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CASE REPORT

Indian male with recurrent Kimura's disease: A case report

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Abstract

Kimura's disease (KD) is an inflammatory disease of unsure etiology which presents as a painless subcutaneous swelling in the head and neck region that involves major salivary glands and regional lymph nodes. KD mainly had seen in head and neck area, and typically presents as tumor-like lesions that could be easily misdiagnosed. The maxillofacial involvement of KD is scarce. Kimura disease is occasionally puzzled with angiolymphoid hyperplasia with eosinophilia, which is usually associated with skin of the head and neck region. The anatomic sites of involvement of KD include posterior-auricular, cervical, and inguinal, epitrochlear lymph nodes, and salivary gland involvement. Here, we report a case of a 33-year-old Indian patient with KD who presented with unilateral nodular swelling in the right cheek region. The diagnosis of KD was made due to the previous history of surgeries, characteristic histopathologic findings along with peripheral eosinophilia, and high serum immunoglobulin E levels.

Keywords: Eosinophilia, Kimura, nephrotic syndrome, parotid gland

Introduction

Kim and Szeto in 1937 diagnosed Kimura's disease (KD).[1] It is a rare chronic inflammatory disorder of unidentified etiology with <120 cases reported worldwide.[2,3] The disease was first described in the Chinese copy as "eosinophilic hyperplasic lymphogranuloma."[3] This disease is widely known after a systematic description by Kimura in 1948 in Japan.[4] The disease is usually associated with the cutaneous and subcutaneous tissue characterized by single or multiple swellings; the patients are usually asymptomatic but sometimes experiences the pain and 10–60 % of them may present renal disorders, namely, nephrotic syndrome (10–12%), characterized by clinically relevant proteinuria (12%–16%). The pathogenesis for this renal impairment is an immune complex-mediated damage or helper T cells dominant immune response disorders.[5] Previous reports have noted that KD involves the parotid tissues and is clinically misinterpreted as a swelling of salivary gland or even a malignancy.[6]

There are many researchers conducted on etiology of KD. To diagnose KD before surgery is quite challenging as clinical examination alone may not give a precise diagnosis.[7] In the present case of KD affecting the preauricular region, the diagnosis was based on the previous history of surgeries, biopsy, and characteristic histopathology findings in combination with peripheral eosinophilia and elevated serum immunoglobulin E (IgE) levels.

Case Report

A male patient of age 33 years came with a chief complaint of a slow-growing swelling on the right side of the face for 3 years. It was initially a small swelling and gradually increased to present size with no other symptoms. There was a history of a similar swelling behind the ear 5 years back which was operated, and the diagnosis rendered was KD based on histopathology report. On examination, the surface of swelling was smooth without any changes in the overlying skin and no local rise in temperature. The manifestations of itching, eosinophilia, and tissue eosinophilic infiltration are characteristic features of an on-going allergic inflammatory reaction. His medical history was normal. The swelling extends anteroposteriorly 2 cm from the commissure of the lip to 2 cm to external auditory meatus. The size of the swelling didn't change during the mastication. On palpation, the lesion was firm in consistency. The right multiple submandibular and 3–4 cervical lymph nodes were palpable and non-tender. Based on the clinical examination, we rendered as a provisional diagnosis of masseter hypertrophy with tuberculosis and KD as differential diagnosis.

The blood investigation and fine needle aspiration cytology were done, and there was increase in eosinophilic count and reactive lymphoid hyperplasia was noted. HIV, hepatitis B surface antigen, and hepatitis B virus were negative. Monteux test was negative with 5TU PPD. The chest X-ray appeared normal.

On ultrasound examination of the lesion revealed multiple discrete and confluent lymph nodes at the right intraparotid and submandibular region. Few lymph nodes were also seen at the right level II and III with short axis.

Based on the clinical examination and investigation, the diagnosis of recurrent KD in the right preauricular region was rendered. We prescribed him antihistamine medication Tab. Montek LC for 1 month, as we hypothesized that an antiallergic agent, such as levocetirizine might be beneficial with Montelukast combination, as Montelukast blocks the leukotriene receptor which controls the inflammation and improves the quality of life. Later, the patient was taken to a physician, and he was put on a course of anthelminthic drugs for the upregulation of the host immune response and on immunosuppressive drug Prednisolone for 3 months, and currently, he is under follow up and there is no change in the size of the swelling, and now, the patient has been advised for the surgical excision as that is best treatment modality.

Discussion

The diagnosis of KD is mainly based on clinical findings, excisional biopsy along with serum analysis showing peripheral eosinophilia and high IgE. The etiology for KD is unclear. The investigations such as CT and MRI examination shown no exact intensity or signal changes. Hence, the oral physicians play an important role in diagnosis as it most commonly manifests in the head and neck region.[4]

Although numerous efforts have been made to establish a definitive treatment for KD, not a single modality has been proved effective. The usual methods of treatment include drug administration such as systemic corticosteroids and immunosuppressive therapy including tacrolimus, radiotherapy, surgery, and photodynamic therapy. To date, conventional surgical excision is considered the most effective as it treats and at the same time provides a sample for histopathological analysis to arrive at accurate diagnosis.[8] According to the study conducted by Ma *et al*., radiation therapy is the primary treatment modality for KD. However, this is in contradiction with other studies which state that the potential risk of radiotherapy-related malignancy surmounts the need for the treatment of this benign disease.[9]

Wang *et al*. reported a case of a patient treated with X-ray radiation therapy. There was a significant reduction in size of the masses after treatment. No recurrence was noted throughout the 68-month follow-up period [10] Tsukagoshi *et al*. reported a case of KD with bronchial asthma and hyperimmunoglobulinemia E. He was treated with an antiallergic drug (suplatasttosilat) with good quality response.[8]

The KD has excellent prognosis, but it is highly recurrent. The recurrence of disease is affected by duration, number, size and boundaries of the lesion, eosinophil count, and serum IgE levels. Shenoy *et al*. reported a recurrent case of KD where there was approximately 80% reduction in size of the lesion within 2 weeks of cyclosporine therapy (5 mg/kg/day in two divided doses) for 4 weeks. It was then tapered by halving the dose every 4 weeks; the complete treatment duration is 14 weeks.[11]

In the present case, there was 3 times recurrence noted. Various medicaments such as leflunomide and methylprednisolone have been tried to prevent the recurrence, and currently, the patient is under follow-up. There are also reports of renal involvement with KD in the form of nephrotic syndrome which worsens the condition of the patient so continuous follow-up is required.[3]

Conclusion

KD is a rare chronic inflammatory benign condition. It is seen mainly among Asian men. It manifests as painless soft tissue swellings with head and neck lymphadenopathy in the head and neck region. Serum analysis revealed peripheral eosinophilia and elevated serum IgE levels. Its diagnosis is confirmed mainly by histopathology. Surgery is the primary treatment modality, but its recurrence rate is high. Systemic steroids are administered to prevent recurrence. Frequent follow-up is required to monitor the condition.

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