ABSTRACT

Educational Objective: To discuss the diagnosis and management of Kimura’s disease, which often presents as a mass in the head and neck. To describe the histopathological and laboratory findings in patients with Kimura’s disease and understand the treatment recommendations.

Objective: Eosinophilic hyperplastic lymphogranuloma, more commonly known as Kimura’s disease, is an inflammatory disorder most frequently found in middle-aged Asian men. It has a predilection for the head and neck region and presents with its own set of clinical characteristics and histologic features. The clinical findings, surgical management, and pathologic description of a patient with Kimura’s disease are presented.

Study design: case report and literature review.

Methods: The patient’s clinical records were examined for history, laboratory values, biopsy results, operative procedure, and pathologic diagnosis. A literature review for Kimura’s disease was also performed in PUBMED.

RESULTS: A fifty-two-year-old Chinese man presented with a left preauricular mass, which initially appeared to be of parotid origin. Laboratory values demonstrated eosinophilia, and fine-needle aspiration biopsy showed clusters of epithelioid histiocytes, lymphocytes, and foamy macrophages. He underwent surgical excision without complication. Pathologic examination demonstrated Kimura’s disease.

Conclusion: Although rare, Kimura’s disease has been reported in the literature originating from countries in Asia since 1937. Surgical excision is the treatment of choice, followed by steroid and/or other immunosuppressive therapy. As the population seeking medical care becomes more diverse, it is important for the otolaryngologist to include Kimura’s disease in formulating the differential diagnosis of head and neck masses.

INTRODUCTION

Eosinophilic hyperplastic lymphogranuloma (EHLG) was first described by Kimm and Szeto in 1957 and later renamed as Kimura’s disease in 1969. It is an inflammatory entity usually affecting middle-aged, East Asian males in the head and neck region. EHLG can often be confused with other lesions of the head and neck caused by inflammatory, congenital, infectious, traumatic, and neoplastic origins. This is a case report that illustrates the clinico-pathologic highlights of Kimura’s disease.

METHODS AND MATERIALS

The patient’s clinical chart was reviewed for history, laboratory values, biopsy results, operative procedure, and pathologic diagnosis. A literature review for Kimura’s disease was also performed in PUBMED.

RESULTS: The patient was a healthy 52-year-old Asian man with a two-year history of intermittent swelling of the left preauricular area. He sought evaluation in the outpatient setting and was noted to have a 2 cm x 3 cm mass of the left preauricular area [Figure 1]. The mass was firm, mobile and indistinguishable from the parotid gland. Laboratory values demonstrated eosinophilia, and fine-needle aspiration biopsy showed clusters of epithelioid histiocytes, lymphocytes, and foamy macrophages. He underwent surgical excision without complication. Pathologic examination demonstrated Kimura’s disease.

Conclusion: Although rare, Kimura’s disease has been reported in the literature originating from countries in Asia since 1937. Surgical excision is the treatment of choice, followed by steroid and/or other immunosuppressive therapy. As the population seeking medical care becomes more diverse, it is important for the otolaryngologist to include Kimura’s disease in formulating the differential diagnosis of head and neck masses.

DISCUSSION

Kimura’s disease is a rare, chronic inflammatory reaction causing swelling with a predilection for the head and neck region. Up to 58% of patients with EHLG have involvement of the parotid gland or the periparotid lymph nodes. Patients less commonly report symptoms of pruritus or overlying skin pigmentation. Kimura’s disease affects an overwhelmingly male majority, particularly of Chinese and Japanese descent, with an average age of 29.6 to 42 years.

Peripheral blood eosinophilia and elevated IgE levels complete the clinical profile of EHLG. Additionally, some patients with Kimura’s disease are also afflicted with nephrotic syndrome. Renal biopsies in these individuals demonstrate mesangial proliferation and tubulointerstitial infiltration by eosinophils.

Physical exam findings and laboratory data cannot distinguish Kimura’s disease from other causes of head and neck masses. Microscopic examination of surgical specimens is definitive. A constant, pathologic finding in Kimura’s disease is the increased presence of lymphoid follicles with active lymphogranuloma. Furthermore, these follicles are densely infiltrated by eosinophils, often leading to folliculolysis and formation of microabscesses. Vascular proliferation is a second prominent component of EHLG. Increased numbers of thin-walled, capillary vessels are found in proximity to the germinal centers. Another entity associated with eosinophilia and vascularization is angiolymphoid hyperplasia with eosinophilia (ALHE). ALHE has a similar clinical presentation, but there are important distinguishing histologic features. In ALHE, lymphoid changes are generally located in the dermis; microabscesses are not seen; and capillary vascularization produces thicker walls with hypertrophied or vacuolated endothelial cells that protrude into the lumen.

The preferred treatment for Kimura’s disease is surgical excision. This offers the advantage of a short treatment with few complications. Although a specific cause is not known, recent studies have demonstrated increased activation of T-helper cells with release of T-helper 2 cytokines in those with Kimura’s disease. Especially with patients who present concurrently with nephrotic syndrome, clinicians recommend 10-40 mg/day of prednisolone with subsequent tapering.

CONCLUSIONS

Kimura’s disease is an uncommon eosinophilic granulomatous reaction of unknown etiology primarily affecting East Asian males with an affinity for the head and neck region. The otolaryngologist should include EHLG when considering the differential diagnoses of eosinophilic lymphoid hyperplasia and parotid masses. Kimura’s disease can be differentiated pathologically, and treatment consists of a combination of surgery, medicine, and radiation.

REFERENCES