

Case report:

Kimura Disease of Parotid Gland

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Abstract:

Kimura disease is a rare, benign, chronic inflammatory disorder. Characteristic features of the disease include painless subcutaneous mass in the head and neck region, elevated serum eosinophilia and markedly raised serum immunoglobulin E. Herein, we report a case of Kimura disease in a patient who presented with a parotid mass and received both medical and surgical management. A course of high dose oral prednisolone and subsequently parotidectomy were the important elements of the management.

Keywords: Kimura disease; Parotid mass; Lymphadenopathy

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Introduction

Kimura disease is a rare, chronic inflammatory disorder of uncertain origin. It was named after Kimura et al. who published a systematic description of the disease in 1948¹. It commonly affects Oriental male population in their second and third decades of life^{2,3}. The disease is characterized by a triad of painless subcutaneous mass in the head and neck region, elevated serum eosinophilia and markedly raised serum immunoglobulin E (IgE)¹. The optimal treatment for Kimura disease has been a subject of debates for many years and still remains controversial.

Case report

A 33-year-old gentleman presented with left parotid swelling which was gradually increasing in size for the past 13 years. It was painless and not associated with facial asymmetry. His mastication was normal. There was no dysphagia, dryness of mouth, ear symptoms or other neck swelling. He

has no other constitutional symptoms. Physical examination revealed a left parotid swelling measuring 12 cm x 10 cm which was firm, non-tender and mobile. It extends superiorly up to upper attachment of auricle and inferiorly to left level II region. Anteriorly, the mass extends to mid left cheek and up to left preauricular region posteriorly (Fig. 1). There were no palpable neck lymph nodes or no facial weakness. Flexible nasoendoscopy was unremarkable.

Laboratory investigations showed significant raised serum of IgE > 5000 kU/L, elevated serum eosinophil protein of > 200 microgram/L and raised ESR of 32mm/hour. However, the leukocyte counts were within normal range. Serum creatinine level were normal.

A magnetic resonance imaging (MRI) were done which showed a large left parotid mass lesion with multiple cervical and supraclavicular lymphadenopathy (Fig. 2). CT scan was done

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Figure 1 (a, b): Anterior and lateral views of the enlarged right parotid gland which extends from left preauricular region to level II

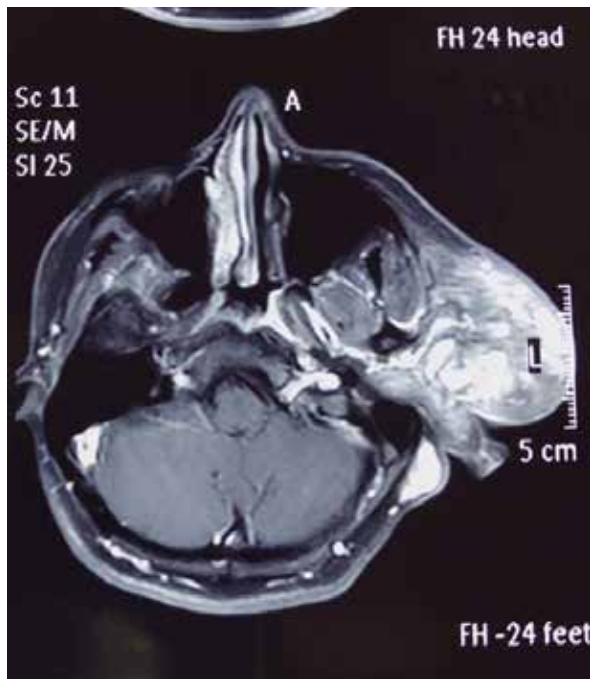


Figure 2: A contrast-enhanced MRI axial view at the level of inferior turbinate showed a well-defined heterogenous lobulated soft tissue mass with its epicentre seen in left superficial lobe of parotid gland

initially and it showed a vascular lesion. Features are suggestive of left parotid primary tumor with nodal metastasis.

FNAC was requested, but it was inconclusive. We then proceeded with left parotid biopsy which

showed features that are consistent with Kimura disease. CTA carotid artery was done and revealed a vascular tumor with arterial supply from branches of the external carotid artery.

Accordingly, the patient was started on high dose oral prednisolone (60 mg) for 2 weeks. The dose was gradually tapered to 30 mg for two weeks and subsequently reduced to 20 mg twice daily for two weeks. After 6 weeks of steroid therapy, the response was good with significant reduction in size of the left parotid swelling to 6 cm x 6 cm.

He underwent left superficial parotidectomy. Intraoperatively, the parotid gland is densely fibrosed and adherent to subcutaneous tissue. Facial nerve was preserved. Post operatively, there was no facial nerve palsy and was restarted back on tapering dose of oral prednisolone 60 mg daily. Histopathological study of the specimen shows normal parotid tissue and numerous lymphoid infiltrates in the subcutaneous tissue. These lymphoid infiltrates show marked hyperplastic and reactive lymphoid follicles with germinal centers formation accompanied by large number of eosinophils, lymphocytes, some plasma cells and mast cells. Eosinophilia with eosinophilic micro abscess formation are seen. Small-sized capillary hyperplasia is also evident. In areas, stroma and perivascular sclerosis are noted. These lymphoid infiltrated are seen at the periphery margin in focal areas. All these histological features are consistent with Kimura disease.

Discussion

The typical presentation of Kimura disease is a non-tender, subcutaneous mass involving the head and neck, predominantly in parotid and submandibular region. Other unusual sites include the limbs, groin, trunk and scalp, and is often associated with regional lymphadenopathy^{4,7}. The lesions gradually increase in size over months or years and spontaneous involution is rare. Kimura disease has a good prognosis. It is a benign disease and malignant transformation has not been documented⁶.

Kimura disease is commonly associated with renal disease, with an incidence rate of 10% to 60%⁸. The most common associated renal disease reported is membranous glomerulonephritis⁹. Immunocomplex-mediated damage or Th2-dominant immune response disorders have been speculated to be the cause of renal impairment⁸. Other associated diseases include autoimmune diseases such as ulcerative colitis⁹ and dermatological problems such as lichen amyloidosis³. Our patient had no renal nor autoimmune disease involvement.

Establishing a diagnosis in Kimura disease might be challenging. Differential diagnosis includes angiolymphoid hyperplasia with eosinophilia (ALHE), Kaposi sarcoma, Sjogren syndrome with parotid involvement, Hodgkin disease, tuberculosis, nodal metastasis, Warthin tumor and low grade angiosarcoma⁷. Characteristic laboratory findings that may facilitate the diagnosis are marked elevation of peripheral blood eosinophils and serum IgE levels^{5,7}. It has been postulated that the size of the lesion may be correlated with the degree of blood eosinophilia, which might be useful to measure the disease activity⁵. Our patient's blood investigations showed both increase in peripheral blood eosinophils and serum IgE.

CT or MRI are useful in aiding the diagnosis of Kimura disease. It helps to distinguish Kimura disease from other conditions, delineate the extension and progression of the mass as well as identifying lymph node involvement^{5,8}. On contrast-enhanced CT, the lesions appear to have ill-defined borders with adjacent enhancing

cervical lymphadenopathy. These lesions also demonstrate intense enhancement which reflects the vascular nature of the lesions. On MRI, tissues involved with Kimura disease show intermediate to high signal intensities on T1-weighted images and hyperintense signals on T2-weighted images⁵. The CT and MRI findings in our patients were compatible with Kimura disease.

Histological examination of the excised lesion is considered as the best method to achieve exact diagnosis of Kimura disease^{7,8}. The histological features in this disease consist of lymphocytic inflammatory infiltrate, forming lymphoid follicles mixed with aggregates of eosinophils and varying degrees of fibrosis in a richly vascular stroma. The inflammatory infiltrates occupy the layers of reticular dermis, subcutaneous tissue, and sometimes, extend to the muscle fascia and the skeletal muscle².

The exact pathogenesis of Kimura disease remains unclear. One of the most widely accepted etiological factor is reactive immune disorder based on the presence of peripheral eosinophils, increased mast cells and increased levels of interleukin (IL)-5 and IgE, which imply an abnormal T cell stimulation similar to a hypersensitivity-type reaction⁴. Other potential cause includes Epstein-Barr virus, human herpes virus-8 and parasitic infection⁵.

There is no consensus yet regarding the optimal treatment for Kimura disease. In asymptomatic cases, conservative observation has been advocated by previous authors^{5,7}. Therapeutic options can be either surgical resection, steroid therapy, cytotoxic therapy and radiotherapy^{4,9}. Medications such as oral retinoids, pentoxifylline, and azathioprine have also been used for the treatment⁴. The preferred treatment of choice in symptomatic cases is surgical excision⁵. However, the recurrence rate of cases treated with surgical excision alone may be as high as 25%⁶. Steroids administration has been documented to arrests progression of the disease considerably⁴. Unfortunately, there is also tendency for lesions to recur once steroid therapy is stopped⁶. Radiotherapy can be considered for recalcitrant cases or lesions not suitable to surgery due to size or unacceptable resultant morbidity⁶.

Our patient had a cosmetically disfiguring parotid swelling which was huge and unlikely to resolve completely with medical therapy. Thus, the option for surgical resection was chosen. He was initially started with tapering dose of oral prednisolone for 6 weeks to reduce the size of the parotid mass and ease the operation process. Post-operatively, he was restarted on oral prednisolone to prevent recurrence.

Conclusion

Kimura disease is a rare disease and high index of suspicion is required to avoid missing the diagnosis. It should be considered as differential diagnosis in Oriental patients presenting with head and neck masses especially if it is associated with raised peripheral blood eosinophilia and serum IgE. Surgeons are also advocated to rule

out renal disease in view of its strong association with Kimura disease. There is a wide range of treatment choices and it depends on the symptoms and patient's expectation. Combination of surgery and oral steroid may be a good choice to reduce the swelling size and prevent recurrence.

Conflict of Interest

No conflict of interest has been disclosed by the authors.

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Authors Contributions

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Collection and assembly of data: MHAG, AO

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