Case report

Kimura Disease: A case report and review of literature
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Abstract
Kimura disease is a rare chronic inflammatory disease with angiolymphatic proliferation of unknown etiology predominantly seen among young Asian males. It classically shows a triad of non-tender subcutaneous masses predominantly in head and neck region with tissue and blood eosinophilia and raised serum IgE level. Here we present a case report of a 24 years male with bilateral pre-auricular and post-auricular swelling for 6 years. The diagnosis of Kimura disease was made on the basis of clinical and histopathological examination.

Keywords: Kimura disease; head neck swelling; lymphadenopathy

Introduction
Kimura disease (KD) is a rare chronic inflammatory angiolymphoid proliferative soft tissue disorder of unknown origin.1 It most commonly occurs in young Asian males.1 The male: female ratio is 3.5:1.2 Typical clinical presentation is characterized by a triad of painless cervical adenopathy or subcutaneous masses predominantly in the head or neck region, blood and tissue eosinophilia, and markedly elevated serum immunoglobulin E (IgE) levels.1 Histopathologically the disease is characterised by hyperplasia of the lymphoid tissue with well-developed lymphoid follicles, marked lymphocyte (eosinophil) infiltration, proliferation of thin walled capillary venules and varying degree of fibrosis.3

Case report
A 24 years male from sub-Himalayan belt presented to surgical OPD with bilateral pre and post-auricular gradually increasing painless swellings [FIG.1] associated with occasional itching over the area for past six years. There was no history of fever, weight loss, tuberculosis, allergy, drug intake, skin lesion, respiratory problems, urinary or gastrointestinal symptoms.

On examination the patient had non tender discrete mobile firm palpable bilateral pre and post–auricular lymph nodes with largest measuring 2.5x2.5 cm located in left pre-auricular area. The patient also had single 3x3 cm lymph node in the central group in left axilla. There was no hepatosplenomegally. Total leukocyte count was 8600 per cubic mm and absolute eosinophil count was 1290 per cmm. Serum IgE level was 4800 IU per ml (reference range 5-100 IU per ml). Routine urine examination and renal function was normal. The patient was negative for HIV I&II. Chest X-ray and abdominal ultrasonography were normal.

FNAC from left pre-auricular and right post-auricular swellings showed polymorphous lymphoid population in different stages of maturation [FIG.2A]. Then excision biopsy from right post-auricular lymph node (1.5x1.4x0.9 cm) revealed angiofollicular hyperplasia with eosinophilia consistent with KD [FIG.2 B,C].

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The patient was treated with antihistaminic (Cetirizine 10mg OD x 2 months) with partial response followed by oral corticosteroids (Prednisolone 30 mg/day x 5 months then gradually tapered over 6 weeks) which led to regression of swelling and patient remained symptom free for 6 months in follow up.

Discussion

KD was first described in 1937 by H.T. Kim and C. Szeto as “Eosinophilic hyperplastic lymphogranuloma”. The name Kimura’s disease was coined following a detailed description of this entity by Kimura et al in 1948, who referred to it as “an unusual granulation combined with hyperplastic changes in lymphoid tissue”. KD mainly affects the young and middle aged Asian males but also reported to occur in non-Asians. The clinical presentation is characterized by triad of painless unilateral cervical lymphadenopathy or subcutaneous swelling, eosinophilia (98%) and markedly elevated serum IgE levels in fewer cases. It most commonly presents with painless, sometimes pruritic, subcutaneous masses in the head and neck region (70%) but rarely involves extremities (12%) and trunk (3%). It may involve axillary and inguinal (15%) lymph nodes, parotid and submandibular glands and very rarely auricle, scalp, orbit, nerves, hard palate, larynx, mediastinal lymph nodes and spermatic cord. KD is primarily an unilateral process but bilateral involvement is documented in literature. Our case is a young Asian male presenting with the classical triad but bilateral involvement and axillary lymphadenopathy present in our case is a relatively rare presentation.

KD is associated with bronchial asthma less frequently with ulcerative colitis. According to previous literature, 12–16% of patients with KD have proteinuria, among whom, 62–79% reach the nephrotic range. Membranous glomerulonephritis is the most common renal lesion reported in patients with KD with others being minimal-change nephrotic syndrome, mesangioproliferative glomerulonephritis, focal segmental glomerulosclerosis and IgA nephropathy. There is no systemic involvement in the present case.

The etiology of KD is still unknown. Although infectious etiologies (human herpesvirus-8, Epstein-Barr virus) have been postulated, it is now believed to be related to an autoimmune or a delayed hypersensitivity reaction. An aberrant allergic response is further supported by the association of the disease with asthma, allergic rhinitis, atopic dermatitis, and peripheral hypereosinophilia, as well as raised serum IgE levels. It is now proposed that Kimura disease is a CD4(+)T helper 2(Th2) allergic reaction due to its raised serum IgE level. Th2 cell produces interleukin IL-4, IL-5, IL-13 which act on B cells which in turn produce antigen specific IgE. Th2 proliferation and the over-expression of cytokines would play an essential role in the development of the disease.

The diagnosis of KD is quite difficult. Imaging studies (USG, CT scan, MRI) show iso or hyper-dense heterogeneous lesions to aid in the diagnosis and extent of lymph node and soft tissue involvement. FNAC can provide initial clue but definitive diagnosis can only be obtained by histopathological examination.

Histologically KD classically shows numerous lymphoid follicles, mixed inflammatory infiltrate composed mainly of eosinophils, increased amount of post capillary venules and variable fibrosis. Considerable confusion was for years between KD and angiolymphoid hyperplasia with eosinophilia (ALHE). In spite of many similarities they are now considered as separate entities with distinctive clinical and histological features. In KD the localization of masses is subcutaneous (nodule) but in ALHE it’s dermal (papule). Lymphadenopathy, salivary gland involvement, eosinophilia and elevated IgE are frequent in KD and rare in ALHE. The predominant cellular component is lymphoid infiltrate in KD while it is vascular hyperplasia in ALHE. Differential diagnosis of KD include conditions like tuberculosis, metastasis, dermatofibrosarcoma protuberans, kaposi sarcoma, salivary gland neoplasm, Langerhans cell histiocytosis, AHLE and angioimmunoblastic lymphadenopathy. Mild to moderate cases may be tried with follow up, anti-allergic drugs and thereafter corticosteroids. Corticosteroid is effective partially and recurrence is common after discontinuation of therapy. They may be used to shrink the lesion before preparation for surgery, particularly surgery in head and neck region. Surgical resection also provides histological diagnosis but complete resection in head and neck region is quite difficult due to subsequent neurological deficit and facial reconstruction and variable chance of recurrence. But, radiotherapy and immunosuppressive agents (Cyclosporine, cyclophosphamide and mycophenolate mofetil / MMF) is more effective in such cases but as not be very appropriate for a benign condition, should be reserved for cases not responding to others.

KD is essentially a benign lesion with no documental
support for malignant transformation.\(^3\) Outcome is good but variable with spontaneous disappearance, waxing and waning, variable response to present therapeutic options and relapse after discontinuation of therapy.\(^3\)

**Conclusion**

This paper is intended to draw attention to a rare chronic inflammatory disease which presents with symptoms and signs that are commonly encountered in practice and which mimics many inflammatory and neoplastic conditions. But KD should always be kept in mind in case of head and neck swelling as early diagnosis has a good prognostic outcome and avoids wandering after many tests.

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**References**