Recognizing Kikuchi Disease: An Unusually Severe Case.

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Kikuchi-Fujimoto disease (KFD) is a rare histiocytic necrotizing lymphadenitis with unclear pathogenesis. Autoimmune and infectious etiologies have been proposed (commonly, EBV), possibly causing over production of IL-6 and Interferon gamma. Worldwide, most frequently reported in young Asian females but can affect all ethnicities. In the USA, 75% cases are Caucasian. Disease course is typically self-limited, presenting with fever, rash, splenomegaly, and lymphadenopathy.

### REFERENCES

5. Up-to-Date and Mayo Clinic hot topic presentation by Dragan Jevremovic, MD

### CASE PRESENTATION

- 45 y.o male PMHx Hashimoto thyroiditis presents with fevers, non-blanching maculopapular/purpuric rash, respiratory symptoms and diffuse lymphadenopathy
  - CT showed extensive lymphadenopathy in neck, chest, abdomen, and pelvis.
  - The patient’s course was complicated by distributive shock requiring mechanical ventilation and pressors, small b/l PE’s, new onset afib with R/R, warm autoimmune hemolytic anemia requiring transfusions, and APF/ATN requiring CRRT.
  - Maculopapular rash bx reports lymphomatoid papulosis type A vs. drug eruption.
  - Initial femoral lymph node core needle bx showed reactive changes & polytypic plasmacytosis. No evidence of lymphoproliferative process.
  - Bone marrow bx w/o evidence of hematopoietic neoplasm.
  - Flow cyto w/o evidence of clonal B or T-cells, increased CD34 blasts or double negative T cells.
  - Workup neg for wide variety infectious etiologies.
  - Rheumatological workup showed type III mixed cryoglobulins and low complement levels. Neg SLE serology.
  - Responded well to pulse steroid therapy and anti-IL6 bx with presumptive diagnosis of multicentric Castleman’s disease.
  - However neg IL-6 levels.
  - Axillary node excisional bx showed necrotizing granulomatous lymphadenitis with numerous histocytes and preserved architecture in non-necrotic areas.
  - Interestingly, in situ hybridization with EBV-encoded small RNA was positive. In setting of neg EBV serologies and positive titer.
  - Bs consistent with Kikuchi vs SLE. Kikuchi dx of exclusion.

- Although cervical lymphadenopathy is most common, nodal involvement maybe variable and diffuse, possibly correlating with multi-organ dysfunction.
- As noted in our patient, a transient rash occurs in approximately 10% of cases and may also indicate a severe course with possible subsequent development of Systemic Lupus Erythematosus (60%).
- Mixed cryoglobulinemia suggestive of autoimmune and chronic infections states is not usually reported and may be underestimated in the diagnosis of KFD.