stain under 10x magnification necrosis with Karyorrhectic debris

Histopathology

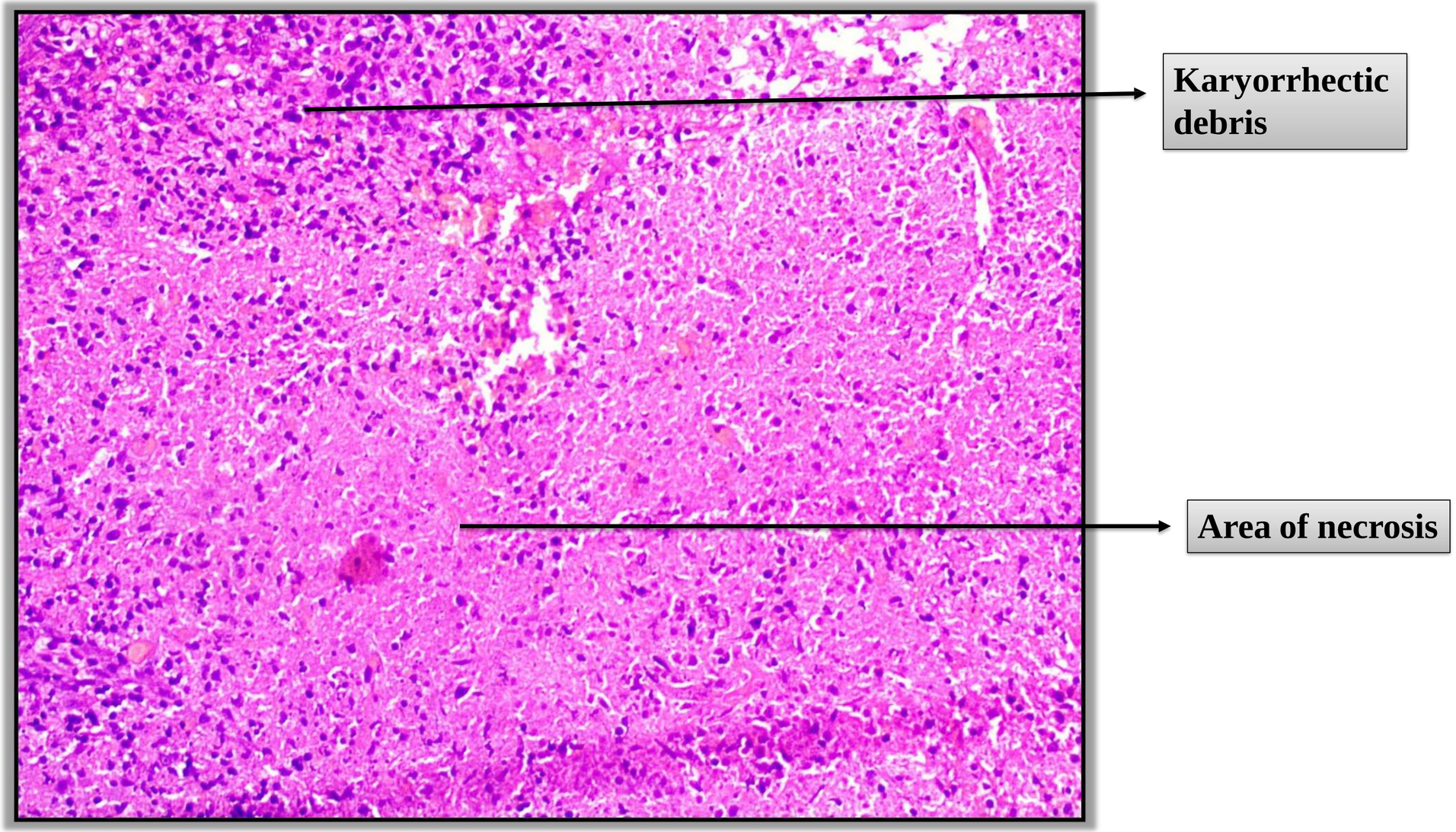

Figure 7: Histopathology of lymph node stained by H and E stain under 20x magnification showing necrosis with karyorrhectic debris

Histopathology

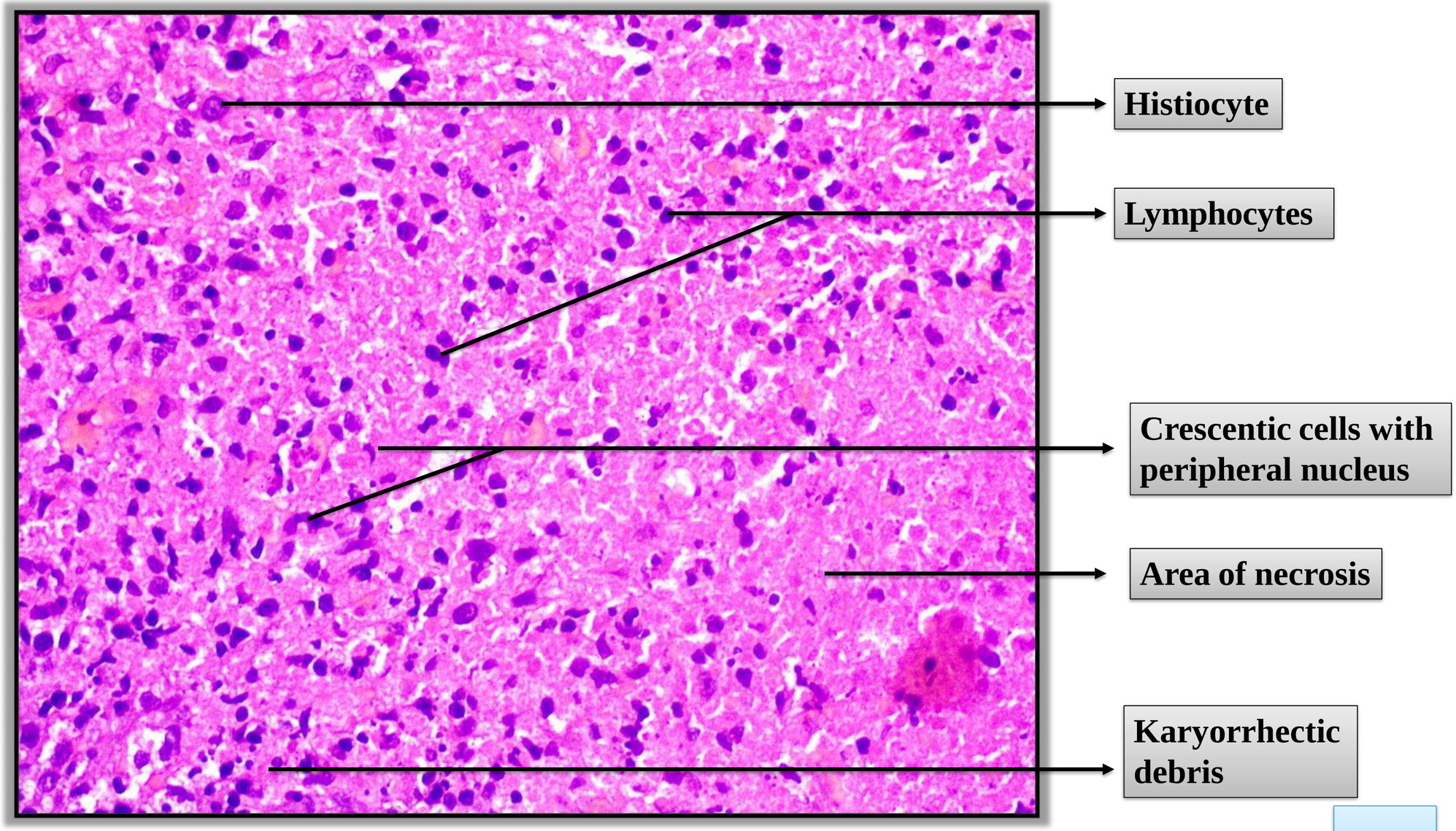

Figure 8: Histopathology of lymph node stained by H and E stain under 40x magnification showing necrosis, karyorrhectic debris with dispersed lymphocytes in the background

Diagnosis and Management

- A final diagnosis of Kikuchi's disease was made based on clinical examinations, and investigations.
- <u>Medications in the hospital:</u>
- INJ AUGMENTIN
- INJ CEFTRIAXONE
- INK AMIKACIN
- TAB PARACETAMOL

Diagnosis and Management

- 13 days following admission, the patient became symptom-free and hemodynamically stable. So, he was discharged with the following advice.
- Vitals at discharge:
  - Heart Rate: 86/ min
  - Pulse: Good volume, regular
  - Respiratory Rate: 20/min
  - Blood Pressure: 96/70 mm/Hg
  - SpO2: 97%
  - Temperature: 98.0 F
- Discharge Advice:
  - Tab PARACETAMOL (500mg) ½ tablet POS SOS for fever
  - Nutritional counseling
  - Review at pediatrics OPD on 10/05/2024.

Discussion

- Kikuchi disease is a self-limiting rare disease commonly seen in the Asian population. It has a female-to-male ratio of 2.75:1.[4]
- Kikuchi's disease has a wide clinical spectrum and is generally self-limiting. However, some patients suffer from prolonged systemic symptoms during the disease, sometimes with severe complications such as HS (Hemophagocytic Syndrome).[3]
- It most commonly affects posterior cervical lymph nodes. It is unilateral, but in 1%-22% of people, there can be generalized axillary, epitrochlear, inguinal, retroperitoneal, lymphadenopathy.[6]
- Multiple infectious agents such as EBV, Parvovirus B 19, herpes simplex virus (HSV) types 1 and 2 (HSV-1 and HSV-2), varicella-zoster virus (VZV), cytomegalovirus (CMV), human herpesvirus (HHV-6, 7, and 8), etc have been suspected as etiological agents, but the causative link has not yet been established.[1][4]
- It also resembles many autoimmune conditions such as SLE, which is different because of the presence of hematoxylin bodies.

Discussion

- Bacterial agents such as Brucella, Bartonella henselae, Toxoplasma gondii, Yersinia enterocolitica, Entamoeba histolytica, and Mycobacterium species have also been suspected to be one of the causes.[2][5]
- It has been postulated that KD represents an exuberant T-cell–mediated immune response to a variety of antigens in genetically susceptible people.
- Compared with the general population, patients with KD more frequently have particular human leukocyte antigen (HLA) class II alleles, specifically HLA-DPA1 and HLA-DPB1.[2]
- Since KD shares age and gender predisposition as well as histologic features with SLE, it has also been proposed that it represents a self-limiting SLE-like autoimmune disorder caused by virus-infected transformed lymphocytes.[6]
- Diagnosis of Kikuchi disease is done based on histopathology of the enlarged lymph node, with immunohistochemistry.
- It has to be differentiated from Kawasaki disease, Lymphoma, Lupus lymphadenitis, etc.

References

1. Masab M, Surmachevska N, Farooq H. Kikuchi-Fujimoto Disease. [Updated 2023 Oct 29]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK430830/
2. Anamarija M. Perry, Sarah M. Choi; Kikuchi-Fujimoto Disease: A Review. *Arch Pathol Lab Med* 1 November 2018; 142 (11): 1341–1346. doi: https://doi.org/10.5858/arpa.2018-0219-RA
3. Lelii, M., Senatore, L., Amodeo, I. *et al.* Kikuchi-Fujimoto disease in children: two case reports and a review of the literature. *Ital J Pediatr* **44**, 83 (2018). https://doi.org/10.1186/s13052-018-0522-9
4. Duan, Wei MD; Xiao, Zheng-Hui MD; Yang, Long-Gui MD; Luo, Hai-Yan MD∗. Kikuchi's disease with hemophagocytic lymphohistiocytosis: A case report and literature review. Medicine 99(51):p e23500, December 18, 2020. | DOI: 10.1097/MD.0000000000023500
5. Racette, S.D., Alexiev, B.A., Angarone, M.P. *et al.* Kikuchi-Fujimoto disease presenting in a patient with SARS-CoV-2: a case report. *BMC Infect Dis* **21**, 740 (2021). https://doi.org/10.1186/s12879-021-06048-0
6. Mahajan VK, Sharma V, Sharma N, Rani R. Kikuchi-Fujimoto disease: A comprehensive review. World J Clin Cases. 2023 Jun 6;11(16):3664-3679. doi: 10.12998/wjcc.v11.i16.3664. PMID: 37383134; PMCID: PMC10294163.

Thank You