

## A case report of KIMURA'S DISEASE with NASAL SEPTUM PRESENTATION

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### ABSTRACT

Kimura disease is a rare chronic inflammatory disorder of unknown aetiology, primarily seen in young Asians. The disease is characterised by painless subcutaneous swelling, blood and tissue eosinophilia and raised IgE levels. Kimura's Disease has three components; cellular component showing lymphoid follicles with germinal centre hyperplasia with inflammatory infiltrate including increased eosinophils, plasma cells, and lymphocytes, surrounded by fibro collagenous and vascular stroma. Kimura disease is a disorder that involves the subcutaneous tissue of the head and neck regions. It is usually associated with regional lymphadenopathy and/or salivary gland involvement.

**KEYWORDS:** Kimura, Nasal septum, Eosinophilia, Subcutaneous nodules

### INTRODUCTION

Kimura's disease is a chronic inflammatory condition of unknown aetiology presenting as multiple painless solitary subcutaneous nodules localized mostly in the region of the head and neck with coexisting lymphadenopathy and peripheral eosinophilia. Kimura's disease is limited to the skin, lymph nodes, and salivary glands. Renal involvement is its only systemic manifestation.<sup>[1]</sup> This rare condition is found almost exclusively in Asian individuals in their 2nd to 4th decade of life and mostly affects males (70–80%).<sup>[2]</sup> Management of this disease is a conservative approach that is best suited for treatment.

### CASE REPORT

A 46-year-old female resident of Hyderabad, India presented with complaints of an insidious onset of a gradually progressive swelling over the nasal bridge, as midline mass for 1 year.

She had no associated symptoms such as nasal or ear discharge. Upon physical examination, a 4×3 cm swelling was observed at the nasal dorsum. This swelling was non-tender, firm to soft and non-compressible. The skin over the swelling was normal. The fine needle aspiration cytology (FNAC) yielded bloody aspirate which was inconclusive. Haematological investigations revealed eosinophilia (27%) and elevated serum IgE (220 U/ml).

A surgical excision was performed and we received two firm, grey-white to grey-brown tissue bits, the largest measuring 2 x2 x1 cm and the smallest measuring 1 x1 cm., Figure 1

Histopathology of all embedded specimens revealed fibrofatty muscular connective tissue stroma showing lymphoid follicles with germinal centre hyperplasia and a few areas with loss of effacement of lymphoid follicles due to eosinophilia. (Figure 2) (Figure 3). These germinal centres of the lymphoid follicles show few congested blood vessels, and polykaryocytes (Figure 4). The paracortical areas show areas of fibrosis with sclerosed blood vessels. There are areas of infiltration with plenty of eosinophils, plasma cells and lymphocytes within the hyalinized stroma (Figures 5, 6 and 7) which are all features suggestive of Kimura's disease.

### DISCUSSION

Kimura's disease was first described in 1937 and later popularized in 1947 by Kimura and his associates, it is a benign disease which involves subcutaneous tissues (preauricular, submandibular), the major salivary gland, and lymph nodes mainly in the head and neck area.<sup>[3]</sup> China and Japan are endemic countries, although sporadic cases are described elsewhere.<sup>[4]</sup> Kimura's disease may affect the kidneys in up to 60% of patients, presenting as all types of glomerulonephritis or as nephritic syndrome (12%).<sup>[5]</sup> Hyper eosinophilia and elevated serum IgE are found in Kimura's disease as well.

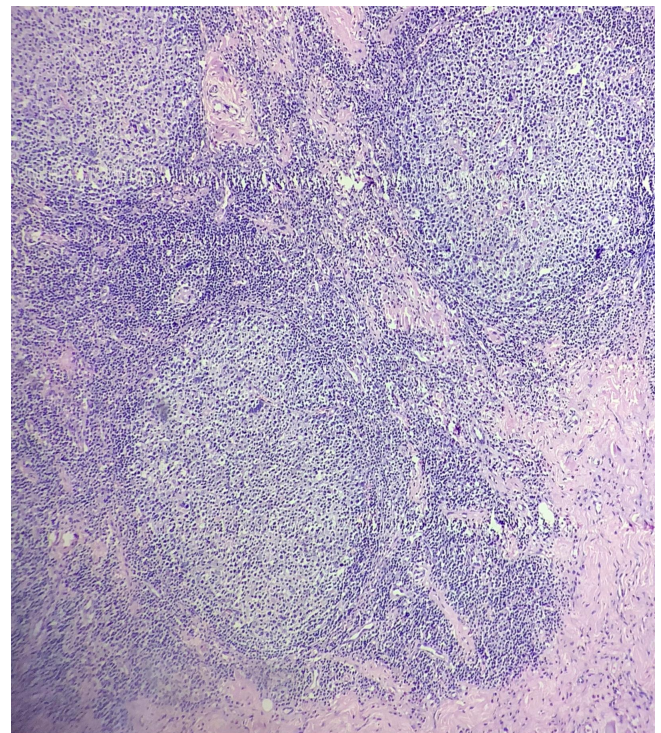




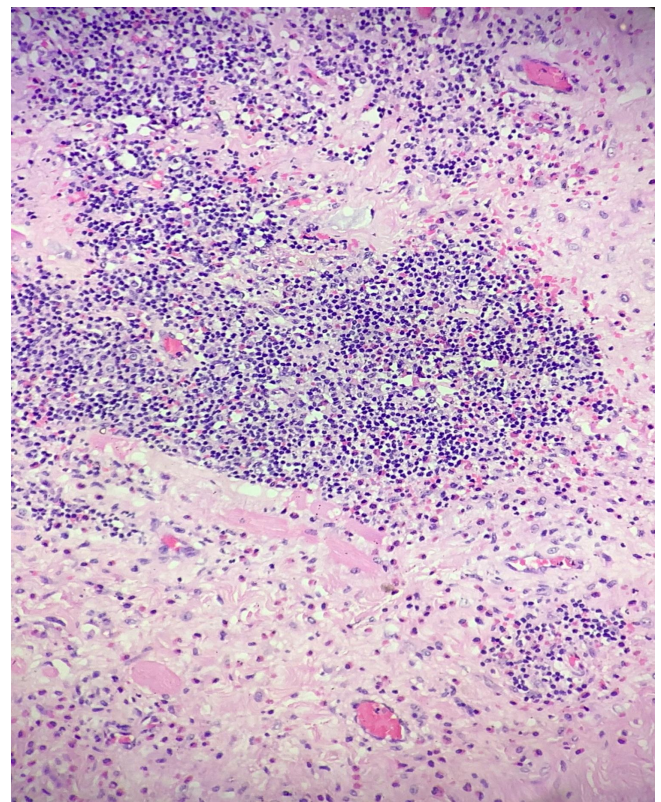
**Figure 1: GROSS SPECIMEN RECEIVED FOR HISTOPATHOLOGICAL EXAMINATION.**

Diagnosis through FNAC is misleading and can easily be mistaken for a malignant disorder. Diagnosis is therefore only established through histopathological examination. T-cell lymphoma, Kaposi Sarcoma, Hodgkin's disease, and angio-lymphoid hyperplasia with eosinophilia are potential differential diagnoses. Differential diagnosis between Kimura's disease (KD) and Angio lymphoid hyperplasia with eosinophilia (ALHE) has been a challenge for a long time. In contrast to ALHE, in Kimura's disease, germinal centres are destroyed due to heavy infiltration of eosinophils and the absence of vacuolated endothelial cells. Immunofluorescence tests show heavy IgE deposits and variable amounts of IgG, IgM, and fibrinogen 1. However, these tests were not performed in our case.

There is no consensus on the management of Kimura's disease; however, a conservative approach may be sufficient with the use of other modalities of treatment. Conserva-

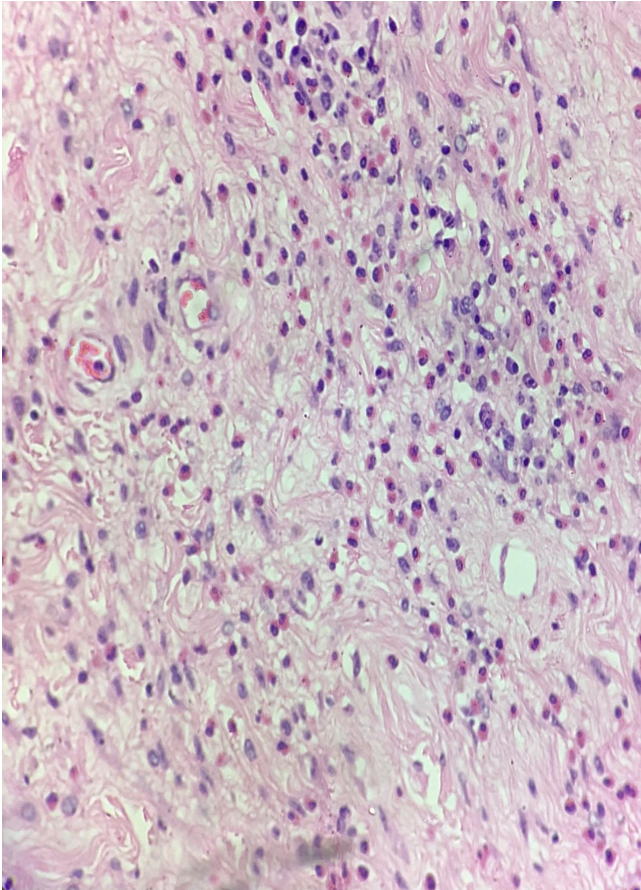


**Figure 2: SECTION SHOWING HYPERPLASTIC GERMINAL CENTRES. [ H&E STAIN, 10X]**

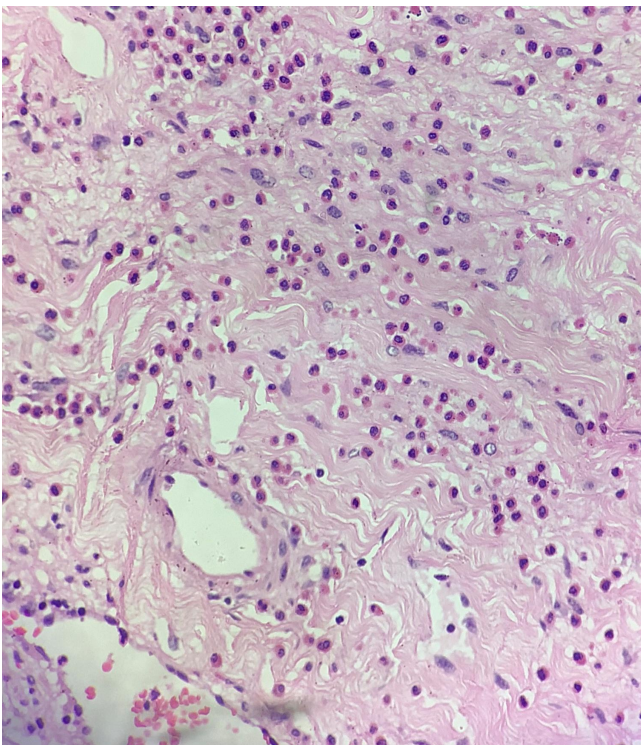


**Figure 3: SECTION SHOWING CONGESTED BLOOD VESSELS AND EFFACEMENT OF LYMPHOID FOLLICLES DUE TO EOSINOPHILIA. [ H&E STAIN, 40X ]**

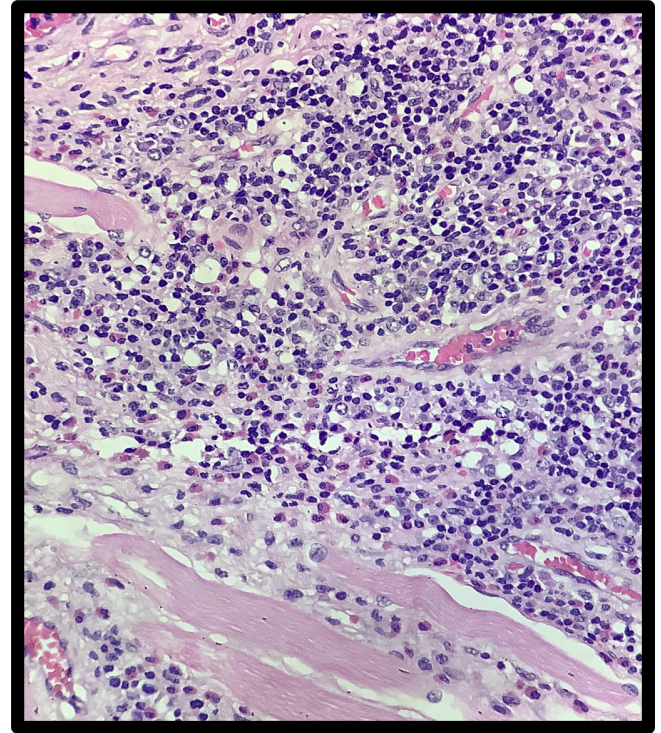




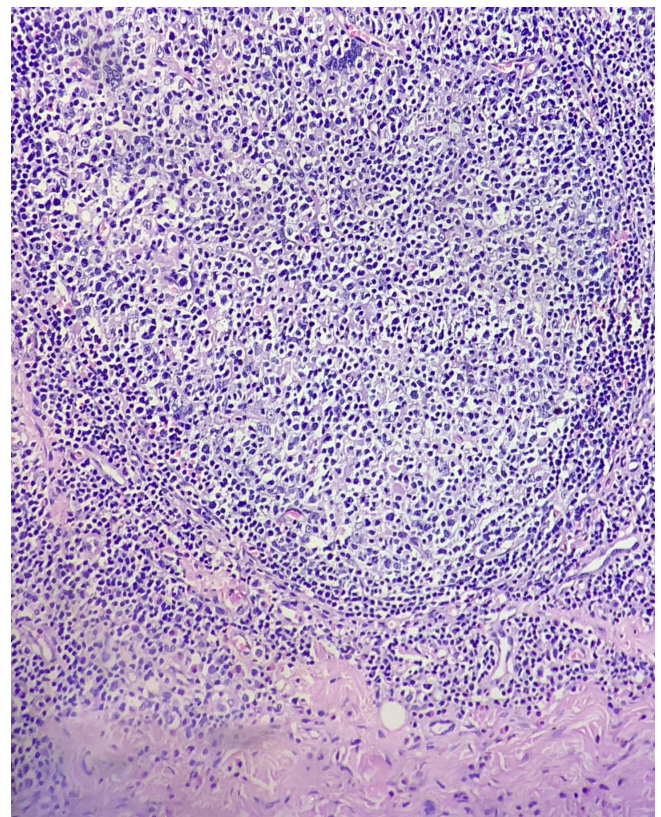
**Figure 5: SHOWS PLENTY OF EOSINOPHILS ALONG WITH PLASMA CELLS, AND LYMPHOCYTES IN HYALANISED STROMAL BACKGROUND. [ H&E STAIN, 40X]**



**Figure 6: SHOWING SCLEROSIS OF BLOODVESSELS.[ H&E STAIN , 40X ]**



**Figure 7: SHOWING EXTENSIVE FIBROSIS WITH CONGESTED BLOOD VESSELS DUE TO EXTENSIVE INFILTRATION OF EOSINOPHILS. [ H&E STAIN, 40X]**



**Figure 4: SECTION SHOWING HYPER-PLASTIC GERMINAL CENTRE WITH POLYKARYOCYTES. [H& E STAIN, 40X ]**



tive treatment includes oral steroids; however, the lesions usually get enlarged again when steroid treatment is terminated. Thus, successful treatment is mainly reassured by a constant low dose of steroids. [2] Surgery and subsequent steroid treatment are proposed as an alternative treatment. Radiation therapy is offered to patients who don't respond to surgery or oral steroids.

This case is exceptional as - 1) It is a sporadic case found in non-Orientals. 2) It shows involvement of the nose with no lymphadenopathy 3) Inconclusive FNAC is seen and histological diagnosis led us to do retrospective investigations and 4) Grossly elevated IgE levels are seen. In addition, there was marked eosinophilia.

## CONCLUSION

Kimura's disease, although difficult to diagnose clinically, should be considered in the differential diagnosis in patients who show primary lymphadenopathy with eosinophilia with or without subcutaneous nodules. It should be investigated accordingly as the disease has an indolent course and good prognosis.

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