

Kimura's Disease- A Diagnostic Challenge

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Abstract

Kimura's disease is a rare, benign, chronic inflammatory disease with an unknown etiology, which predominantly affects Asian males. It has predilection for the head and neck region, and primarily involves the major salivary glands and is accompanied with loco-regional lymphadenopathy. It is therefore often misdiagnosed for a salivary gland tumor or lymphadenopathy secondary to metastases, lymphoma or tuberculosis. We present a case of Kimura's disease in a middle aged Chinese male patient manifesting as painless lumps over the left jaw and neck. Based on the CT scan, the differential diagnoses of lymphomatous/metastatic adenopathy affecting the left cervical and parotid nodes or a primary left parotid malignancy with ipsilateral metastatic adenopathy were considered. However, a fine needle aspiration biopsy of the left sided adenopathy yielded the diagnosis of Kimura's disease. The triad of subcutaneous masses in head and neck region, regional lymphadenopathy and masses in one or more of the major salivary glands should raise the suspicion of Kimura's disease.

Keywords: Kimura; Salivary glands; Regional lymphadenopathy; Inflammatory condition; Computed tomography

Introduction

Kimura's disease is an indolent, benign but locally disfiguring disease. Its true importance lies in its ability to mimic a number of benign inflammatory and neoplastic conditions of the head and neck [1-3]. Its protean manifestation is challenging to head and neck surgeons, radiologists and pathologists [4]. Since its initial description in 1937 by Kim and Szeto [5] followed by systematic description in 1948 by Kimura [6], Kimura's disease remains an enigmatic condition to this date.

Case Report

A 50-year-old Chinese male with multiple firm, painless lumps over the left jaw and neck, gradually increasing in size over a period of eighteen months, was referred for a computed tomography (CT) scan of the neck. The contrast-enhanced CT revealed a soft tissue density mass with ill-defined margins in the superficial lobe of the left parotid gland (Figure 1). Another ill-defined subcutaneous mass was identified over the left parotid lesion with CT attenuation similar to the left parotid lump (Figure 2). Enlarged well defined round to oval solid lymph nodes were seen in the ipsilateral neck, from levels II-V of the left cervical lymph node chain (Figure 3). The differential diagnoses included lymphomatous/metastatic adenopathy affecting the left cervical and parotid nodes or a primary left parotid malignancy with ipsilateral metastatic adenopathy. The patient underwent a fine needle aspiration (FNA) of the left sided cervical adenopathy and a final diagnosis of Kimura's Disease was made, based on classical cytological features. The patient was offered conservative management and is currently on follow up.

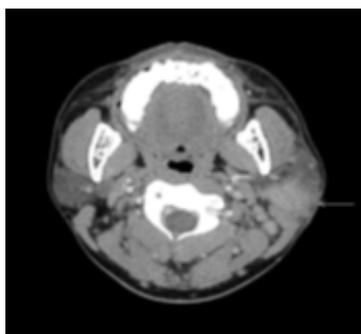


Figure 1: Axial image of the contrast enhanced CT reveals an enhancing mass with ill-defined margins noted in the superficial lobe of the left parotid gland.

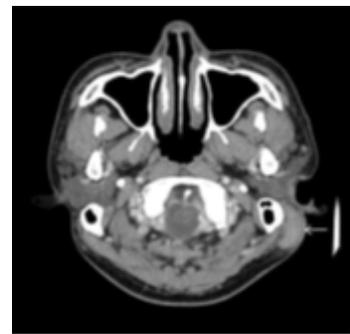


Figure 2: Axial image of the contrast enhanced CT at the level just inferior to the parotid gland reveals a well-defined mass with attenuation similar to the parotid mass in the subcutaneous region of the neck.



Figure 3: A sagittal reformatted CT image depicts enlarged enhancing cervical lymph nodes from levels II-V of the neck on left side. The left parotid mass lesion is also well seen.

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Discussion

Although typically described as a disease of the Far East, Kimura's disease has been reported in non- Oriental populations as well. It is predominantly seen in young middle-aged males, between the 2nd and 4th decades of life with reported male to female sex ratios between 3:1 to 10:1 [2-4]. The classical clinical presentation is with a triad of painless masses in the subcutaneous regions of the head and neck, in the major salivary glands and local lymphadenopathy [1,7]. Involvement of the oral cavity, axilla, groin, limbs and trunks have been described [7,8]. A high association with marked peripheral blood eosinophilia (10% to 70%) and elevated serum IgE levels (800-35,000 U/ml) suggests the possibility of an immunologically mediated disorder [8]. The disease is associated with systemic connective tissue diseases and nephrotic syndrome [1].

Histopathological findings include prominent germinal centers with fibrous, vascular and cellular proliferation; the cellular component consists of dense eosinophilic infiltrates in a background of abundant lymphocytes, plasma cells, eosinophilic micro-abscesses with central necrosis and Warthin-Finkeldey-type polykaryocytes. Increased postcapillary venules in para-cortex and sclerosis without arteriovenous shunts are seen. Immunofluorescence studies have revealed IgE deposits in follicle centres [4,8,9].

On ultrasound, the masses of Kimura's disease are predominantly solid and hypoechoic, with homogeneous or heterogeneous internal architecture and high internal vascularity. The affected lymph nodes are often spherical, well-defined, and hypoechoic with a preserved echogenic hilum [2]. On CT scan, various imaging appearances have been described. Gopinathan et al. [7] in their study of 13 cases with Kimura's disease described two specific morphologic patterns in salivary glands, namely (1) Type I and (2) Type II masses. In their study, Type 1 masses had a well-defined nodular configuration, while Type 2 masses were ill-defined with plaque like configuration related to salivary glands, as was seen in our patient. Type I lesions were seen in a younger age group and enhanced avidly on post contrast examinations while Type II lesions were noted in an older age group and did not demonstrate impressive enhancement. These features prompted them to suspect that they represent the two opposite ends of the spectrum in an inflammatory disease continuum. Lim et al. [3] also described a similar temporal progression of findings in the same patient and attributed this phenomenon to progressive fibrosis and sclerosis around the post-capillary venules, which eventually obliterate and result in diminished enhancement on contrast-enhanced imaging.

Involvement of lymph nodes is reported to be in the 42% to 100% range across the literature [7]. The enlarged lymph nodes are generally well-defined, round to oval in shape and may depict marked enhancement. In our case, unilateral lymphadenopathy from levels II-V was noted which led us to suspect more sinister diagnoses like metastases or a lymphoproliferative disease.

On imaging, Kimura's disease essentially should be a diagnosis of exclusion. Salivary gland neoplasms, ALHE (angiolymphoid hyperplasia with eosinophilia) and lymphadenopathy due to lymphoma, metastases (from breast, colorectal and nasopharyngeal carcinoma) and tuberculosis should be considered before the possibility of Kimura's

disease is proposed [1,7]. Primary salivary gland tumours are usually well-encapsulated and not associated with subcutaneous extension, as is usually seen in Kimura's disease. ALHE lacks racial predilection and is more common in middle aged women. Association of ALHE with lymphadenopathy or peripheral eosinophilia is rare [4,10]. The affected lymph nodes in tuberculosis typically demonstrate necrotic centers. Lymphomatous and metastatic adenopathy are difficult to differentiate from Kimura's disease based on imaging findings alone, especially if subcutaneous and/or salivary gland masses are not present in the latter. However, a long clinical course, as is seen in Kimura's disease, is unusual in these conditions [7,8].

Conclusion

The treatment options for Kimura's disease are varied and not well established [1]. Surgical excision, steroids, radiotherapy, cryotherapy and laser fulguration are utilized to varying success. Steroids provide transient relief and the disease often recurs on cessation. Local irradiation is effective in a majority of cases [1]. Surgical excision is often associated with recurrence of the disease [10,11].

The triad of subcutaneous masses in the head and neck region, regional lymphadenopathy, and masses in one or more of the major salivary glands should raise the suspicion of Kimura's disease. However, more common sinister conditions like salivary gland neoplasm and neoplastic/infective adenopathy should be meticulously excluded. The coexistence of peripheral blood eosinophilia and raised IgE levels in these patients may help to further strengthen the diagnosis although histopathological confirmation is usually required.

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