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Recognizing Kikuchi Disease: An Unusually Severe Case

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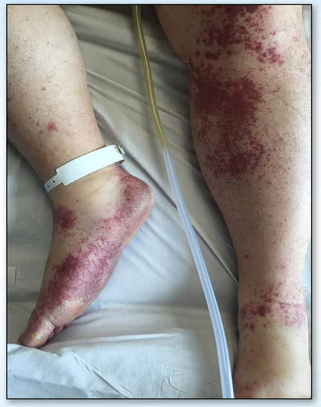
INTRODUCTION

- 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.

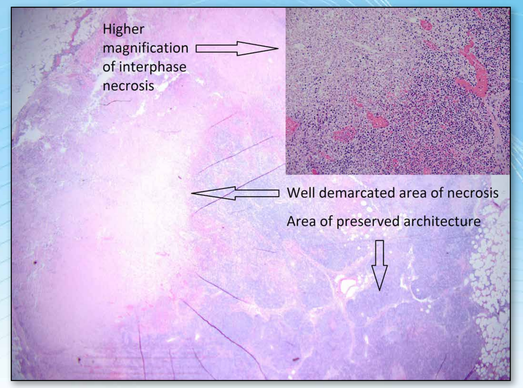
CASE PRESENTATION

- 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 PE's, new onset afib with RVR, warm autoimmune hemolytic anemia requiring transfusions, and ARF/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 tx with presumptive diagnosis of multicentric Castleman's disease
    - However negative IL-6 levels
  - Axillary node excisional bx showed necrotizing granulomatous lymphadenitis with numerous histiocytes 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 titers
    - Bx consistent with Kikuchi vs SLE. Kikuchi dx of exclusion



RESULTS

- 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.

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