Kikuchi Fujimoto disease (KFD), or Kikuchi histiocytic necrotizing lymphadenopathy, is an extremely rare disease known to have a worldwide distribution with a higher prevalence among Japanese and Asians. The cause of KFD is thought to be viral or autoimmune. KFD is a benign, self-limited disease that resolves in 1 to 4 months. An early biopsy can be instrumental in preventing unnecessary investigations and potentially harmful treatments. We report a case of a woman who presented with this rare condition.

A 31-year-old Caucasian woman presented to the hospital with progressively worsening, painful right cervical and right axillary lymphadenopathy of 10 days duration. She denied fever, chills or malaise. She had similar episodes of recurrent painful lymphadenopathy in the past. During these episodes she was treated with empiric antibiotics, followed by partial resolution of the symptoms. Her other medical history was significant for asthma and chronic bronchitis. She denied any sick contacts or recent travel. She did not have any pets at home and denied any recent contact with cats or insects. She did not have high-risk behaviors for HIV.

Physical examination revealed enlarged, tender lymph nodes in the right posterior cervical, axillary and supraclavicular regions. A complete blood count showed an absence of leukocytosis and a mild predominance of lymphocytes. A CT scan of the neck confirmed multiple enlarged lymph nodes in the right posterior neck, carotid sheath, submandibular and supraclavicular regions. A CT scan of the chest revealed lymphadenopathy in the anterior mediastinum, left para-tracheal, pre-tracheal and bilateral axillary regions, while the abdomen and pelvis were unremarkable. HIV RNA, Bartonella antibodies, EBV, immunoglobulin panel, CMV, RPR, and ANA were negative.

Biopsy of the right cervical lymph node revealed crescentic histiocytes admixed with nuclear debris and apoptotic cells (Figures A, B, C), consistent with Kikuchi lymphadenopathy. Immunostaining of lymphoid cells was positive for CD3, CD4 and CD8 T cells. She was treated symptomatically with pain relief and hydration and was discharged after her symptoms had improved.

The diagnosis can only be made by histological examination of the node, which may show the following changes typical for KFD: paracortical necrotic foci, surrounded by histiocytic aggregates, irregular rounded eosinophilic areas of different sizes in the paracortex and cortex with the presence of numerous histiocytes, lymphocytes, immunoblasts, plasmacytoid monocytes and eosinophil granulated cellular debris. It is typical not to find epithelioid cells and neutrophil granulocytes in the necrotic centers. In particular, the disease needs to be distinguished from high-grade lymphoma and SLE lymphadenitis.
In KFD painful, palpable lymphadenitis is localized to one or two sites in the head and neck territories in young adults (third decade) and is associated with fever. Other clinical manifestations are infrequent: arthralgias, cutaneous rash, sweating, splenomegaly. Leukopenia is present in 50% of the cases with sometimes “atypical” lymphocytes.

References

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Disclosures
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