

## Case Report

# A rare case of Kimura lymphadenopathy in elderly female

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### ABSTRACT

Kimura's disease (KD) is a chronic inflammatory disease, a rare variety. It usually presents as non-tender subcutaneous swelling in head and neck region, predominantly in preauricular and submandibular area and is often associated with cervical lymphadenopathy, marked peripheral eosinophilia and an elevated immunoglobulin E (IgE) level. Renal involvement is the only systemic manifestation. Nephrotic syndrome is the most common manifestation of renal disease in KD. Diagnosis through Fine needle aspiration cytology (FNAC) is misleading and can easily be mistaken as a malignant disease. So, diagnosis is therefore only established by histopathological examination. The treatment of KD involves one of three major approaches are surgical excision, irradiation, or steroid therapy. Surgical excision is recommended as the treatment of choice but carries recurrence rates of 33-50%. In our study, we are reporting a case of elderly female who presented with left sided cervical lymphadenopathy for which excision biopsy was done and the histopathology examination shown as kimura lymphadenopathy, a rare case.

**Keywords:** Kimura's disease, Nephrotic syndrome, Cervical lymphadenopathy

### INTRODUCTION

Kimura's disease (KD) is a chronic inflammatory disease, endemic in Asian subcontinent but rare entity in the West. Etiology is not clear yet. The typical clinical presentation is characterized by painless, subcutaneous nodules, predominantly in the head and neck region. This condition is often associated with cervical lymphadenopathy, raised serum eosinophil counts, and markedly elevated serum immunoglobulin E (IgE) levels.<sup>1,2</sup> The submandibular, axillary, or inguinal lymph nodes may also be palpable in some individuals. Initial diagnosis of KD can be usually arrived with comprehensive history from the patient and complete physical examination. Additional definitive tests have to be performed for confirmation of specific systemic illness.<sup>3</sup> Renal involvement is the only systemic manifestation.<sup>4</sup> Nephrotic syndrome is the most common manifestation of renal disease in KD.<sup>5,6</sup> This rare chronic inflammatory condition is found almost exclusively in

Asian male individuals (70-80%) in their 2nd to 4th decade of life.<sup>7,8</sup>

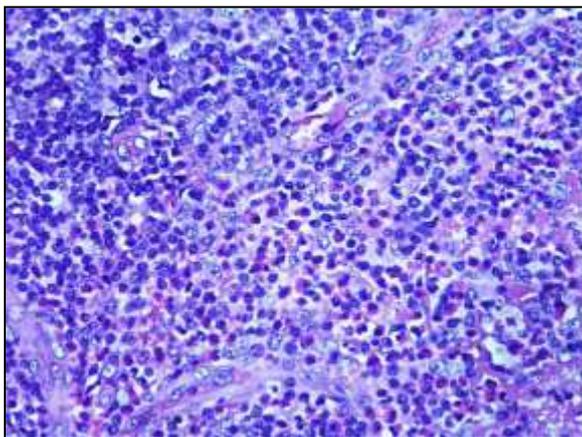
### CASE REPORT

A 61-year old female came to the outpatient department (OPD) with complaints of swelling over the left side of the neck and submandibular region for the past 6 months. Patient denied having fever, weight loss, cough or breathlessness, but she had past history of extra-pulmonary tuberculosis which was diagnosed after the excision of the swelling which was present just above her left elbow 8 months back. Soon after the diagnosis she was started on antitubercular treatment (ATT) drugs and completed the full course under revised national tuberculosis control programme (RNTCP) guidelines. She was a hypertensive and was under regular medication. Lab investigation revealed to have slightly elevated liver function test (serum glutamic-oxaloacetic transaminase (SGOT) 30 IU/L and alkaline phosphatase (ALP) 168 IU/L), whereas the rest of

the lab values were found to be within the normal range. Clinically patient had a single swelling measuring 3×2 cm present in the left lateral aspect of the neck, which was firm, non-tender and immobile in nature. On further examination there was another 2 swelling, one with 0.3×0.3 present just below the former one with the same features and another swelling of 2×2 cm was found to be in left side just below the angle of mandible with the features of the above swelling. So clinically suspected tuberculous cervical lymphadenopathy since already had tuberculosis history, planned for excision biopsy under local anaesthesia. Intra-operatively a group of submandibular lymph node were excised along with the cervical lymph nodes and the specimens were sent for histopathological examination which revealed features are consistent with Kimura's lymphadenopathy. Wound healed and all sutures removed in 12th postoperative period.



**Figure 1: Postoperative picture of the patient.**



**Figure 2: Histopathology slide.**

## DISCUSSION

This entity was first described in China by Kim and Szeto in 1937.<sup>14</sup> However, it did not become much popular till Kimura and colleagues described cases in Japan in 1948 by their histologic findings of “unusual granulation combined with hyperplastic changes in lymphoid tissue”. Later this disorder came to be known as Kimura's disease (KD). It usually presents as non-tender subcutaneous

swelling in head and neck region, predominantly in preauricular and submandibular area and is often associated with cervical lymphadenopathy, marked peripheral eosinophilia and an elevated IgE level (Messina et al).<sup>15</sup> Sometimes it may be mistaken for a malignant disorder. Fine needle aspiration cytology (FNAC) is misleading and so diagnosis is established only on histopathological examination. Renal involvement is the only and common systemic manifestation which may affect up to 60% of patients as membranous glomerulonephritis, minimal change glomerulonephritis, diffuse proliferative glomerulonephritis, mesangial proliferative glomerulonephritis and also nephritic syndrome (12% of cases) Messina et al, Hui et al.<sup>4,15</sup> KD commonly involves parotid glands, the epitrochlear, axillary, and inguinal nodes. Although the masses enlarge slowly, patients remain asymptomatic otherwise. Pruritus and dermatitis may occur, and also involve kidneys, orbits, ears, spermatic cord, and nerves. Common type of renal involvement is nephrotic syndrome.<sup>10,11</sup> Widespread disseminated intravascular thrombosis is also reported in literature, affecting mesenteric and renal veins (thrombotic storm).<sup>11</sup> The etiology is not clear yet but could be an abnormal immune response to an unknown antigenic stimulus.<sup>3</sup> Diagnosis through FNAC is misleading and can easily be mistaken as a malignant disease. So diagnosis is therefore only established by histopathological examination. T-cell lymphoma, kaposi sarcoma, hodgkin's disease, and angio- lymphoid hyperplasia with eosinophilia are the potential differential diagnoses (Churia et al). Differential diagnosis between KD and angiolymphoid hyperplasia with eosinophilia (ALHE) has been a challenge for a long time. In contrast to ALHE, in KD, germinal centers are destroyed due to heavy infiltration of eosinophils and absence of vacuolated endothelial cells. Immunofluorescence tests show heavy IgE deposits and variable amounts of IgG, IgM, and fibrinogen.<sup>7</sup> The treatment of KD involves one of three major approaches are surgical excision, irradiation, or steroid therapy. The clinical course of KD is generally benign. Non-steroidal anti-inflammatory drugs (NSAIDs), corticosteroids, cryotherapy, intralesional chemotherapeutic agents (bleomycin, vinblastine and tacrolimus), surgical excision, radiotherapy, laser therapy are the various options for treatment. Surgical excision is recommended as the treatment of choice but carries recurrence rates of 33-50%.<sup>12</sup> The recurrence of disease can be various factors like disease duration time, lesion size diameter, blood eosinophil count, well-defined lesion boundaries, serum IgE levels and single or multiple lesions.<sup>13</sup>

## CONCLUSION

Kimura's lymphadenopathy is a chronic autoimmune inflammatory condition causing cervical lymphadenopathy. Even though it is endemic in Asia, it is not a common cause for cervical lymphadenopathy in clinical practice like tuberculosis or malignant conditions. So clinical diagnosis of KD is not common in routine

practice. Hence the idea behind this article is to draw the attention of the clinician to this disease in diagnosis and management. KD should be considered in differential diagnosis in patients who present with primary cervical lymphadenopathy with eosinophilia and investigated accordingly as the disease has an indolent course and good prognosis. We present one such case of KD an uncommon autoimmune condition of unknown etiology in the cervicofacial region.

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