AN UNUSUAL CASE OF LYMPHADENITIS

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Guided by

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Complaints

- 14 yr old girl presented with
- Bil. neck swellings since 1 month,
- Fever since 1 wk prior to admission
- Generalised erythematous rashes associated with fever

History of Presenting Illness

- Bil. neck swelling 1 mth prior to admission gradually progressing in size associated with mild pain.
- Oral antibiotics followed by fever and rash 4 days prior to admission

PAST HISTORY

- Mitral valve prolapse on treatment

FAMILY HISTORY

- No h/o contact with known case of TB

Examination

- Generalised flushed appearance
- Right superficial cervical lymph nodes >2cm &left posterior cervical lymph nodes 1-2 cm firm ,mobile with some matting
- ENT normal
- R/S- clear

- P/A- soft, no organomegaly
- CVS & CNS normal
- No other significant lymphadenopathy

Investigations

- CBC

5/6-TC-3300

6/6-TC-2400

12/6-TC-3800 , platelets-2.4 lac

- DC-N-69%,L-25%,M-1%

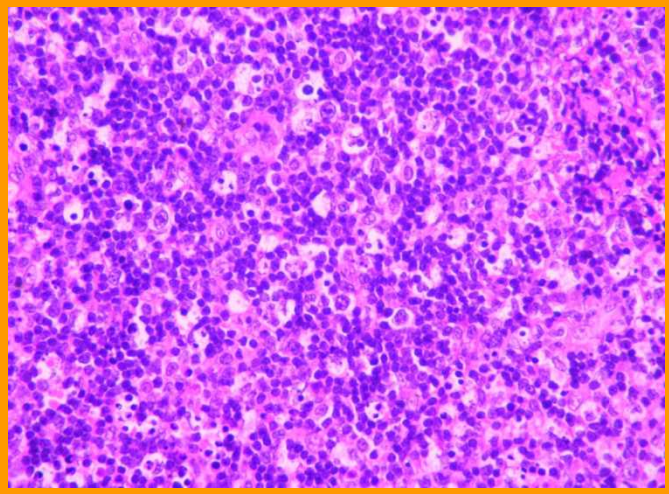
- P.S-leukopenia with reactive lymphocytes

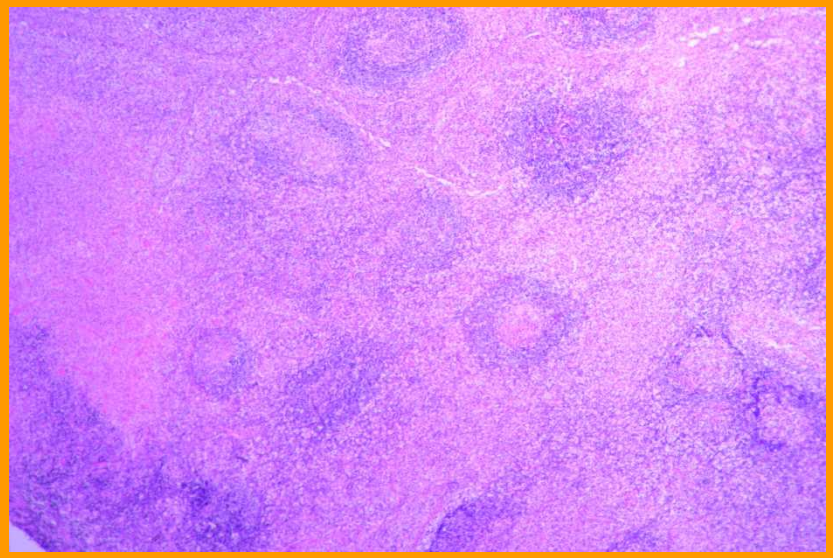
- MPQBC –negative
- Chest x ray normal study
- Mantoux –negative
- Urine routine normal
- Blood culture & urine culture no growth
- Paul Bunnell test

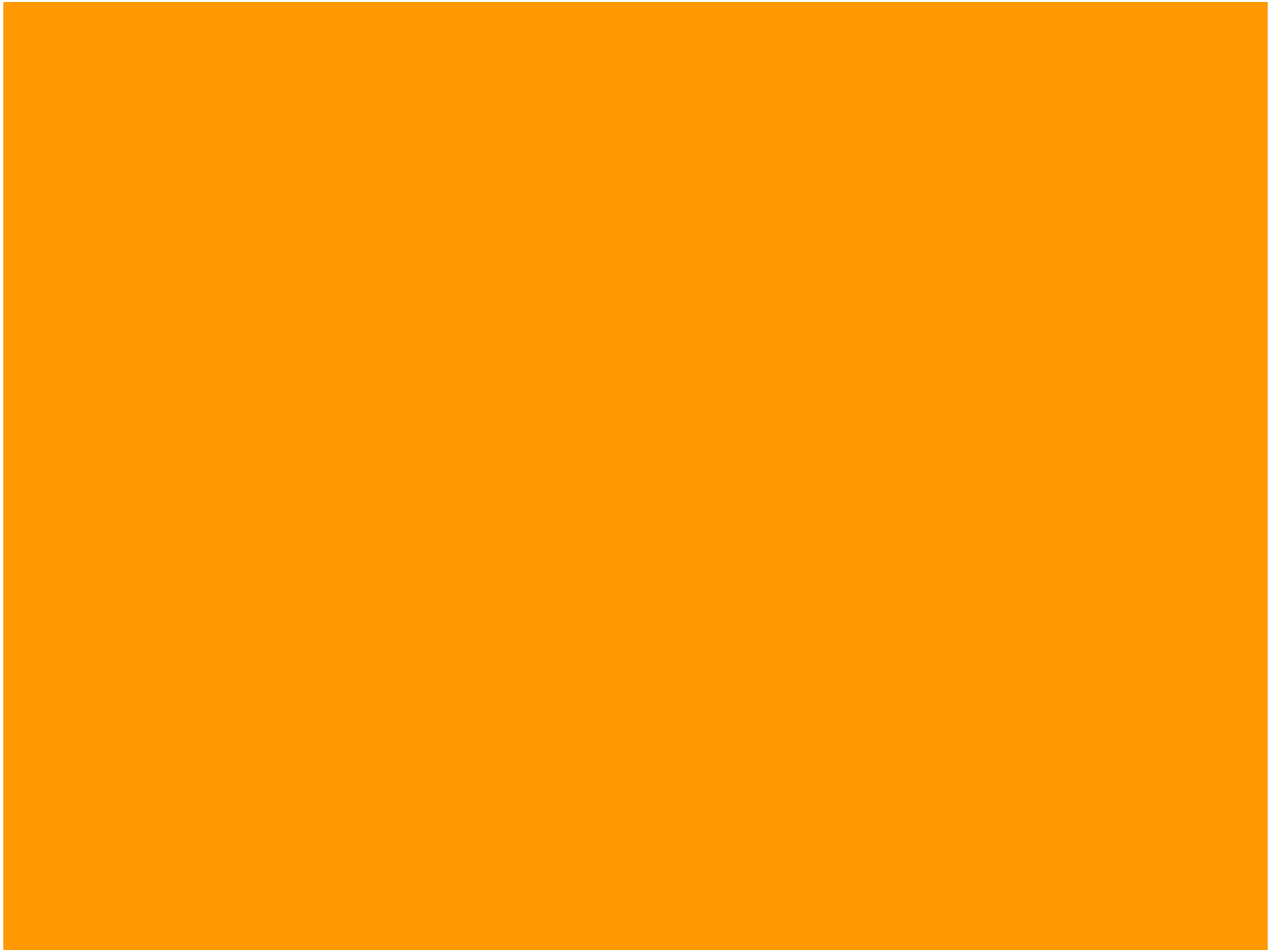
- Vasculitic package negative
- S.LDH was normal

LYMPH NODE BIOPSY

- Extensive necrosis with karyorrhexis
- Scattered residual reactive lymphoid follicles with prominent germinal centres
- Necrotic areas infiltrated by sheets of histiocytes with crescenteric nuclei
- Starry-sky appearance
- No granulomas identified
- Stain for AFB negative







Kikuchi-Fujimoto Disease Background/Epidemiology

- Kikuchi-Fujimoto disease, Kikuchi's disease, or Kikuchi's histiocytic necrotizing lymphadenitis
- First described in Japan in 1972 in 2 young women
- Has since been described all over the world including the United States and Europe
- Literature reviews estimate up to 80% are of Far Eastern descent
- Females to males ratio of about 4:1
- Age distribution of 6 to 80 years reported.

Background/Epidemiology

- Rare, BENIGN disease characterized by the presence of enlarged and inflammed lymph nodes
- No strong genetic predisposition established
- Rare familial variants have been reported, primarily from Japan and Saudi Arabia

Pathogenesis

- UNKNOWN, however, histologic changes suggest an immune response of T cells and histiocytes to an infectious agent
- Infectious agents proposed include EBV, HHV 6 and 8, parvovirus B19, paramyxo virus, parainfluenza virus, yersinia enterocolitica to name a few
- Apoptotic cell death mediated by cytotoxic CD8 T lymphocytes is the principle mechanism of cellular destruction
- Single or multiple paracortical foci with necrosis and a histiocytic cellular infiltrate on lymph node bx

Pathogenesis

- Biopsy show 2 phases- a proliferative phase and a necrotizing phase
- Proliferative phase- follicular hyperplasia, paracortical expansion by lymphocytes, T and B cell blasts, plasmacytoid monocytes and histiocytes (presence of numerous blast cells may confuse with lymphoma, EBV, or HSV infection)
- Necrotizing phase- necrosis without neutrophilic infiltrate; progressive dominance of histiocytes as major cell type

** Absence of neutrophils distinguishes from SLE**

Clinical Presentation

- Most common clinical presentation is fever and cervical lymphadenopathy in a previously well young woman
- Fever is typically low grade and persists for 1 week, rarely up to 1 month
- Other symptoms include fatigue, joint pain/arthritis, night sweats, weight loss
- Some reports describe skin manifestations including facial erythema, erythematous papules, plaques, nodules, ulcers etc in up to 40% of patients

Clinical Presentation- Lymphadenopathy

- Usually CERVICAL and LOCALIZED, particularly post cervical involvement
- Usually only moderately enlarged (1-2 cm) but often assoc. with dull or acute pain
- Typically firm, smooth, discrete, and mobile

Differential:

- Lymphoma (Hodgkin's and non-Hodgkin's)
- Infections including:
  - -EBV/CMV
  - -HIV
  - -Cat Scratch Disease
  - -Tuberculous adenitis
- Autoimmune – in particular , SLE is an important consideration as many patients initially diagnosed with Kikuchi's have subsequently developed SLE (tubuloreticular structures in the lymphocytes and endothelial cells in SLE have been observed similar to those seen in Kikuchi's disease)

Diagnosis

- ** Made by Lymph Node Biopsy (especially to rule out badness like Lymphoma since Kikuchi's is a self limited disease)
- Lab Studies usually NORMAL across the board (no one abnormality pointing toward Kikuchi's disease); however, case series have shown:
  - Leukopenia in up to 30% with atypical lymphocytes in 25%
  - ESR elevated (even above 60 in 70% of pts in one series)
  - LFTs elevated mildly
  - Elevated serum LDH

Diagnosis

- ANA and RF generally negative (but ANA needs to be done in any pt. suspected of having Kikuchi's)
- Infectious serologies should be done including:
  - EBV/CMV
  - HIV
  - Toxo
  - Bartonella
  - Yersinia enterocolitica
- CT Imaging- shows perinodal infiltration and homogeneous nodal contrast enhancement of lymph nodes

Treatment

- No standard treatment has been established for Kikuchi-Fujimoto disease

- Many patients receive antibiotics intially, however, unless infectious agent identified this is not recommended

Prognosis

- This is a benign, self-limiting condition with excellent prognosis
- It can recur over time, quoted at 3% (one case report had patients with 4 different episodes over 18 year period)
- Patients need to be followed for recurrences, as well as for predilection to developing SLE which is not completely understood at this time

TAKE HOME MESSAGE

- Rare cause of prolonged fever with lymphadenopathy
- Autoimmune predilection