Kimura disease

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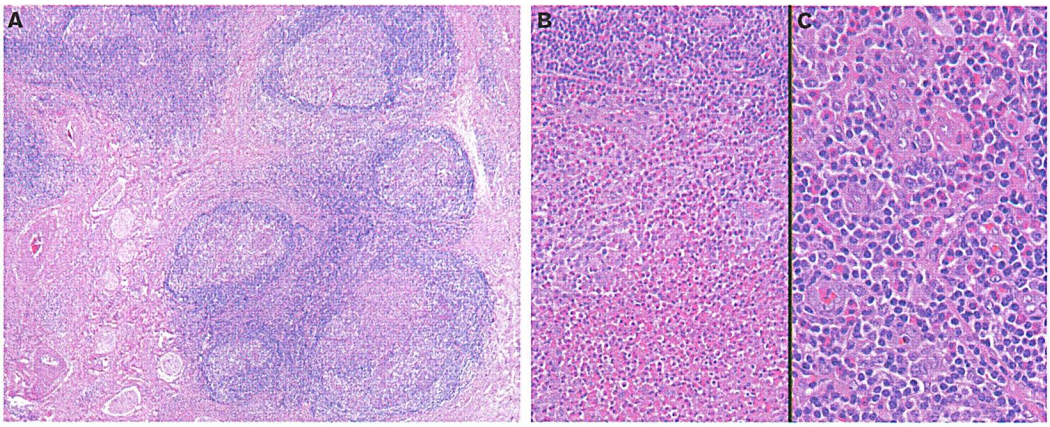

*Figure. **A:**. Marked fibrosis, follicular hyperplasia, and vascular proliferation with eosinophilic infiltrates are seen in the germinal centers. **B:** Eosinophils form a microabscess in a destroyed germinal center. **C:** The vascular proliferation of endothelial cells is seen within the germinal center.*

Kimura disease is a rare, chronic inflammatory disorder that involves subcutaneous tissues, predominantly those in the head and neck region. Its etiology is unknown, but it is frequently associated with regional lymphadenopathy and/or salivary gland involvement. Kimura disease has a predilection for males of Asian descent. Clinically, it can simulate a neoplasm, and most patients have peripheral blood eosinophilia and elevated serum immunoglobulin E (IgE) levels.

These solitary masses invariably affect lymph nodes and subcutaneous soft tissues. Their characteristic histologic features include preserved nodal architecture, florid follicular hyperplasia with reactive germinal centers, protein and IgE deposits in germinal centers, germinal center necrosis, eosinophilic infiltrates, proliferation of postcapillary venules, sclerosis, polykaryocytes, eosinophilic folliculolysis, and prominent eosinophilic microabscesses (figure).

Kimura disease is sometimes confused with angiolymphoid hyperplasia with eosinophilia, Hodgkin lymphoma, angioimmunoblastic T-cell lymphoma, florid follicular hyperplasia, Castleman disease, and lymphadenopathy secondary to drug reactions and parasitic infections. Appropriate cultures, histochemical stains, and immunophenotypic studies will help separate these unique and distinct lesions.

Kimura disease is a chronic disorder—usually a localized process without systemic symptoms—that follows an indolent clinical course. Itchiness, urticaria, and chronic eczema, along with renal disease, are occasionally associated. Surgery is the mainstay of therapy, although regional or systemic corticosteroid therapy, cytotoxic therapy, and radiation have been used. Recurrence after surgery or following discontinuation of steroid treatment is common.

Suggested reading

Hui PK, Chan JK, Ng CS, et al. Lymphadenopathy of Kimura's disease. Am J Surg Pathol 1989;13:177-86.

Ioachim HL, Ratch H. Kimura lymphadenopathy. In: Ioachim HL, Ratch H, eds. Ioachim's Lymph Node Pathology. Philadelphia: Lippincott-Raven, 2002:209-11.

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