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Kimura Disease Presenting As A Recurrent Mass In Left Parotid Region, In A Case With Preexisting Renal Disease

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Abstract

Background: Kimura's disease is a distinctive angiolymphoid proliferative disorder characterized by an unknown etiology.

Case presentation: We presented a case of a 30-year-old male who presented to surgery OPD with complaints of swelling on his left cheek which was insidious in onset and gradually progressive since eight months. The patient was a known case of CKD since fifteen years and was on maintenance dialysis with a frequency of thrice a week. The case was misdiagnosed as a nerve sheath tumour and the excision of the swelling was done. The swelling was sent for histopathological examination which was suggestive of Kimura's disease.

Conclusion: Kimura's disease is a rare disease which needs consideration as a differential diagnosis when a case is presented as a lymph node with eosinophilic infiltration and prominent follicular hyperplasia. Accurate diagnosis can be achieved by employing rigorous histological criteria, especially when complemented by pertinent clinical information and laboratory findings. Thus, it is advisable to avoid radical surgery, as no such instances are reported of malignant transformation in the literature.

Keywords: NIL

Introduction

Kimura disease presents as an uncommon chronic inflammatory condition affecting subcutaneous tissues, primarily in the head and neck area, often accompanied by regional lymph node enlargement and/or salivary gland participation. It is more prevalent among males of Asian origin and can mimic a neoplastic growth clinically. Distinguishing Kimura disease from angiolymphoid hyperplasia with eosinophilia, which primarily affects the superficial skin of the head and neck region, is crucial to avoid confusion. [3].

Common sites of involvement in Kimura's disease include the periauricular, axillary, and inguinal lymph nodes, as well as the parotid glands. Kidney involvement represents the most substantial systemic manifestation of the disease, occurring in approximately 10% to 16% of KD patients. Other systemic manifestations are considerably less frequent.[2]

Case Report

A 30-year-old male patient presented to the Surgical Outpatient Department (OPD) with the complaint of swelling over left cheek since eight months (figure 1) which was insidious in onset and gradually progressive. He was a known case of CKD since fifteen years and was on maintenance dialysis twice

weekly since one year. He had history of hypertension since fifteen years. He had been taking steroids since fifteen years for renal disease. Renal biopsy was not done. During the period of admission, he was on maintenance dialysis with the frequency of three times a week. He also had an abdominal ultrasound which revealed reduced size of kidneys. He had no history of diabetes mellitus, tuberculosis or asthma.

Haematological examination revealed Hb 9.4 gm/dl, TLC 5620 /cumm, Eosinophils 20.7%, Platelet count 1,20,000 /cumm. Urea 34 mg/dl, Creatinine 5.2mg/dl, Na 136 mmol/L, Potassium 4.1mmol/L.

On examination, the swelling was of size 6x6cm, firm in consistency, no evidence of tenderness or pus discharge or any discoloration or redness.

A fine needle aspiration was performed from the swelling which revealed only haemorrhagic fluid. Clinically the case was diagnosed as a nerve sheath tumour and excision of the swelling was done. Specimen was sent for histopathological examination.

The specimen was received in four pieces, the largest (labelled as tumor) measuring 4.5 x 4 x 1.4 cm and remaining smaller nodules measuring 1 x 0.5 x 0.5 cm and 0.6 x 0.6 x 0.2cm. Separate specimen labelled as parotid gland measuring 7 x 5 x 2.5 cm was also received (figure 2A0. Cut surface of tumour and smaller nodules was homogenous grey white (figure 2B).

Figure 1 - Clinical picture showing swelling over left parotid region for the first time

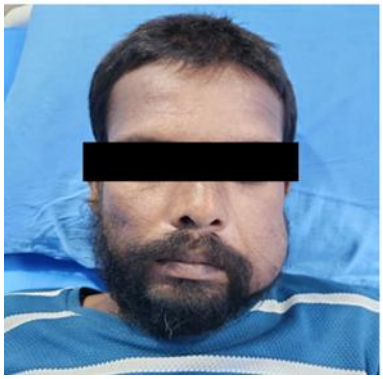
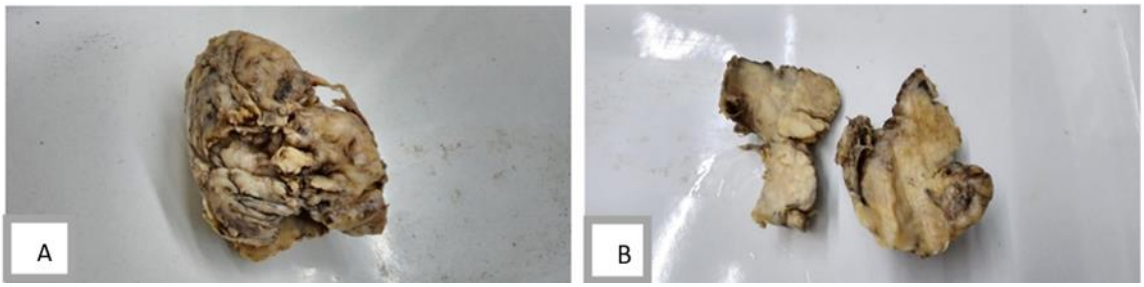


Figure 2 A/B. Gross appearance of the tumour with cut surface



Microscopy

Sections taken from tumour and smaller nodules revealed similar features. They showed hyperplastic follicles with germinal centres associated with expansion of interfollicular areas by abundant eosinophils, often forming eosinophilic micro abscesses admixed with polymorphous populations of lymphocytes and histiocytes. Intervening fibro-connective tissue showed mononuclear inflammatory infiltrate (figure 4, 5, 6). Many congested blood vessels and areas of haemorrhage were also seen. Warthin-Finkeldy giant cells were not seen. Sections from the parotid gland showed unremarkable histology. Histological features were suggestive of Kimura's disease.

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Figure 4. 40x view H/E showing lymphoid follicles are abundant eosinophils in interfollicles areas forming microabscesses at places

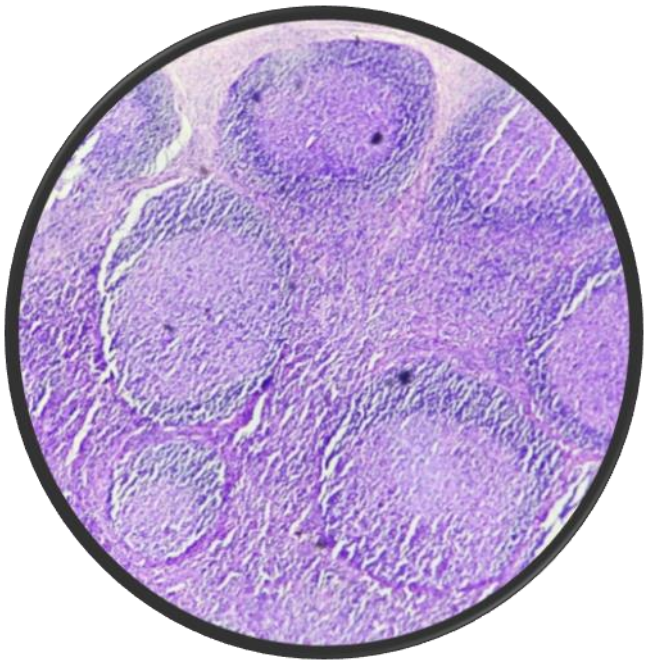

Figure 5. X10 magnification using Hematoxylin and eosin stain (H&E) showing fibroconnective tissue with lymphoid follicles and polymorphous inflammatory infiltrate

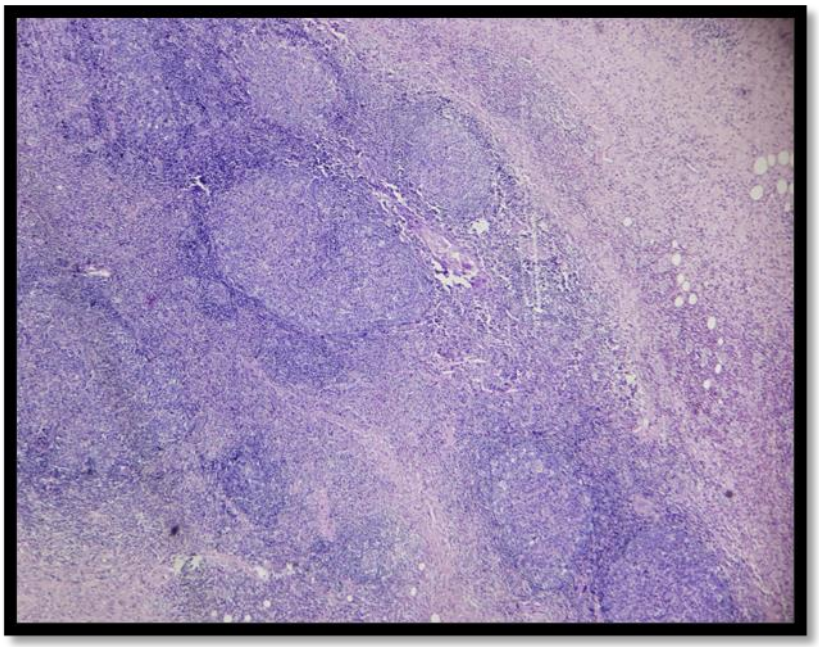

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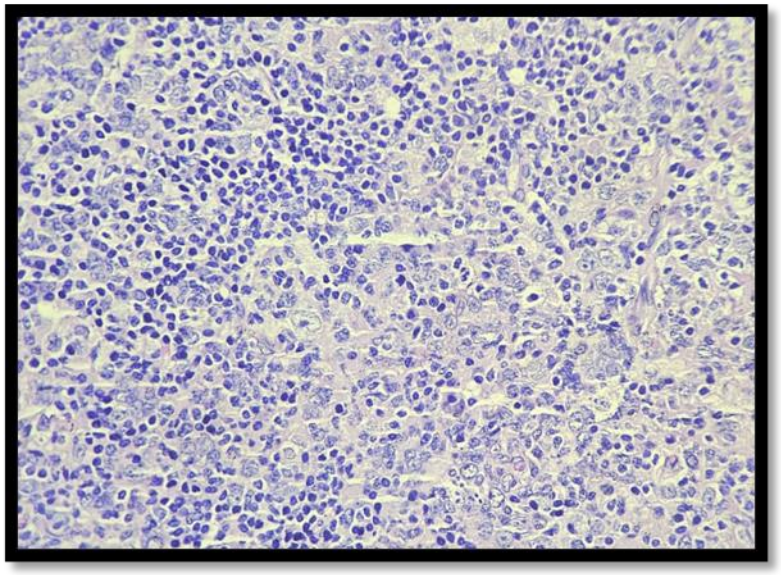
Figure 6. 100x view showing polymorphous population of cells, many lymphocytes, eosinophils and macrophages.



He had recurrence of swelling at the same site within 1 month of excision. The swelling gradually increased in size and was excised 18 months later. The recurrent swelling measured 5.1 x 4.2 x 1.9 cm. On gross multiple, irregular, brownish white, soft tissue masses were received total measuring 8 x 7.5 x 1 cm. Histopathology showed similar features. The patient is well 6 months after re-excision.

Discussion

Kimura's disease (KD) is a non-cancerous chronic inflammatory condition of the soft tissues, marked by elevated levels of eosinophils and IgE. While the exact aetiology of the disease remains unclear, some researchers have suggested that immune responses to viral infections and certain concurrent neoplasms could contribute to its development by disrupting T-cell immunoregulatory functions. [2].

The cause of KD remains unknown, though it could involve immune dysregulation, atopic reactions to persistent antigenic stimuli like arthropod bites, viruses, or neoplasms. An intriguing theory implicates Candida as a potential source of persistent antigenemia, despite the absence of isolated hyphae or spores. KD is characterized by hyperplasia of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate suggest KD may be a type of hypersensitivity reaction involving lymphocytes, possibly mediated by T-helper 2 cells. [1].

Renal involvement in Kimura's disease (KD) has been the subject of several studies, particularly concerning findings from renal biopsies. Ren et al. found that between 59% to 78% of patients exhibited nephrotic syndrome characterized by mesangial proliferative glomerulonephritis and membranous nephropathy, findings were consistent with those reported in other studies. KD combined with kidney involvement is seen in 10-60% cases [7]. However, in our case CKD was present for 10-12 year prior to development of Kimura's disease which is also reported in literature (3,9,10).

Several conditions can mimic KD, and the differential diagnosis should include angiolymphoid hyperplasia with eosinophilia (ALHE), Kaposi's sarcoma, tuberculosis, nodal metastasis (from breast and rectal cancer), hamartoma, epithelioid hemangioma, lymphocytoma, and low-grade angiosarcoma. [5]

In our case, the diagnosis was missed clinically and was suspected as a nerve sheath tumour.

In the literature, a significant debate has revolved around the distinction between KD and ALHE. Some view KD and ALHE as either identical or as various stages of a single disease process. Both conditions exhibit numerous similarities, such as soft-tissue masses primarily affecting the head and neck area, a

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protracted clinical course, and a favourable prognosis. Histologically, both conditions show a proliferative vascular pattern along with eosinophilic and lymphoid infiltration within the lesion. However, there are distinct clinical and histological criteria available to differentiate between KD and ALHE. [5]

In contrast, (ALHE) affects individuals across all racial demographics, showing a slight female predominance. Symptoms manifest as small, superficial nodules on the skin, often red in color, which can lead to bleeding, itching, and the development of tumours. While regional lymph node swelling, increased eosinophil levels in the blood, and elevated IgE levels are uncommon. [6]

KD commonly appears in young males, typically during their second and third decades of life. The lesions usually emerge as deep-seated, sizable soft-tissue masses within the subcutaneous tissue or salivary glands, with no significant changes in the overlying skin. Patients often experience regional lymphadenopathy (ranging from 67 to 100%). Nearly all cases exhibit peripheral blood eosinophilia and elevated serum IgE levels. [5].

KD treatment options include surgical excision, regional or systemic steroid therapy, and radiation. Among these, surgical resection stands out as the primary treatment, especially for localized masses, as was the case with our patient. However, despite its effectiveness, surgical resection carries a high reported rate of recurrence.[2].

Reports indicate that steroids can have an impact on Kimura's disease (KD); however, discontinuation of steroid treatment often leads to tumor recurrence. Furthermore, despite ongoing therapy, some tumors may become resistant to steroid treatment. Radiation therapy has been employed for refractory lesions, but concerns regarding the potential for secondary malignancies have tempered enthusiasm for its use in this typically benign condition. Furthermore, there is no documentation of the use of cyclosporine for treating KD. Surgical excision may be considered as an initial option, particularly for localized lesions, although recurrence remains a possibility. However, it is advisable to avoid radical surgery, as there are no reported instances of malignant transformation in the literature. [5]

Conclusion

While Kimura disease is uncommon, it warrants consideration as a potential diagnosis when encountering a lymph node with eosinophilic infiltration and prominent follicular hyperplasia. This condition has unique clinicopathological features, underscoring the importance of distinguishing it from drug reactions, hypersensitivity, and infectious etiologies. Accurate diagnosis can be achieved by employing rigorous histological criteria, especially when complemented by pertinent clinical information and laboratory findings. (3)

Though the condition is rare, awareness about this condition and its common clinical presentation can help the clinicians in arriving at the correct diagnosis. Case studies illustrating rare condition presentations and diagnostic challenges serve as valuable educational tools for clinicians.

References

1. Mohamed Ashraf Fouda, Osama Gheith, Ayman Refaie, et al.: Kimura Disease: A Case Report and Review of the Literature with A New Management Protocol. Int J Nephrol. 2010,1-4. 10.4061/2010/673908
2. Askar K. Alshaibani, Abdullah H. Al-Mulla, Mustafa Alhamoud, et al.: Isolated Kimura disease presenting a brow mass in a Saudi male: a rare case report. Cureus. 2022, 14(3): e23000. 10.7759/cureus.23000
3. Hong Chen, Lester D. R. Thompson, Nadine S. Ives Aguilera, et al.: Kimura Disease A Clinicopathologic Study of 21 Cases. Am J Surg Pathol. 2004, 28:505-513. 10.1097/00000478-200404000-00010
4. Cheng-Feng Tseng, Hsin-Ching Lin, Shun-Chen Huang, etal.: Kimura's disease presenting as bilateral parotid masses. Eur Arch Otorhinolaryngol. 2005, 262:8-10. 10.1007/s00405-003-0677-9
5. Harshi Dhingra, Ruchi Nagpal, Asif Baliyan, et al.: Kimura disease: case report and brief review of literature.
6. Song Ren, Xin Yi Li, Fang Wang, et al.: Nephrotic syndrome associated with Kimura's disease: a case report and literature review. BMC Nephrology. 2018, 19:316. 10.1186/s12882-018-1123-y
7. Manish Balwani, Charulata Bawankule, Amit Pasari, et al.: Minimal Change disease and Kimura's disease responding to Tacrolimus

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Therapy. Saudi J Kidney Dis Transpl. 2018, 30(1): 254-257. 10.4103/1319-2442.252921
8. Chien Te Lee, Chao Cheng Huang, King Kwan Lam, et al.: Kimura's Disease in a Chronic Hemodialysis patient. Am J Nephrol. 2001, 21:47-50. https://doi.org/10.1159/000046218
9. Yamada A, Mitsuhashi K, Miyakawa Y, Kosaka K, Takehara K, Iijima M, Tanaka K, Shibata S. Membranous glomerulonephritis associated with eosinophilic lymphfolliculosis of the skin (Kimura's disease): report of a case and review of the literature. Clin Nephrol. 1982 Oct;18(4):211-5.
10. Rajpoot DK, Pahl M, Clark J. Nephrotic syndrome associated with Kimura disease. Pediatr Nephrol. 2000 Jun;14(6):486-8. doi: 10.1007/s004670050799.

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