Case Report

A rare cause of bilateral postaural lymphadenopathy: Kimura’s disease

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INTRODUCTION

Kimura’s disease (KD) is a chronic inflammatory disorder with angiolymphatic proliferation of unknown etiology, usually affecting young men of Asian race but is rare in other races. Affected Indians are very rare. Herein, we present an atypical manifestation of Kimura’s disease occurring in an Indian male who presented with bilateral postaural lymphadenopathy. The diagnosis of Kimura’s disease can be very difficult and misleading; it is important not to ignore histopathological features.

Keywords: Kimura’s disease, Lymphadenopathy, Eosinophilia

ABSTRACT

Kimura’s disease (KD) is a chronic inflammatory disorder with angiolymphatic proliferation of unknown etiology, usually affecting young men of Asian race but is rare in other races. Affected Indians are very rare. Herein, we present an atypical manifestation of Kimura’s disease occurring in an Indian male who presented with bilateral postaural lymphadenopathy. The diagnosis of Kimura’s disease can be very difficult and misleading; it is important not to ignore histopathological features.

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INTRODUCTION

Kimura’s disease was first reported in Chinese literature in 1937 as “eosinophilic hyperplastic lymphogranuloma”. The disease became widely known only in 1948 when it was reported in Japanese literature by Kimura and Ishikawa and came to be known as Kimura’s disease.1 It is a chronic inflammatory condition of unknown etiology presenting as multiple solitary subcutaneous nodules localized mostly in the region of head and neck with coexisting lymphadenopathy. The disease is characterized by angiolymphoid proliferation with peripheral eosinophilia and elevated serum IgE. This rare disease almost exclusively affects Asian males in their 2nd to 4th decade of life.2,3

One such case of Kimura’s disease, which is uncommon in Indian population, is reported.

CASE REPORT

Sixteen year old male presented with insidious onset bilateral postaural multiple swellings progressively increasing in size over past six years. The patient has no history of ear discharge, seborrheic dermatitis, swelling in other part of body or other constitutional symptoms.

On physical examination, a 5×4 cm swelling was present on right postaural region (Figure 1) and 5×3 cm swelling on left postaural region (Figure 1). There was another swelling of size 2×2 cm present on left side just posterior to aforementioned swelling. The swellings were non tender, firm and not fixed to overlying skin. The skin over the swelling was normal. There was no cervical, axillary or inguinal lymphadenopathy.

Ultrasonography (USG) examination showed heterogenous lesions in bilateral postaural region of size 4×3 cm on right side, 4×3 cm and 1.5×1.5 cm on left side showing fatty hilum with no vascularity likely lymphadenopathy. USG of neck showed no enlarged lymph nodes and abdominal USG showed normal kidneys.

Hematological investigation revealed eosinophilia (32%) with absolute eosinophil count of 1400 cells/mm³ and
elevated serum IgE levels (2500 IU/ml). Fine needle aspiration cytology showed reactive hyperplasia with eosinophils.

Figure 1: Pre-operative photograph.

Figure 2: Large quantity of eosinophilic granulocytes (H&E staining, microscopic magnification of ×40).

So, excisional biopsy of right sided swelling was performed. Histopathology revealed a 4×2.5 cm lymph node showing marked follicular hyperplasia with eosinophilic infiltrate in interfollicular zones (Figure 2). These features were consistent with the diagnosis of Kimura’s disease. The patient was then started on low dose steroids (30 mg/day) for three months. The swellings decreased in size in initial one month and stopped responding thereafter and left sided swellings were excised. The patient is under follow-up in outpatient department and has been asymptomatic for 12 months.

DISCUSSION

Kimura’s disease was described as “eosinophilic hyperplastic lymphogranuloma” by Kim and Szetu in 1937 but it was only in 1948 that it was named as Kimura’s disease after a detailed description by Kimura et al. It is a rare, benign, chronic inflammatory disorder of unknown etiology. A number of theories have been suggested for the origin of Kimura’s disease, including impairment of immune regulation, atopic reaction to a persistent antigenic stimulus by certain parasites, virus and neoplasm. This hypothesis is supported by eosinophilia and increased IgE in the peripheral blood. The most interesting hypothesis suggests Candida acting as a source of persistent antigenaemia, although neither hyphae nor spores have been isolated.

It predominantly manifests as a painless mass in head and neck region, commonly involving major salivary glands and lymph nodes. It can also involve axillary, inguinal or epitrochlear nodes although infrequent. Co-existing renal disease is common, with an incidence ranging from 10% to 60%. In these cases it may present as all types of glomerulonephritis or as nephrotic syndrome.

Hypereosinophilia and elevated serum IgE level are considered as important markers in pathology and diagnosis of Kimura’s disease. Iwai et al had described blood eosinophilia at mean count of 35.2% with elevated IgE in all Kimura’s cases. The patient reported here also has eosinophilia and elevated serum IgE levels. Kimura’s disease can be mistaken easily for a malignant tumor like acute lymphocytic leukemia, T-cell lymphoma, Hodgkin’s etc., among others. That is why differential diagnosis should be performed very carefully taking into account all clinical and histological findings.

The constant histological features seen in this disease are preserved lymph node architecture, lymphatic follicular hyperplasia, vascular proliferation and eosinophilic tissue infiltration. Additional features which may be present include prominent germinal centre, fibro collagenous deposition and marked fibrosis. Fine needle aspiration cytology is of limited value and substantial amount of tissue specimen is required for confirmation of diagnosis. Therefore hematological, serological and imaging studies are valuable for provisional diagnosis but only excisional biopsy provide with a definite histological diagnosis.

Several reports have described the findings of Kimura’s disease in computed tomography (CT), magnetic resonance imaging (MRI) and USG but all these modalities only provide non-specific picture and diagnosis is not possible solely on basis of radiography. Therefore, the role of imaging is to provide the extent and dimensions of the tumor to aid in its resection.

There is no consensus on the management of Kimura’s disease till date. Many treatment options including surgical excision, oral steroids, cytotoxic therapy and radiotherapy have been tried. Complete surgical excision is preferable to others as it is both therapeutic and diagnostic. Also, the patients are spared from harmful effects of radiation and cytotoxic therapy. Conservative treatment includes oral steroids which have been reported to decrease the size of enlarged lymphnodes as well as decrease renal symptoms. But there is no evidence of reduction in size of affected salivary gland and the lesions usually get enlarged again after termination of steroids. The success of treatment is mainly reassured by a constant low dose steroids but the side effects of long
term steroid administration should also be taken into consideration.3,8

Radiation therapy is useful as an effective adjuvant therapy for incomplete resection cases or salvage treatment for recurrence cases and effective dose of radiation to be given is between 20-30 Gy.9 Other modes of treatment include retinoids, immunosuppressants and monoclonal antibodies (imatinib). Beccastrini et al had described a case in complete remission for 8 years with long term low dose cyclosporine A therapy.10 However, more comprehensive studies are required to determine the safety and therapeutic effects of drugs.

CONCLUSION

Kimura’s disease is a rare disease entity. Although difficult to diagnose clinically, it should be considered as a differential diagnosis in patients with primary lymphadenopathy and associated eosinophilia. The patients should be followed rigorously for long term as it is known to recur.

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