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Case Report

A comprehensive case report of kimura's disease: diagnosis and treatment insights

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ABSTRACT

Kimura's disease is a rare chronic inflammatory disorder primarily affecting the head and neck region. We present a case of a 35-year-old male patient with swelling over the right preauricular region and right eyebrow for 5 months. The patient's blood investigations revealed elevated eosinophil count, increased absolute eosinophil count, and significantly elevated serum IgE levels. Ultrasonography suggested lipomatosis, while fine-needle aspiration cytology (FNAC) was inconclusive. Excisional biopsy of the swellings confirmed the diagnosis of Kimura's disease based on histopathological findings of hyperplastic lymphoid follicles with micro abscesses. The patient was initiated on intravenous dexamethasone, resulting in a reduction in swelling size and a subsequent decrease in serum IgE levels and eosinophil count. This case report highlights the clinical presentation, diagnostic challenges, and therapeutic response of Kimura's disease.

Keywords: Kimura's disease, Lipomatosis, Eosinophilia, Hyperplastic lymphoid follicles

INTRODUCTION

Kimura's disease (KD) is an uncommon, non-malignant, chronic inflammatory condition characterized by the presence of subcutaneous nodules, swollen lymph nodes, and elevated levels of eosinophils. It was initially documented by Kimura and colleagues in 1948 and primarily affects individuals of Asian ancestry, with a higher occurrence among males.[1] The typical clinical manifestations involve painless, solid swellings beneath the skin in areas such as the preauricular, parotid, or supraorbital regions.[2]

In India, KD is infrequent, with only 200 cases reported globally based on histopathological confirmation.[3] It predominantly affects young adults, with the majority of patients falling between 20 and 40 years of age.[4] Commonly affected sites include the areas around the ears, groin, orbit, and eyelids.[5] KD often coincides with renal disease, with an incidence ranging from 10% to 60%.[6] The exact cause of Kimura's disease remains uncertain, although it is thought to involve a combination of immunological, infectious, and genetic factors.[7,8] This case study highlights the clinical features, diagnostic process, and management of a patient with Kimura's disease, emphasizing the difficulties in diagnosing the condition and the importance of early intervention.

CASE REPORT

A 35-year-old male patient presented with a chief complaint of swelling over the right preauricular region and right eyebrow persisting for the past 5 months. The patient was previously healthy and had no significant medical or surgical history.

The swellings gradually increased in size without associated features, and there was no involvement of other body parts. Physical examination revealed a diffuse swelling measuring 5×5×0.5 cm over the right preauricular region and a well-circumscribed swelling

measuring 2×2 cm on the right eyebrow. The swellings were firm, non-tender, and mobile.

Investigations

Blood investigations showed a raised eosinophil count of 25%, with an absolute eosinophil count of 1500 /cumm. Serum IgE levels were highly elevated at 14352.5 IU/ml. Ultrasonography (USG) was suggestive of lipomatosis. Fine-needle aspiration cytology (FNAC) was inconclusive, revealing cellular smears with nucleated and anucleate squamous cells, few cyst macrophages, and abundant lymphocytes against amorphous necrotic blood-filled material.

Histopathological examination

Given the inconclusive FNAC findings, excisional biopsy of the swellings was performed. Histopathological examination revealed hyperplastic lymphoid follicles with micro abscesses. There was no evidence of granuloma or malignancy, confirming the diagnosis of Kimura's disease involving the parotid and supraorbital regions.

Treatment and follow-up

After the definitive diagnosis of Kimura's disease, the patient was initiated on intravenous dexamethasone 8 mg twice daily for 2 weeks. The size of the swellings started to decrease gradually during the course of treatment. Repeat serum IgE levels showed a serial decrease, measuring 12489 IU/ml and then 11200 IU/ml.

The absolute eosinophil count decreased to 238 and 189/cumm. At the time of discharge after 2 weeks of treatment, the size of the swelling had significantly reduced, indicating the effectiveness of the initial treatment in controlling the disease.

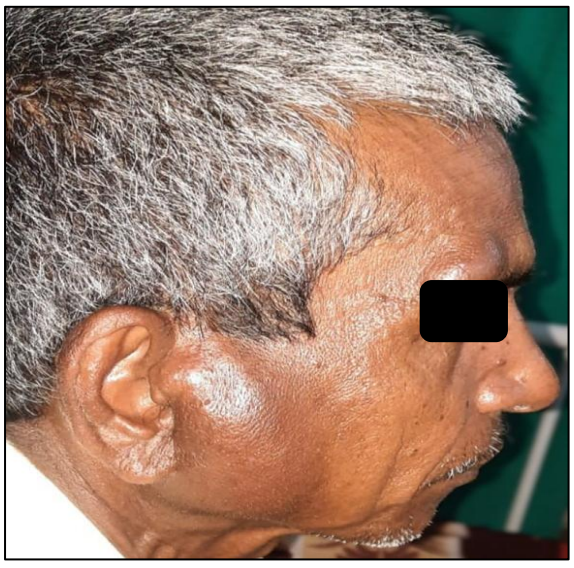

Figure 1: Other complications.

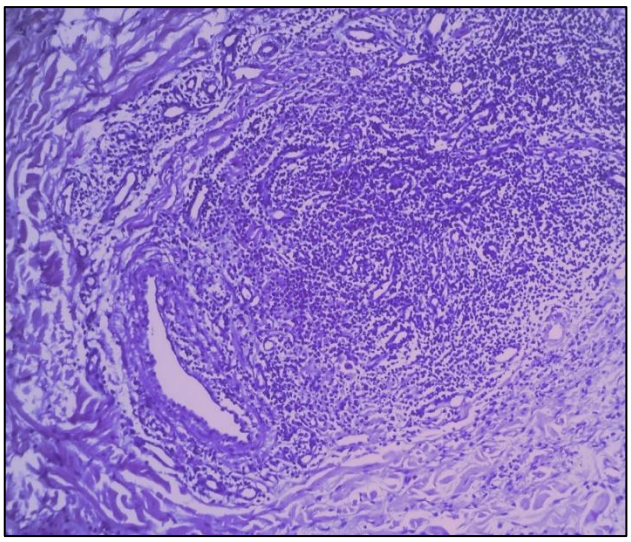

Figure 2: Histopathological examination of biopsy specimen showed hyperplastic lymphoid follicles with micro abscesses typical of kimura's disease.

DISCUSSION

Kimura Disease is an uncommon chronic inflammatory disorder that was initially described in the Chinese literature as "eosinophilic hyperplastic lymphogranuloma" and later known as Kimura's disease after its description in the Japanese literature.[1]

The disease is more frequently observed in the second and third decades of life, with a slight male predominance.[1] Various studies have shown typical Kimura manifestation, and so is our case. Here, this case report presents the clinical features, diagnostic workup, and management of a 35-year-old male patient with swelling over the right preauricular region and right eyebrow for the past 5 months.[2,9-12]

Although the exact etiology and pathogenesis of Kimura's disease remain unclear, it is believed to involve a self-limited allergic or autoimmune response triggered by an indefinite persistent antigenic stimulus.[7,8] The clinical presentation of Kimura's disease is characterized by the expansion of subcutaneous nodules in the head and neck region, commonly associated with regional lymphadenopathy.[2] Additional sites of involvement include the salivary glands, orbit, eyelid, palate, pharynx, axilla, groin, and arm.[5,8] In our case, the swelling started over the right preauricular region and right eyebrow, gradually increasing in size, without associated features or swelling in other parts of the body. This was consistent with the finding of similar cases.[9,10]

Local examination of our case revealed a diffuse swelling over the right side preauricular region and a separate swelling of 2×2 cm on the right eyebrow. These swellings were well circumscribed, firm, non-tender, and mobile. This was in accordance with the findings of case report

by Alshaibani et al, where palpable mobile upper medial mass (1×2 cm) over the left eyebrow was observed.[9]

One of the hallmarks of Kimura's disease is the association with elevated eosinophils and IgE levels in peripheral blood.[5,8] Cases reported raised Eosinophil levels up to 24% and 11.7% in studies by Dhingra et al and Malhotra et al respectively.[2,11] In our case, blood investigations revealed a raised eosinophil count of 25% and an absolute eosinophil count of 1500/cumm, indicating eosinophilia. Additionally, the serum IgE level was significantly elevated at 14352.5 IU/ml, further supporting the diagnosis.

Laboratory findings, along with the characteristic clinical presentation, aid in the differential diagnosis of Kimura's disease from other inflammatory, neoplastic, and infectious conditions. The disease should be distinguished from angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin's disease, Kaposi sarcoma, eosinophilic granuloma, Castleman's disease, tuberculosis, dermatopathic lymphadenopathy, lymphadenopathy of drug reactions, and parasitic lymphadenitis, among others.[2,9,10]

Histopathological examination reveals distinctive features of Kimura's disease, such as intact nodal architecture, prominent germinal center hyperplasia, infiltration of eosinophils, and proliferation of postcapillary venules. Other possible findings include sclerosis, polykaryocytes, vascularization of germinal centers, proteinaceous deposits, eosinophilic abscesses, and reticular IgE deposition within germinal centers. While the nodal architecture is usually preserved, it is common to observe capsular fibrosis with subcapsular sinusoid obliteration and involvement of the surrounding soft tissues.[2,8,9] Histopathologic findings in our case revealed hyperplastic lymphoid follicles with micro abcesses suggestive of kimuras disease of parotid and supraorbital region. Similar results were reported in studies by Alshaibani et al, Zhao et al, Dhingra et al, Malhotra et al and Qureshi et al.[2,9-12]

Imaging studies, such as computed tomography (CT) and magnetic resonance imaging (MRI), can provide valuable information in diagnosing and staging Kimura's disease. They help assess the extent of disease involvement, including lymph node enlargement and the presence of lesions in other sites such as the orbit.[2,9,10]

Treatment options for Kimura's disease include surgical excision, systemic steroids, and radiation therapy.[2,10] Surgical resection is considered the first-line therapy, especially for localized masses.[13] However, recurrence rates after surgical excision have been reported to be as high as 25%.[9,10] Systemic steroids, such as oral prednisolone, have shown efficacy in controlling disease progression and reducing symptoms. However, abandonment of steroids can often lead to relapse. Radiation therapy is reserved for cases resistant to steroids or those with recurrent or unresectable lesions.[10]

Renal involvement is a significant complication of Kimura's disease, occurring in a considerable proportion of patients. It manifests as extra membranous glomerulonephritis and nephrotic syndrome. While renal biopsies have reported findings consistent with mesangial proliferative glomerulonephritis and membranous nephropathy, our case did not undergo a renal biopsy. Instead, an excisional biopsy of the mass was performed, and the histopathology report disclosed lymphoid follicles with germinal center hyperplasia, fibrosis, plasma cells, interfollicular eosinophils, and eosinophilic abscesses, which were consistent with Kimura's disease.[9,10]

CONCLUSION

Kimura's disease is a rare chronic inflammatory disorder primarily affecting the head and neck region. This case report highlights the clinical presentation, diagnostic challenges, and therapeutic response in a patient with Kimura's disease involving the parotid and supraorbital region. Early recognition, accurate histopathological diagnosis, and prompt initiation of corticosteroid therapy is crucial in managing this rare condition.

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