A teenage girl of Pakistani ethnicity presented with massive lymphadenopathy involving the posterior cervical lymph nodes. An MRI neck showed nodal conglomerates measuring 6 × 4 cm, raising a suspicion of lymphoma or an infectious etiology. USS abdomen was unremarkable. Extensive viral serologies/PCR were non-contributory. TB Elispot was negative.

An excision biopsy of the cervical lymph node was done and histopathology revealed necrotising lymphadenitis which was unequivocally consistent with Kikuchi disease, a close differential being lupus lymphadenitis with no evidence of lymphoproliferative malignancies (slides further reviewed by onco-pathologists), Special stains for micro-organisms and further fungal/mycobacterial cultures and 16s rDNA PCR were negative. Immunology showed a borderline ANA of 1:80 (speckled pattern) with immunoglobulins, ENA, RF, dsDNA, complements and antiphospholipid antibodies being negative or normal. There were no features to support a diagnosis of lupus.

**Results:** In view of extranodal/systemic features she was commenced on an oral course of steroids. This resulted in effective resolution of fevers and shrinkage of the bulky lymphadenopathy enabling discharge. ESR and CRP normalized at this stage with ferritin trending down to 599. LDH came down to 408 and Vit B12 levels dropped to 1200.

An attempt to wean steroids within a couple of weeks resulted in resurgence of high fevers, constitutional symptoms and enlargement of cervical lymphadenopathy to a mass measuring 10×6 cm. ESR, CRP and ferritin increased slightly to 38, 17 and 342 respectively but LDH rose significantly to 1632. Addition of Hydroxychloroquine was of little benefit. Increasing oral steroids back to full dose had little effect although splitting the dose seemed to alleviate fevers. IV methyl Prednisolone was deferred, as conceived to have little advantage over oral steroids. IVIG (1g/kg) was administered over 2 days, which resulted in complete resolution of fevers, dramatic reduction in bulky lymphadenopathy and swelling and normalization of ESR, CRP and ferritin. LDH came down to 730. This has enabled further successful weaning of steroids.

(Pictures available if accepted)

**References**


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**25. KIKUCHI DISEASE WITH EXTRANODAL INVOLVEMENT:**

**RESPONSE TO IVIG AND ROLE OF INFLAMMATORY MARKERS**

Akhila Kavirayani, et al (TBC)

**Aims:** To describe a clinical case of Kikuchi disease with systemic features, response to IVIG therapy and to look at the role of possible biomarkers

**Background:** Kikuchi-Fujimoto disease, an extremely rare cause of lymphadenopathy is histiocytic necrotizing lymphadenitis. It can run a benign course although severe/extranodal forms are known to occur, which might warrant systemic immunosuppression. A few cases might further evolve into lupus.

**Methods:** A teenage girl of Pakistani ethnicity presented with massive unilateral tender cervical lymphadenopathy involving the posterior triangle and accompanying high-grade fevers/constitutional symptoms for 4 weeks unresponsive to antibiotics, alongside papular rashes over arms. CRP was 108, ESR was a maximum of 65, ferritin was 4577 at its highest, with no other features to support a concurrent HLH; LDH 857, ALT 58, Vit B12 > 2000 (upper limit 900). There were no cytopenias. An MRI neck showed nodal conglomerates measuring 6 × 4 cm, raising a suspicion of lymphoma or an infectious etiology. USS abdomen was unremarkable. Extensive viral serologies/PCR were non-contributory. TB Elispot was negative.