Kimura’s disease is a benign, uncommon, chronic inflammatory condition that usually presents with painless subcutaneous nodules or plaques in the head and neck region. Although the disease is predominantly found in Asian populations, there are occasional cases reported among Caucasians and rare occurrences in African populations. The etiology and pathogenesis of the disease are unknown and the clinical presentation can mimic several benign and malignant disease states. The accurate diagnosis of Kimura’s disease is based on clinical and histopathological findings. There is no evidence of malignant transformation and occasional spontaneous resolution occurs. Various treatment modalities have been suggested in the management of this condition. Oral corticosteroids have been the mainstay of therapy, even though steroid withdrawal can result in lesion recurrence.

Kimura’s disease is an uncommon, chronic, inflammatory condition that usually presents with painless subcutaneous nodules or plaques in the head and neck region. Originally described in Chinese literature in 1937 as “eosinophilic hyperplastic lymphadenopathy,” the disorder became more widely known as Kimura’s disease following a report by Kimura et al in 1948. Kimura’s disease shares some of the clinicopathologic features of angiolymphoid hyperplasia with eosinophilia (ALHE), although they are considered distinct entities by most authors. The etiology and pathogenesis of the disease are unknown, however, an allergic or autoimmune process that incorporates lymphocytes, eosinophils, and blood vessels has been proposed.

The clinical presentation of Kimura’s disease mimics several benign (eg, pyogenic granuloma) and malignant (eg, lymphoma) disease states, and physicians who encounter patients with unexplained swelling, nodules, or plaques in the head and neck should include Kimura’s disease in their differential diagnosis. The epidemiology of the condition follows a predominantly Asian distribution with occasional cases reported among Caucasians and very rare occurrences in African populations. We present a case of Kimura’s disease without eosinophilia in an African American male.

Case Report

A 40-year-old African American male presented with a raised lesion along his right preauricular area that had increased in size over a 6-week period. The patient also related a 3-year history of intermittent swelling along this area, in addition to his right and left zygoma. He denied any pain or discharge from the site and had been treated empirically for this condition with amoxicillin/clavulanate in the past. The patient’s medical history was remarkable for tobacco use, hypercholesterolemia, and mild hypertension that was attributed to excess alcohol use. He was not currently taking any medications. On review of systems, there was no facial trauma, focal infection of the head or neck, or history of herpes simplex infection.

On physical examination, he was afebrile and normotensive, and the remainder of his vital signs were within normal limits. The patient’s head and neck examination revealed a 2- by 1-cm nodule along the right preauricular area (Figure 1, A and B). No additional nodules, lesions, or lymphadenopathy were noted. Laboratory results included an erythrocyte sedimentation rate of 9 mm/h, total white blood cell count of 9.5 x 10^9/UL with a differential of 50% granulocytes, 44% lymphocytes, and 6% monocytes. No eosinophils were reported, and the remainder of the hematologic laboratory values were normal. Blood chemistry values...
were within normal limits, with the exception of an elevated cholesterol of 260 mg/dL. Serum IgE levels were not drawn. A differential diagnosis of this presentation included malignant/neoplastic (low-grade non-Hodgkin’s and Hodgkin’s lymphoma, metastatic carcinoma with primary site from head and neck region) and nonmalignant, primarily infectious (chronic sialadenitis, AIDS-related lymphadenopathy, regional abscesses or infection) etiologies.

The patient underwent excisional biopsy of the lesion and had an unremarkable postoperative course. A 2-cm ellipse of skin and subcutaneous tissue was received and contained a vaguely nodular firm region that was grossly visible in the subcutaneous region. Microscopically, histologic sections revealed nonencapsulated angiolymphoid lesions composed of small benign vessels embedded in a dense lymphoid infiltrate, which was composed of reactive follicles with germinal centers and admixed numerous eosinophils (Figure 2). A few plasma cells with associated fibrosis were also identified. A proliferation of epithelioid endothelial cells was not observed, and no necrosis or granulomas were present. The epidermis and dermis were normal. The histopathologic findings were interpreted to be diagnostic of Kimura’s disease. On follow-up, the patient was counseled as to the pathogenesis and clinical course of the disease, and no therapy was instituted. After 1 year, the patient continued to have intermittent, bilateral, asymptomatic facial swelling, primarily along his left and right zygoma.

**Discussion**

Kimura’s disease and ALHE have some overlapping features, with most cases of Kimura’s disease occurring in an Asian population. Some studies support the concept that there are 2 distinct disorders based on clinicopathologic features. ALHE, also known as histiocytoid hemangioma and epithelioid hemangioma, may represent a benign vascular neoplasm. Kimura’s disease has no conclusive pathogenetic mechanism; however, a benign inflammatory process or reactive or immune-mediated response is most commonly proposed. The detection of elevated serum IgE levels and presence of IgE deposits in lymphoid germinal centers and glomeruli of patients with associated nephrotic syndrome support an immunologic pathogenesis.

The distinction between Kimura’s disease and ALHE has important clinical implications. Kimura’s disease has been associated with renal disease, particularly nephrotic syndrome. Asthma and Löeffler’s syndrome have also been reported with Kimura’s disease. The patient in our case had no clinical or laboratory features of these conditions. Kimura’s disease and ALHE share common features, such as a predilection for the head and neck region, development of soft-tissue masses, and a tendency to recur. However, ALHE is found in an older, predominantly female population with smaller (<1 cm), intradermal, or more superficial lesions. Although Kimura’s disease presents predominantly in young Asian males, ALHE can occur in all races without a predilection for age or gender.

The accurate diagnosis of Kimura’s disease, based on clinical and histopathologic findings, is important because clinically it can be mistaken for a malignancy. Biopsy and histopathologic examination are required to confirm the diagnosis and rule out malignancy. Kimura’s disease involves subcutaneous tissue and sometimes salivary glands and lymph nodes of the head and neck. Histologic sections show a dense, chronic, inflammatory infiltrate composed of lymphocytes, plasma cells, and epithelioid cells.
phocytes that form lymphoid follicles with occasional germinal centers admixed with numerous eosinophils that can form eosinophilic abscesses. Lesser numbers of plasma cells and mast cells are admixed. The inflammatory infiltrate is accompanied by a proliferation of small blood vessels and fibrosis. The main differential diagnosis is ALHE with eosinophilia, which also occurs in the head and neck region. Histologically, ALHE is composed of a lympho eosinophilic inflammatory infiltrate associated with prominent blood vessels, thus sharing several overlapping features with Kimura’s disease. ALHE does not involve lymph nodes or salivary glands like Kimura’s disease. ALHE is usually located in the dermis and is more superficial than Kimura’s disease, which occurs in the subcutis. Although lymphoid follicles are not always present in ALHE, a histologic feature considered distinctive of ALHE is the presence of epithelioid or histiocytoid endothelial cells that line the vessels and give rise to the term epithelioid hemangiomma. Epithelioid endothelial cells are not seen in Kimura’s disease.

In the present case, we observed the lesion in the subcutaneous tissue, which contained prominent lymphoid aggregates, some with germinal centers. In addition, the lesion lacked vessels demonstrating histiocytoid endothelial cells, all features favoring the diagnosis of Kimura’s disease over ALHE.

Radiographic studies that have been used to evaluate patients with suspected Kimura’s disease are variable and oftentimes nonspecific. Ultrasound may demonstrate nodes that are solid, round or oval, or hypoechoic. Contrast enhancement on computed tomography may be demonstrable, but does not appear to be a consistent finding. There are no characteristic features of Kimura’s disease with magnetic resonance imaging; however, magnetic resonance imaging can help to define the extent of the disease in more than one plane. TI-201 SPECT scanning, a tumor-seeking diagnostic agent and tool, will show elevated uptake on early and delayed imaging.

Kimura’s disease is a benign condition that often follows a prolonged, indolent course. There is no evidence of malignant transformation, and occasional spontaneous resolution occurs. Because of the significant association of Kimura’s disease with renal disease, periodic assessment of kidney function is recommended. Various treatment modalities have been suggested in the management of this condition. Surgical excision has been promoted and may be indicated when lesions encroach or compromise vital head and neck structures (eg, facial nerve, parotid gland) or for cosmetic reasons. Despite this aggressive therapy, lesions tend to recur. There is one report of a trial of radiation therapy, followed by surgical excision and postoperative antiallergics and anti-inflammatory agents. The author concluded that this intervention resulted in a lower rate of recurrence; however, the risks associated with this modality far outweigh the benefits of expectant management in this benign disease process.

The mainstay of therapy has been oral corticosteroids, even though steroid withdrawal can result in lesion recurrence and a proportion of lesions are refractory to this treatment modality. There have been reports of poor results due to disease relapse with the use of intrallesional steroid injection, argon, and CO₂ laser vaporization. Radiation therapy has been used for refractory disease; however, only 50% of patients were able to wean off oral steroids after radiation treatment. Other therapeutic interventions, including retinoic acid, cytotoxic agents, and electrodessication and curettage, have had variable success rates.
Acknowledgment—The authors thank Cynda A. Johnson, MD, MBA, for her review of the manuscript. Dr. Daaleman is supported by the Robert Wood Johnson Foundation Generalist Faculty Scholars Program.

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