

Rare Presentation of Kimura's Disease Involving The Parotid Gland in An Indian Male: A Case Report

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Abstract: Kimura's disease (KD) is a rare chronic inflammatory disorder that involves the subcutaneous tissues and occurs predominantly in the head and neck region, frequently associated with regional lymphadenopathy or salivary gland involvement. Although, not an uncommon condition in the Asian countries, it is quite rare among the occidental civilization and in the western countries is more commonly diagnosed among Asian migrants. Kimura's disease is sometimes confused with angiolymphoid hyperplasia with eosinophilia, which occurs in the superficial skin of the head and neck region. The classical features of angiolymphoid hyperplasia with eosinophilia are characterized by a triad of painless subcutaneous masses in the head and neck region, blood and tissue eosinophilia, and markedly elevated immunoglobulin E (IgE) levels. Here, we report a case of a 33-year-old Indian male with KD who presented with unilateral nodular swelling in the left parotid region. The diagnosis was based on characteristic histopathologic findings in conjunction with peripheral eosinophilia and elevated serum IgE levels.

Keywords: Angiolymphoid hyperplasia with eosinophilia, Kimura's disease.

1. Introduction

Kimura's disease (KD) was initially diagnosed by Kim and Szeto in 1937 in the Chinese literature as 'eosinophilic hyperplastic lymphogranuloma'.¹ The definitive histologic description was published by Kimura et al in 1948, which they termed 'Kimura's disease'.² Kimura's disease is an inflammatory disorder of unknown etiology that tends to occur among Asians, chiefly in Chinese and Japanese populations. Recent studies have shown that KD occasionally shows a clonal proliferation of T-cells.³ The disease commonly involves the dermis and subcutaneous tissue characterized by one or multiple nodules and asymptomatic but sometimes painful masses may be present. Previous studies have noted that parotid tissues are often involved in KD and clinically misdiagnosed as a salivary gland swelling or even as malignancy.⁴ Clinically, KD can be confused with a lymphoma or an infectious process but a definitive diagnosis can be established on biopsy

2. Case Report

- 33 years old man hailing from Mysuru reported to the JSS hospital with a chief complaints of slow – growing swelling on the left side of the face since 3 years. The swelling started as a small nodule and gradually increased to the size of 6 x 4 cm on presentation with no history of any constitutional signs and symptoms.
- Local examination – the swelling extended from tragus to one cm below angle of mandible superoinferiorly and 4 cm anteroposterior direction. No local rise of temperature and changes in the overlying skin. No signs of involvement of facial nerve. No increase in the size of swelling observed during food intake. Preauricular firm lymph nodes were palpable.

Investigations

Differential leukocyte count was showing 26 percents of eosinophils.

Peripheral blood smear revealed eosinophilia.

Urine routine suggesting nil urine albumin level.

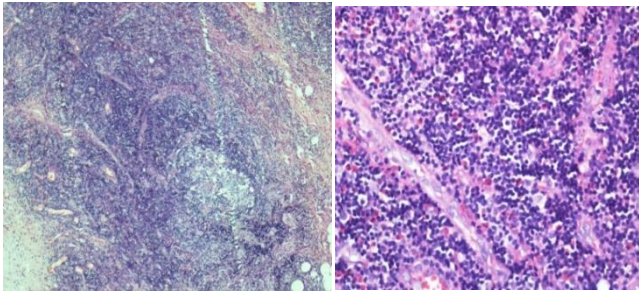
FNAC Parotid gland showed reactive hyperplasia

Management and HPE report

- Superficial conservative parotidectomy along with enlarged lymph node excision done under GA. Specimen sized 7 x 5 x 2 cm, brownish black colour, firm in consistency, rough surface with irregular borders.
 - HPE revealed parotid gland with areas of fibrosis, lymphoid follicles, dense infiltration by eosinophils forming abscess at places and vascular proliferation.
- Features of those of Kimura disease.**



Clinical photograph showing a swelling in the Left parotid region.



Photomicrograph of the histopathologic section of the lymph node showing fibrous connective tissue septa separating germinal centers

3. Discussion

Kimura's disease first described in China by Kim and Szeto in 1937 under the term 'eosinophilic hyperplastic lymphogranuloma'¹ is currently more widely known as KD after a systematic description published in 1948 by Kimura et al.² It is commonly seen in the young and middle aged Asian males of Chinese and Japanese population.⁵ The characteristic feature of this disease is its typical presentation as painless subcutaneous masses with adenopathy in the head and neck region. The disease usually involves subcutaneous tissues about the parotid and submandibular glands with regional lymphadenopathy (mainly periauricular, axillary or the inguinal group) and the oral mucosa is rarely affected.⁶ Kimura's disease involving the auricle⁷ as well as the scalp and orbit⁸ has been reported. This case manifests as a unilateral asymptomatic swelling of the parotid gland and a similar presentation has been reported by Tham et al.¹⁰ Kimura's disease is a benign condition and is usually self-limiting, occasionally presenting with renal involvement with nephrotic syndrome being the most common associated with this disease. Proteinuria may occur in 12–16% of the cases.¹⁰ However, in our case, renal function tests were within normal limits with no evidence of proteinuria. The characteristic histopathologic feature of KD is the presence of prominent germinal centers in the involved lymph nodes containing cellular, vascular, and fibrous components. The cellular component consists of dense eosinophilic infiltrates in a background of abundant lymphocytes and plasma cells, eosinophilic microabscesses with central necrosis⁶ Other features are the proliferation of small blood vessels, many of which are lined by enlarged endothelial cells with uniform ovoid nuclei and intracytoplasmic vacuoles. These endothelial cells have been described as having a cobblestone appearance. In addition, perivascular and interstitial infiltrate composed primarily of lymphocytes and eosinophils are also seen.⁶ Clinical differential diagnosis includes malignant lymphoma, parotid tumors, hemangioma, pyogenic granuloma, Mikulicz's disease, and Kikuchi's disease.⁷ Other conditions to be considered in the differential diagnosis are Kaposi's sarcoma, angiosarcoma, eosinophilic lymphoma and angioimmunoblastic lymphadenopathy, and various parasitic diseases which are accountable for subcutaneous masses with an associated lymphadenopathy such as tissue invasive helminthes infections, cysticercosis, sparganosis, and toxocariasis. Imaging studies including computed tomography and

magnetic resonance imaging scans can be helpful to delineate the extent of the disease.⁸ The difficulty in distinguishing KD from angiolymphoid hyperplasia with eosinophilia (ALHE) is due to certain similar clinical and histopathologic features. Common histopathologic characteristics of ALHE and KD are as follows. In both the dermis and subcutaneous tissues are involved with an inflammatory infiltrate composed of lymphocytes and eosinophils and blood vessels with endothelial cells that may be protruded with abundant cytoplasm. Both may present with fibroplasia with a plasma cell infiltrate. However, the epithelial and nonepithelial adnexal structures may be spared.⁴ The treatment of KD is variable, with surgical excision being the obvious choice. However, the lesion has a tendency to recur. Other therapeutic options such as radiation, systemic corticosteroids, and cytotoxic agents have been tried with inconsistent results. The management of KD differs with the condition of the patient with surgical, medical, or radiotherapy being the line of treatment.⁹ Surgery should be contemplated in younger patients with localized primary lesions or in localized recurrences. In patients with recurrent disease with renal involvement both intralesional and oral steroids, cyclophosphamide and anti-platelet drugs have been used with reasonable success rates. However, radiotherapy may be indicated for unresectable masses and recurrent masses unresponsive to medical therapy.

4. Conclusion

This is a rare case of KD in an Indian male patient affecting the parotid gland, clinically simulating chronic sialadenitis. With poor socio-economic conditions and a rapid increase in HIV infections, KD should be considered in the differential diagnosis in patients who present with primary lymphadenopathy and eosinophilia. The purpose of this article is to emphasize the importance of the knowledge regarding KD among the clinicians and the pathologists since it can mimic various benign inflammatory and neoplastic conditions in the head and neck region leading to misdiagnosis and mismanagement.

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