Kimura’s Disease: A case report
Shilpa.C.Patel, Vishal.R. Dave

Abstract
Kimura’s disease is a rare inflammatory disorder of unknown cause, primarily seen in young Asian males. The disease is characterized by a triad of painless subcutaneous masses in head and neck region, blood and tissue eosinophilia and moderately elevated serum immunoglobulin E levels. Here, we present a case report of this rare entity of unusual age presentation with classical histopathological features.

Key Words: Kimura’s Disease; Eosinophilia; Parotid Gland; Lymphadenopathy

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Introduction
Kimura’s disease (KD) is a rare chronic angiolymphoid proliferative soft tissue disorder of unknown origin.(1) It occurs most often in young and middle aged Asian male. It is most common in head and neck region, with a predilection for preauricular area. Typical clinical presentations are painless subcutaneous masses, regional lymph node enlargement, blood and tissue hyper eosinophilia, and markedly elevated serum IgE levels.(1) According to the previous medical literatures, Kimura’s disease has a high recurrence rate so early & definitive diagnosis of the disease is vital for effective treatment plan.(1) Here we report a case of Kimura’s disease with clinico-pathological differential diagnosis.

Case Report
A 65 year old man reported to outpatient department with chief complaint of painless swelling on right side of face in parotid region since 4 months but it was painful on mastication. Here we summarized details of present case in the following Table-1. With a provisional diagnosis of salivary gland neoplasm, an incisional biopsy was performed. The section stained with Haematoxylin & Eosin revealed unencapsulated ill-defined lesion with lymphoid follicle formation, fibrosis and chronic inflammatory cell infiltration with predominant eosinophils (Figure 1a, 1b, 1c, 1d). Inter follicular areas show increased number of venules with endothelial cell proliferation. In some areas lymphoid follicle were infiltrated by eosinophils with resultant eosinophilic micro abscesses. With a diagnosis of Kimura’s disease, the patient was undergone superficial lobe parotidectomy with removal of multiple regional lymph nodes with vicinity.

The excisional biopsy specimen received was firm nodular soft tissue measuring approximately 4.2X2X1.5 cm³ and two in number. The histopathological feature of excisional biopsy was matched with that of incisional one. The patient was supplemented with oral Dexamethasone for total duration of 6 weeks (0.5 mg twice daily for 1 week and then tapered over 5 weeks). The patient was apparently normal without any local recurrence & systemic involvement within 6 month of follow-up period.

Discussion
Kimura’s disease first described in1937 in Chinese literature by H.T.Kimm and C.Szeto and they termed it as “eosinophilic hyperplastic lymphogranuloma”.(2) The disease became widely known as Kimura’s disease after Kimura and colleagues reported two cases of unusual granulation combined with hyperplastic changes of lymphoid tissue.(1) The etiopathogenesis of KD remains unknown and it is considered nowadays as an allergic disease and it seems to be a systemic immunological disorder. Eosinophilia and increased serum IgE levels make KD be considered a CD4 (+) T helper 2 (Th2) allergic reactions. Th2 cells would produce interleukins (IL) IL-4, IL-5 and IL-13, which, in turn, would act in B cells favoring the production of antigen-specific IgE. Th2 cell proliferation and the overexpression of cytokines would play an essential role in the development of the disease.(3)

It occurs predominantly as a unilateral manifestation in the head and neck and it is frequently associated with regional lymphadenopathy with or without involvement of salivary glands.(4) In the present case, e presence unilateral parotid gland swelling with regional lymphadenopathy led to the clinical diagnostic dilemma of other pathologic entities affecting salivary glands & lymph nodes. The differential diagnosis of Kimuras disease frequently in previous literatures are
Lymphomas, salivary gland neoplasms, benign lymphoepithelial lesions (BLL / Mikulicz’s disease), Angiolymphoid hyperplasia with eosinophilia (ALHE/ Epitheloid hemangiomma) and angioimmunoblastic lymphadenopathy (AIL). Constant classical features of KD include numerous lymphoid follicles, mixed inflammatory infiltrate composed mainly of eosinophils and increased amount of post capillary venules.(5)

<table>
<thead>
<tr>
<th>Age/Sex</th>
<th>location</th>
<th>Physical findings</th>
<th>lymphadenopathy</th>
<th>Systemic involvement</th>
<th>Laboratory investigation</th>
</tr>
</thead>
<tbody>
<tr>
<td>65/Male</td>
<td>Parotid (right)</td>
<td>3.5X3.5 cm firm, not tender &amp; normal overlying skin</td>
<td>Regional lymphadenopathy</td>
<td>No renal involvement</td>
<td>Eosinophilia (36%) Elevated erythrocyte sedimentation rate (ESR) Raised IgE levels</td>
</tr>
</tbody>
</table>

Table 1: clinical & investigation details of present case

Figure 1a. Lymphocytic infiltration with follicle formation and fibrosis (4x), 1b. Well defined lymphoid follicle with prominent germinal centre, (10x) 1c. Chronic Inflammatory cell infiltration with predominant eosinophils with microabscesses.(40x), 1d. Interfollicular areas showing mixed inflammation with prominent eosinophils and proliferation of venules.(40x), 1e. Interfollicular areas showing increased number of venules with endothelial cell proliferation.(40x)

Till date there is no definite treatment for KD is well established.(2) However the treatment plan should aim at preservation of vital structures associated with the lesions & cosmetic rehabilitation, while preventing recurrences. Treatment options range from observation & follow-up of mild & symptomatic cases to conservative surgical approach, medication & radiotherapy in symptomatic & recurrent cases. If the lesion is primary, localized or present in young age then surgical approach is more preferable. According to previous literature, only surgical approach had high incidence of recurrence.(1) If the lesion is recurrent with systemic involvement, application of medication like corticosteroid and immunosuppressive agents have been shown to decrease size of the lesion.(2) Irradiation should be considered in patients resistant to the steroid or to prevent the patient from deleterious effect of long term use of steroid.(5)

**Conclusion**

In conclusion this paper draws attention on such a rare chronic inflammatory disease which mimics neoplastic conditions. This disease should be considered in differential diagnosis of patients presented with head & neck mass and lymphadenopathy and investigated accordingly as this disease has good prognosis. Knowledge of signature features of Kimura’s disease put the physicians in a better position to evaluate its clinical outcome and optimal treatment regimen.

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**References**


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