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 affects all ethnicities. In the USA, 75% cases are Caucasian.

Disease course is typically self-limited, presenting with fever, rash, splenomegaly, and lymphadenopathy.

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 PEs, new onset afib with RVR, warm autoimmune hemolytic anemia requiring transfusions, and AF/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

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 underutilized in the diagnosis of KFD.

CONCLUSION

Our case demonstrates the importance of excisional biopsy and histopathology in recognizing KFD within the broad differential for generalized lymphadenopathy.

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