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Kimura Disease – A Rare Cause of Chronic Swelling and Inflammation

Dr. Keerthana Patta[1*], Dr. Priyanka Barphe[2], Dr. Devidas B Dahiphale[3], Dr. Saurabh Joshi[4]

[1*]Junior resident at Department of radiodiagnosis, MGM Medical College & Hospital Chhatrapati Sambhajingar
Email ID: keerthanapatta@gmail.com
[2]Assistant Professor at Department of radiodiagnosis, MGM Medical College & Hospital Chhatrapati Sambhajingar
Email ID: barphep@gmail.com
[3]Head of the department at Department of radiodiagnosis, MGM Medical College & Hospital Chhatrapati Sambhajingar
Email ID: drdbdahiphale@gmail.com
[4]Associate professor at Department of radiodiagnosis, MGM Medical College & Hospital Chhatrapati Sambhajingar
Email ID: saurabh21j@gmail.com

***Corresponding Author:**
Dr. Keerthana Patta
Department of radiodiagnosis, MGM Medical College & Hospital Chhatrapati Sambhajingar
Email ID: keerthanapatta@gmail.com

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ABSTRACT
Kimura Disease (KD) is a rare, benign chronic inflammatory condition characterized by painless subcutaneous nodules, lymphadenopathy, eosinophilia, and elevated serum IgE levels. This report describes a 35-year-old female with a painless left-sided head and neck mass. Diagnosis was confirmed by clinical, radiological, and histopathological evaluation. Due to its overlapping features with malignancy and other inflammatory conditions, timely recognition of KD is essential for proper management.

K Keywords - *Kimura Disease, Lymphadenopathy, Eosinophilia, Radiological evaluation*.

INTRODUCTION

Kimura Disease (KD) is a rare, chronic inflammatory disorder primarily affecting young to middle-aged Asian males. It presents as painless subcutaneous nodules, often in the head and neck, with regional lymphadenopathy, peripheral eosinophilia, and elevated serum IgE levels [1]. Initially described in 1937 and characterized by Kimura in 1948, KD was once confused with Angiolymphoid Hyperplasia with Eosinophilia (ALHE) but is now recognized as a distinct entity [1]. Though the exact cause remains unclear, KD is thought to result from an allergic or autoimmune response. A Th2-mediated immune mechanism, with elevated IL-4, IL-5, and IL-13 levels, contributes to eosinophil activation and vascular proliferation [2,3]. Infectious or environmental triggers have been suggested but are unproven [3].

Clinically, KD manifests as slow-growing, firm, painless swellings in the periauricular, submandibular, and parotid regions, with no systemic symptoms, distinguishing it from malignancies and infections [4]. Imaging typically shows hypoechoic, hypervascular lesions on ultrasound, and homogeneously enhancing soft tissue masses on CT and MRI, with no necrosis or bone erosion. Diffusion-weighted MRI can help differentiate KD from malignant tumors [5].

Histopathology is diagnostic, revealing eosinophilic infiltration, lymphoid follicular hyperplasia, and vascular proliferation, particularly of postcapillary venules. Fibrosis may be present, but granulomas are absent, excluding conditions like tuberculosis or sarcoidosis [6].

Treatment involves surgical excision for localized lesions, though recurrence is common. In recurrent or extensive cases, steroids, immunosuppressants (e.g., cyclosporine A, imatinib), biologics (e.g., omalizumab), and radiotherapy are used [7]. Long-term follow-up is essential due to the relapsing nature of KD.

Case Presentation

A 35-year-old female presented to the Department of Radiology at MGM Aurangabad with a two-year history of painless swelling in the left external auditory canal (EAC) and postauricular region, accompanied by hearing difficulty. No systemic symptoms, such as fever, night sweats, or weight loss, were reported, narrowing down the differential diagnosis. On physical examination, two well-defined, non-tender, firm swellings (2 × 2 cm) were observed. The overlying skin had a bluish discoloration, suggesting vascular involvement, but there was no local rise in temperature. Systemic examination was unremarkable.

Figure 1- Bluish painless mass protruding from the left external auditory canal (EAC)

Laboratory findings revealed peripheral eosinophilia (10%) and elevated serum immunoglobulin E (IgE), suggesting an allergic or immune-mediated disorder. Ultrasound showed hypoechoic swelling with increased vascularity in the postauricular region and enlarged hypoechoic lymph nodes with increased hilar vascularity in the parotid gland. Non-contrast CT revealed homogeneous soft tissue density lesions in both the EAC and postauricular regions. MRI demonstrated an isointense lesion on T1-weighted images, hypointense on T2-weighted images with multiple flow voids, suggesting vascular proliferation. Post-contrast T1 images showed significant homogeneous enhancement, indicating hypervascular tissue. Enlarged lymph nodes in the left parotid caused obliteration of the left parapharyngeal space without bony erosion.

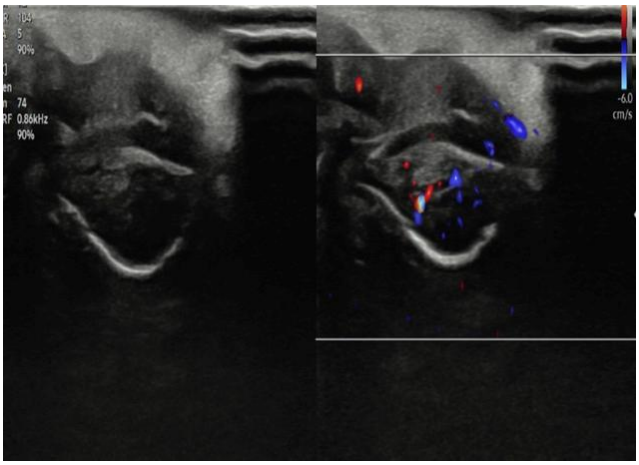

Figure 2-left postauricular swelling -appears hypoechoic and showed raised vascularity on USG

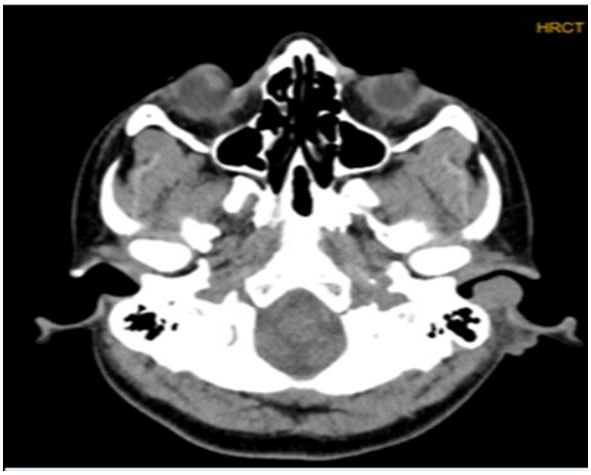

Figure 3-Axial NECT- homogenous soft tissue density lesion in left EAC and left post auricular region

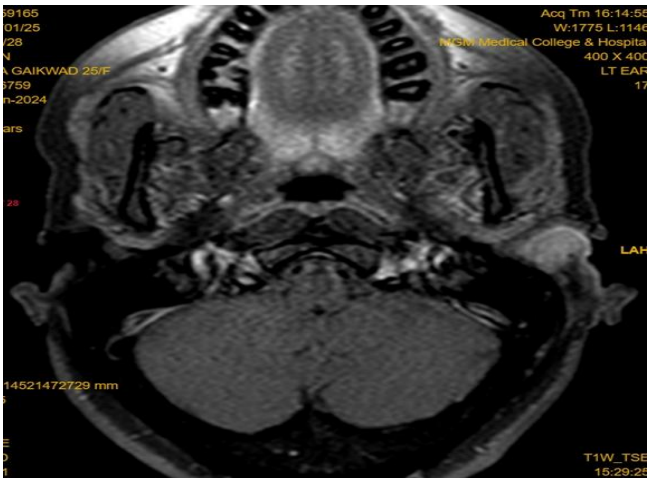

Figure 4-Axial T1 post contrast images-significant homogenous post contrast enhancement of lesion in EAC

Surgical excision was performed, and histopathological analysis confirmed Kimura Disease, with eosinophilic infiltrates, vascular proliferation, and lymphoid follicular hyperplasia. The absence of granulomas ruled out tuberculosis and sarcoidosis. The diagnosis of Kimura Disease was confirmed, and the patient was advised long-term follow-up for monitoring recurrence.

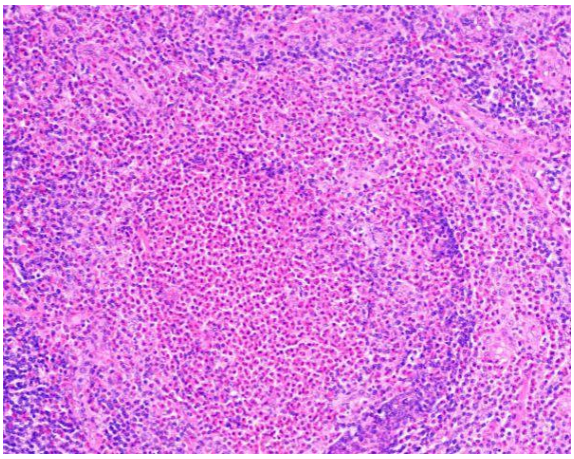

Figure 5- Histopathology images showing lymphoid follicular hyperplasia with eosinophilic infiltrate and vascular proliferation

DISCUSSION

A 35-year-old female presented with painless swellings in the left external auditory canal (EAC) and postauricular region, unusual for Kimura Disease (KD), which is more common in males. The patient had no systemic symptoms like fever, night sweats, or weight loss. Laboratory investigations revealed eosinophilia (10%) and elevated IgE levels, which are characteristic of KD. Ultrasound imaging showed hypoechoic, hypervascular nodules, while CT revealed homogeneous soft tissue density lesions in the affected areas. MRI demonstrated isointense lesions on T1-weighted images, hypointense lesions with flow voids on T2-weighted images, and significant homogeneous enhancement, indicating vascular proliferation. Histopathological examination confirmed KD with eosinophilic infiltrates, vascular proliferation, and follicular hyperplasia, without granuloma formation, ruling out conditions like tuberculosis or lymphoma.

The pathophysiology of KD is thought to involve an immune-mediated process, with a significant role played by T-helper 2 (Th2) cytokines like IL-4, IL-5, and IL-13, leading to eosinophil recruitment and IgE production. While infectious triggers such as Epstein-Barr virus (EBV) have been proposed, no definitive causative agents have been identified. Imaging findings in this case align with previous reports, such as a 32-year-old male with bilateral parotid gland enlargement (5). KD is frequently misdiagnosed due to its overlapping features with other lymphoproliferative and inflammatory conditions. The primary differential diagnoses for Kimura Disease (KD) include:

- **Angiolymphoid Hyperplasia with Eosinophilia (ALHE)**: Similar to KD but affects skin, lacks significant lymphadenopathy, and shows epithelioid endothelial cells (8).
- **Lymphomas**: Distinguished by the absence of systemic B symptoms and eosinophilia, with lymphoma showing atypical lymphoid cells (9).

- **Metastatic Lymphadenopathy**: Malignancies like nasopharyngeal carcinoma show necrotic nodes, unlike KD's homogeneous, well-defined lesions (10).
- **Chronic Infections**: Tuberculosis lacks eosinophilic infiltration and granulomas, helping differentiate it from KD (11).

  Treatment typically involves surgical excision, with high recurrence rates. Corticosteroids, immunosuppressants, and targeted therapies like imatinib or omalizumab are used in recurrent cases. Regular follow-up is crucial to monitor for recurrence, as KD has a benign course but is prone to relapse (13, 16).

CONCLUSION

Kimura Disease is a rare yet distinct chronic inflammatory disorder that poses diagnostic challenges due to its resemblance to other lymphoproliferative and inflammatory conditions. This case highlights the importance of considering KD in patients presenting with painless head and neck masses, particularly when eosinophilia and elevated IgE levels are present. Comparison with similar case reports reinforces the characteristic imaging and histopathological findings that aid in diagnosis. While surgical excision remains the mainstay of treatment, recurrence is common, necessitating a multimodal approach for optimal disease management. Further research is needed to identify novel targeted therapies and improve long-term outcomes for affected patients.

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