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Case Report

Kimura's disease: a rare post auricular recurrence

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ABSTRACT

Kimura's disease is a rare, chronic inflammatory condition of unknown cause, predominantly affecting individuals of Southeast Asian descent. It typically manifests in the head and neck region, presenting as deep subcutaneous nodules or masses, with peripheral blood eosinophilia and elevated serum immunoglobulin E (IgE) levels. Regional lymphadenopathy and salivary gland involvement are often associated with the disease. We report a case of a 38-year-old Indian woman diagnosed with Kimura's disease. She presented with a painless swelling on the right side of her face, located behind the ear, which had been gradually increasing in size over six months. Routine hematological, imaging, and histopathological evaluations confirmed the diagnosis. The patient was started on prednisolone at a dosage of 1 mg/kg/day, with close monitoring and additional supportive treatment. She showed remarkable clinical improvement, with a substantial reduction in the size of the swelling and lymph nodes, exceeding 50%. This case underscores the importance of considering Kimura's disease in the differential diagnosis for middle-aged South Asian adults with gradually expanding facial swellings. It also emphasizes the significance of early diagnosis and prompt intervention to reduce complications and facilitate quick recovery.

Keywords: Kimura disease, Lymphadenopathy, Salivary gland, Facial swelling

INTRODUCTION

First described in 1938 by Kimura and Szeto as "eosinophilic hyperplastic granuloma," Kimura disease is a rare, chronic inflammatory condition affecting deep subcutaneous tissue and lymph nodes in the head and neck, as well as the ante-cubital fossa and inguinal region.[1] It is a male preponderance condition, with a male-to-female ratio of 3:1, and occurs most commonly in the second and third decades of life.[2]

It typically manifests with painless regional lymphadenopathy or enlargement of salivary glands. Kimura et al further characterized it in 1948 in a paper titled "On the unusual granulation combined with hyperplastic changes of lymphatic tissue".[2] Previously considered associated with angiolymphoid hyperplasia with eosinophilia (ALHE), it is now recognized as a distinct entity with an unknown cause.

Proposed etiologies include allergic reactions, candida infections, dysregulation of eosinophil dynamics and IgE synthesis, and altered systemic immune-mediated reactions. The condition usually presents as a painless tumor-like mass in the head and neck area, accompanied by regional lymphadenopathy, blood and tissue eosinophilia, and elevated serum immunoglobulin E levels, suggesting an immunological basis.[3]

Diagnosis of Kimura disease relies on histopathological examination, with no reported instances of malignant transformation to date. There are no definitive management guidelines; however, treatments such as surgery, thalidomide, cyclosporine, interferon-α, pentoxifylline, omalizumab (an anti-IgE antibody),

corticosteroids, and radiotherapy have been recommended. Recurrence rates can be as high as 62%.

CASE REPORT

A 38-year-old female presented to the hospital on 22 May 2024, with chief complaints of a right-sided, painless facial swelling behind the ear for the past 6 months. The swelling has gradually increased in size. The patient is relatively asymptomatic, with no fever, nausea, vomiting, ear pain, discharge, or headache.

She reports no changes in vision, no weight loss, and a normal appetite. Both her family and personal medical history are insignificant. She experienced similar symptoms in 2010, for which she underwent surgical resection of the swelling followed by a short course of antibiotics for postoperative infection prophylaxis; however, no records are available for that treatment.

Upon examination, the patient's general condition was good. She was conscious and well-oriented to time, place, and person. Physical examination revealed a temperature of 100.4°F, heart rate of 86 beats per minute, respiration rate of 16 breaths per minute, blood pressure of 128/84 mm Hg, and transcutaneous oxygen saturation of 96% on ambient air, with no visible pallor or icterus. A well-defined swelling, characterized by a soft to firm consistency and tenderness, was palpated in the posterior auricular region, adjacent to the right parotid gland. The overlying skin appeared erythematous. Additionally, numerous enlarged, discrete, non-tender lymph nodes were detected in the submental, submandibular, upper anterior, and posterior jugular regions, predominantly on the right side. No axillary or inguinal lymphadenopathy was evident. Hepatosplenomegaly was absent. Systemic examination revealed no abnormalities.

Initial investigations (Table 1) suggested a hemoglobin level of 13.1 g/dl, a mean corpuscular volume of 83 fl, a total leukocyte count of 5,400/µl with 42% neutrophils, 26% lymphocytes, 4% monocytes, 18% eosinophils, serum IgE levels of 2193.9 IU/ml, and an erythrocyte sedimentation rate (ESR) of 8 mm/hour.

Other blood examination findings, including urine routine chemistry and microscopy, random plasma glucose, renal function tests, coagulation studies, and infectious markers (HBsAg, HCV, and HIV), were unremarkable. Tests for malaria (Paracheck), dengue (NS-1), and typhoid (Widal test) were negative.

Plain and enhanced axial computed tomography (CT) scans of the neck (Figure 1a and b) revealed a well-defined, irregular, peripherally enhancing collection with air foci in the subcutaneous plane of the right postauricular region, inferior to the pinna, and adjacent to the right parotid gland, with mild surrounding inflammation. The approximate size of the lesion is 2.4×2.2×2.4 cm (AP×TRA×CC). Both parotid and submandibular glands appear normal in size. There is mild compression over the superior pole of the parotid gland, with loss of the fat plane with the parotid gland. The fat plane with the sternocleidomastoid muscle is preserved. These findings suggest post-excision biopsy inflammatory changes with a thin rim of residual lesion adjacent to the parotid gland. Multiple subcentimeter to enlarged, homogeneously enhancing lymph nodes are seen in bilateral levels IB, II, and III, and the periglandular region of the bilateral parotid gland. The largest lymph node, measuring 8×14×11 mm (AP×TRA×CC), is located at level II on the right side, suggesting the possibility of nonspecific reactive pathology.

Table 1: Hematological investigations.

Test	Observed value	Reference range
Hemoglobin (g/dl)	13.1	12.0-15.0
RBC count (millions/c.mm)	4.40	3.8-4.8
Haematocrit (%)	38.0	36-46
Platelet count (cells/c.mm)	2,02,000	1,50,000-4,10,000
Total WBC count (cells/c.mm)	5,400	4,000-10,000
Neutrophils (%)	42	40-80
Lymphocytes (%)	36	20-40
Monocytes (%)	04	2-10
Basophils (%)	00	0-2
Eosinophils (%)	18 %	1-6
Absolute eosinophil count (cells/c.mm)	972	0-500
Serum IgE levels (IU/ml)	2,193.9	1.5-378

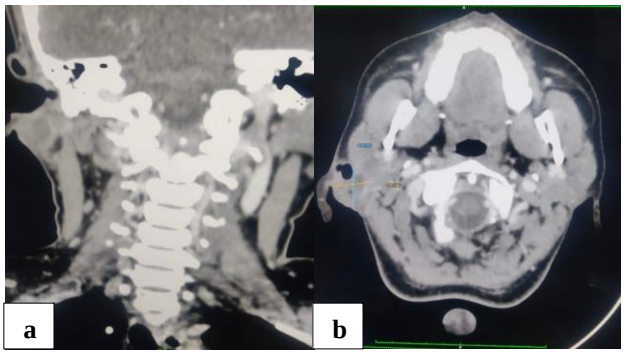

Figure 1: (a) Plain CT neck, and (b) enhanced axial CT scan through the neck.

An excisional biopsy of the swelling was performed, revealing multiple brown soft tissue fragments measuring 2×2 cm in aggregate. The cut sections exhibited white and brown areas. Microscopically, as shown in Figures 2a-d, the sections display squamous epithelium with underlying lymphoid tissue surrounded by adipose and fibroconnective tissue. The lymph nodes exhibit marked follicular hyperplasia with prominent germinal centers.

Numerous proliferating capillaries lined by prominent epithelioid endothelial cells are also present. There is significant eosinophilic infiltration, with the formation of eosinophilic abscesses. The surrounding adipose and fibroconnective tissues also show vascular proliferation and eosinophilic infiltration with abscess formation. There is no evidence of tuberculosis or neoplastic lesions.

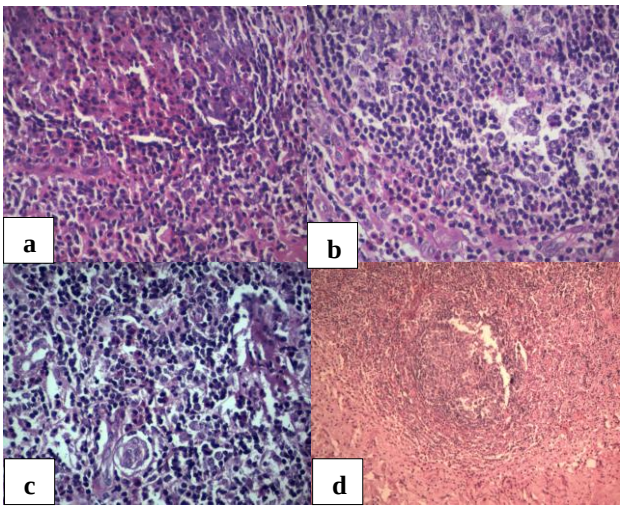

Figure 2: (a) Extensive eosinophilic infiltration forming an eosinophilic abscess (H&E×40), (b) hyperplastic follicles and diffuse eosinophilic infiltration (H&E × 40), (c) hyperplastic follicles and diffuse eosinophilic infiltration (H&E×40), and (d) hyperplastic follicles and diffuse eosinophilic infiltration (H&E×10).

Clinical evidence of swelling in the parotid region, along with marked lymphadenopathy in the head and neck area, peripheral eosinophilia, elevated serum IgE levels, and characteristic histopathological features, led to the diagnosis of Kimura's disease. The patient was started on prednisolone at a dosage of 1 mg/kg/day with close monitoring and other supportive treatments. Remarkable clinical improvement was observed, evidenced by a substantial reduction in the size of the swelling and lymph nodes by more than 50%.

DISCUSSION

Kimura's disease is a rare condition that predominantly affects Asian males, with sporadic cases reported worldwide.[4] It shows a significant male predominance, with a ratio ranging from 3.5 to 7:1, and usually presents during the third decade of life. Chen et al suggested that, although rare, Kimura's disease should be considered in the differential diagnosis of any lymph node showing eosinophilic infiltrate and prominent follicular hyperplasia, irrespective of the patient's race.[5,6]

The precise cause and pathogenesis of Kimura's disease remain unclear, but it is thought to result from a self-limited allergic or autoimmune response triggered by an unidentified persistent antigen. Research suggests that CD4+ T cells, particularly CD4 T-helper 2 (Th2) cells, and their cytokines—such as granulocyte-macrophage colony-stimulating factor, tumor necrosis factor-α, IL-4, IL-5, eotaxin, and RANTES—contribute to lymphoid follicle formation and elevated IgE levels.[7] Additionally, clonal T-cell populations have been linked to disease development and recurrence.[8] The immune response associated with Kimura's disease makes patients susceptible to allergic conditions such as asthma, chronic urticaria, pruritus, and rhinitis. The condition involves eosinophilia and elevated serum IgE levels, indicating an allergic or hypersensitivity process, although the specific antigens remain unidentified. Current understanding suggests that Kimura's disease represents an unusual allergic response involving Th2 cytokines.[9]

Clinically, Kimura's disease typically presents with one or more subcutaneous indolent nodules, peripheral eosinophilia, and elevated serum IgE levels. These nodules are usually painless, slowly enlarging, and located predominantly in the head and neck region, occasionally causing pruritus or pain.[3] Lymphadenopathy and enlargement of salivary glands may also be observed. In some cases, renal involvement can manifest as proteinuria and nephrotic syndrome. Lesions primarily affect subcutaneous soft tissues around the postauricular region and may occasionally involve salivary glands and lymph nodes.[10] Rarely, Kimura's disease has been reported in other sites, including the oral cavity, conjunctiva, eyelid, tympanic membrane, skeletal muscle, prostate, kidney, peripheral nerves, and epiglottis.

Characteristic histological findings include preserved nodal architecture, germinal center hyperplasia, eosinophilic infiltration, and postcapillary venule proliferation. Sclerosis, increased vascularization, proteinaceous deposits in germinal centers, micro-abscesses, and peripheral blood eosinophilia are commonly observed. Approximately 12% of patients' exhibit associated renal disease characterized by proteinuria.

Imaging techniques such as computed tomography (CT) and magnetic resonance imaging (MRI) are useful for assessing the extent and progression of the disease, particularly lymph node involvement. Fine-needle aspiration cytology can provide diagnostic insights in certain cases. Definitive diagnosis requires histologic examination of excised lesions, which reveal marked hyperplasia with eosinophilic infiltration, vascularization of the paracortex, and hyaline material deposition within follicles. Immunostaining may show IgE deposition around follicles and on non-degranulated mast cells.[7] Although intense eosinophilia and focal necrosis might resemble parasitic infection, no parasites are identified. The polymorphous infiltrate with eosinophilia and the presence of giant cells may raise suspicion of Hodgkin's disease, particularly on fine-needle aspiration, necessitating excisional biopsy if initial investigations are inconclusive. Persistent nodal architecture with prominent

germinal centers, eosinophilic infiltration, and postcapillary venule proliferation are consistent features.[11]

Differential diagnosis

Involving the parotid gland: Mikulicz's disease, infective parotitis, salivary gland tumors, and Sjögren's syndrome.

Lymphoid proliferation and eosinophilia: Hodgkin's lymphoma, Langerhans cell histiocytosis, and parasitic lymphadenitis.

Additional differentials include angiolymphoid hyperplasia with eosinophilia (ALHE) which is typically presents as a subcutaneous mass in the head and neck region. Microscopic findings may include aggregates and lobules of vascular endothelium lined by plump cuboidal or hobnail endothelial cells, often involving large muscular vessels. Regional lymphadenopathy, serum eosinophilia, and elevated IgE levels are uncommon in ALHE. Others include epithelioid hemangioma, angiofollicular lymphoid hyperplasia, low-grade angiosarcoma, atypical or pseudo pyogenic granuloma, and eosinophilic granuloma of the soft tissues.[6,12]

Treatment strategies vary, with surgical excision being the preferred method, often complemented by postoperative corticosteroid therapy. Systemic steroids can effectively control disease progression but may lead to relapse upon withdrawal.[13] Radiation therapy is considered for steroid-resistant lesions, though it carries risks such as xerostomia. Due to high recurrence rates after surgery, alternative therapies are being explored with varying success, including all-trans retinoic acid, leukotriene receptor antagonists like montelukast, and H_1 receptor blockers.

CONCLUSION

Kimura's disease is documented in only a limited number of cases within medical literature. It remains largely unrecognized among healthcare professionals, often evading detection or being misdiagnosed for years as other ailments. Its rarity and unusual presentation pose significant challenges to diagnosis. However, a careful history and clinical examination can provide clues, with confirmation typically relying on histopathological examination. Excision biopsy of the lesion remains the primary diagnostic tool for Kimura's disease. Therefore, Kimura's disease should be considered in the differential diagnosis of parotid gland swellings and conditions related to non-specific lymphadenitis. Despite recent advancements in understanding its clinical features, laboratory findings, and imaging, further investigation modalities are needed to overcome current study limitations. Despite its inflammatory nature, determining the most effective treatment approach for Kimura's disease remains unpredictable. Thus, diagnosing and managing this condition continues to present a significant challenge for both clinicians and patients. While Kimura's disease does not typically lead to complications, accurate diagnosis is crucial for effective management. Conservative management followed by radical treatment and regular follow-up is essential to prevent recurrence, as recurrence rates following treatment may be as high as 62%.

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