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– CASE REPORT –

Kimura Disease – Not Every Lymphadenopathy is Malignant

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Abstract

Introduction. *Any obvious lymphadenopathy should be given careful attention and thorough evaluation. With a spectrum of diseases presenting with lymphadenopathies, the clinical presentation, along with histo-pathological correlation, is important for the final diagnosis.*

Case. We are presenting a case of symptomatic lymphadenopathy for 1 month in a middle-aged woman, who, on evaluation, was found to have Kimura disease, a chronic inflammatory condition with unknown etiology, on excision biopsy. The patient was initially treated with corticosteroids, causing the disappearance of lymph nodes and symptomatic improvement. After five months, she had a recurrence of symptoms on steroid tapering, which was managed with an increase in the dose of oral corticosteroids, leading to resolution of symptoms.

Conclusion. *Kimura disease is a chronic inflammatory condition. Patients present with lymphadenopathy along with B symptoms.Surgery, immunosuppression and radiotherapy are treatment options.*

Keywords: *Kimura Disease, lymphadenopathy, lymph nodes*

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Introduction

Diagnostic advancements have made it possible to diagnose rare diseases. They present with common symptoms, but often remain undiagnosed. The diagnosis of these cases is often complicated by a range of symptoms similar to those of endemic disease. Lymphadenopathy is one of the most common symptoms occurring with varied differential diagnoses, according to

age, not strictly so. It needs a histopathological examination if it persists for a long time. In India, Lymphadenopathy is most commonly attributed to tuberculosis and other endemic diseases. [1] Lymphoma accounts for only nearly 5% of patients with lymphadenopathy. [2] Patients of middle age presenting with generalized lymphadenopathy often raise suspicion of malignancy; sometimes, rare diseases are contributory. Kimura disease is a non-malignant condition caused by chronic inflammation of unknown etiology. The disease has seen a preponderance towards the Asian population, with more males affected than females.[3] Patient presents with painless subcutaneous nodules with or without lymphadenopathy, mainly in the head and neck region, with a few presenting with salivary gland and renal involvement.[4] Lymphadenopathy in the visceral region is documented but occurs rarely. Salivary gland involvement is more common in males, while lymphadenopathy is more common in females. [5] Serum IgE is elevated in most cases, along with peripheral eosinophilia.[6-7] Patients may have systemic symptoms such as fever, weight loss, and night sweats, so a thorough assessment is necessary. Diagnosis is established by a tissue biopsy demonstrating features of tissue eosinophilia, florid follicular hyperplasia, interstitial fibrosis, and eosinophilic abscess.[5] Treatment includes surgical excision, prolonged corticosteroid therapy, immunosuppression, and radiotherapy. [8] We are reporting a case of a 47-year-old female with neck swelling with B symptoms.

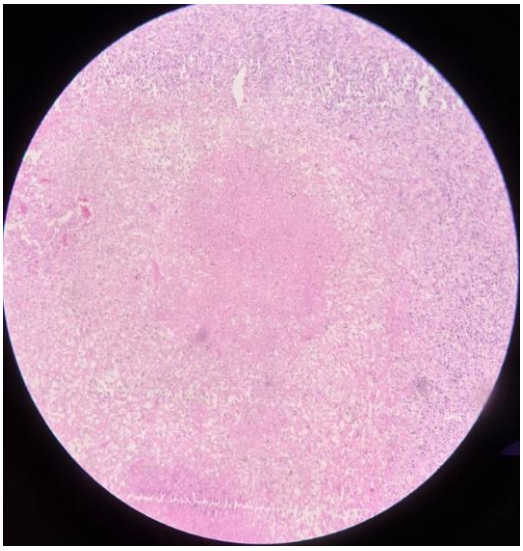

Image 1a

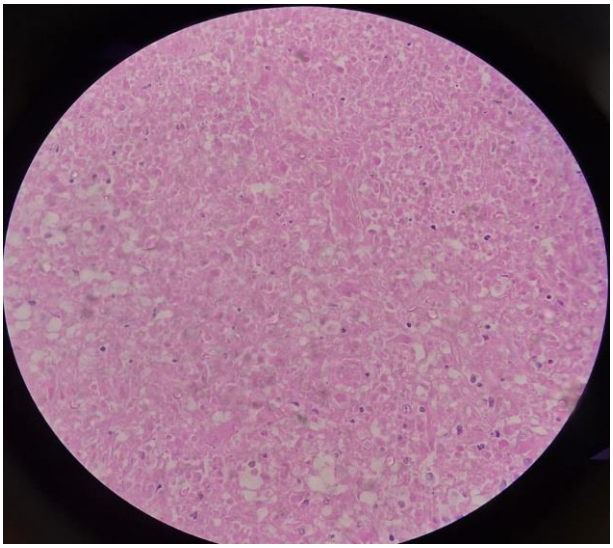

Image 1b

Image 1a and 1b*:Histopathological examination of the excised lymph node showing eosinophilic microabscess surrounded by polymorphous inflammatory cells. (Hematoxylin and eosin stain)*

Case report

A 47-year-old female with no comorbidities presented to the outpatient department of Clinical Haematology with left-sided neck swelling , low-grade fever with evening rise of temperature , fatigue and weight loss of 5 kg over a period of 2 months. The neck swelling was insidious in onset, gradually increasing in size over 2 months. On examination, the largest was a 4 x 2 cm non-tender neck swelling in the cervical region at level III, along with supraclavicular, bilateral inguinal, and femoral lymph nodes. The overlying skin was unremarkable. She had injected conjunctiva with a puffy face. Other systemic examination was unremarkable. Her initial blood workup is shown in **Table 1.**

Parameters	Baseline	After 1 months of corticosteroid therapy
Hemoglobin (Hb) (gm/dl)	10.3	10.1
Total Leucocyte count (TLC) (µL)	7230	8100
Platelets (µL)	140000	416000
Differentials (%)	N41 L19 E36	N40 L51 E2
Lactate Dehydrogenase (LDH) (IU)	364	201
Erythrocyte sedimentation rate(ESR) (mm/hr)	60	20
Absolute Eosinophil Count(AEC) µL	2600	220
Total IgE (IU/ml)	485	100
C- reactive protein (mg/L)	< 5	-

Table 1. *Laboratory Parameters Before and After Treatment (N- Neutrophils, L-Lymphocyte, E-Eosinophil)*

PET CT suggested FDG-avid intraparotid, bilateral cervical, supraclavicular, bilateral axillary , anterior diaphragmatic , bilateral lateral chest wall, mediastinal, abdominal, pelvic, bilateral inguinal and bilateral femoral lymph nodes. Biopsy of the cervical lymph node suggested benign chronic inflammatory disease with an eosinophilic microabscess in a background of polymorphous inflammatory cells, suggestive of Kimura disease. Immunohistochemistry (IHC) showed CD3 and CD20 in reactive T and B cells, respectively, with negative BCL2 and CD15. Immunoblasts were positive for CD30.

After discussing treatment options with the patient, she was started on prednisolone oral therapy 1mg/kg, which substantially reduced lymphadenopathy along with B symptoms. Corticosteroids were tapered gradually. Five months from treatment, she had similar complaints as initial complaints, which were managed with an increase in the dose of ongoing corticosteroid therapy along with blood pressure and blood sugar monitoring, resulting in remission of these symptoms.

Discussion

Any obvious lymphadenopathy should be given careful attention and thorough evaluation. Given the spectrum of diseases presenting with lymphadenopathy, the clinical presentation, along with histopathological correlation, is important for the final diagnosis. Our patient had generalised lymphadenopathy with B symptoms, presenting at middle age, suggesting a malignancy; however, generalised lymphadenopathy along with B symptoms has also been reported in Kimura disease.[9] Also, a case series of 23 patients with Kimura disease showed that lymphadenopathy was a prominent feature in females, whereas salivary gland involvement was more common in males.[5] Our patient had injected conjunctiva with facial puffiness, which is one of the findings associated with Kimura disease, occurring in patients with a history of atopy.[10] Our patient had no history of Bronchial asthma or skin rashes, but the presence of injected conjunctiva along with facial puffiness indicates the presence of an underlying allergic process. Identifying the allergic process is important, as the probable pathogenesis of Kimura disease involves an immune response to persistent antigenic stimulation, believed to be driven by the interplay between Th2-mediated cytokines and IgE. [11]

The differential diagnosis for Kimura disease, when the diagnostic dilemma arises, includes angiolymphoid hyperplasia with eosinophilia (ALHE) and IgG4-related disease. ALHE generally affects the young to middle-aged population with no sex predilection and is more common in the Western population, and is rarely associated with lymphadenopathy.[12-13] The presence of high IgE levels in our patient, along with generalised lymphadenopathy, makes the diagnosis of ALHE unlikely. Moreover, histopathologically, vascular proliferation is more prominent in ALHE than in Kimura disease, in which germinal centre hyperplasia, interstitial fibrosis, excessive

eosinophilic infiltration, and abscess formation are characteristic features. [14]

IgG4-related disease is associated with the pancreas, salivary glands, orbits, kidney, and lung, where biopsy shows increased IgG4+ plasma cells, a feature also seen in Kimura disease, along with other similar features such as high levels of IgE and peripheral eosinophilia in the background of an immune-mediated disease process. [15] Visceral involvement, such as the kidneys, is also common in both diseases, and both respond well to corticosteroid therapy, making them difficult to distinguish. In IgG4-related disease, storiform fibrosis and obliterative phlebitis are characteristic features, while interstitial fibrosis and eosinophilic abscess are characteristic features of Kimura disease.[5,16] Best modality for the treatment of Kimura disease is surgical excision followed by radiotherapy. Relapses are common and often compel to start immunosuppressive therapy handling the immune dysregulation.

Conclusion

Kimura disease is a rare benign condition characterised by chronic inflammation triggered by an unknown factor. The presentation could be similar to malignancy; however, the disease is self-limiting and has a good response to excision, radiotherapy and corticosteroids, though relapses are common.

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Ethics Statement and Conflict of Interest Disclosures

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Ethics Consideration: The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national laws. Written informed consent was provided by the participant in this study.

Conflict of interest: No known conflict of interest correlated with this publication.

Availability of data and materials: The data used and/or analyzed throughout this study are available from the corresponding authors upon reasonable request.

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