Case Report

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Kimura Disease: A Case Report in an Unlikely age group

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ABSTRACT

Kimura disease is a rare chronic inflammatory disorder of unknown etiology. The disease is primarily seen in young Asian males and is characterized by painless subcutaneous swelling. It is often accompanied by renal involvement, predominantly manifesting as nephrotic syndrome. We describe a case of Kimura disease in a 69-year-old Indian male, presenting with left postauricular swelling. Clinical and histopathological characteristics are described along with a brief review of the literature.

Introduction

Kimura disease is a rare benign chronic inflammatory disorder which usually presents as subcutaneous mass in the head and neck region.[1],[2] It was first described in 1937 by Kim and Szeto in the Chinese literature as “eosinophilic hyperplastic lymphograuloma”[10] but was not widely recognized until Kimura described it in Japanese literature in 1948. It is frequently associated with lymphadenopathy, salivary gland involvement, peripheral blood eosinophilia and raised immunoglobulin E (IgE) concentration in the blood. In 70% of cases, Kimura disease occurs in young and middle-aged men and is endemic in Asia. [3]

Coexisting renal disease has an incidence ranging from 10% to 60%. [4]. Kidney disease is often associated with autoimmune diseases such as ulcerative colitis. Immunocomplex-mediated damage, especially a defect in Th2-cell immunoregulation might be involved in the disease pathogenesis and renal involvement. [5, 6]

Because the presentation has a head and neck mass as its presentation, Kimura disease needs to be differentiated from certain malignancies. The peripheral eosinophilia can also mimic an allergic reaction or a parasitic infection.

In the present case report, we describe the case of a 69-year-old male presenting with a postauricular swelling.

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Case Presentation

A 69-year-old male patient presented to the Ear Nose Throat Outpatient Department (OPD) with the complaint of a left postauricular swelling for the past 7 years which was insidious in onset and gradually progressive. He had no history of any constitutional symptoms. On examination, USG neck showed an ill-defined solid hypoechoic heterogenous lesion of size 4.2 x 2.8 x 1.5 cm in the posterior auricular region. Multiple mildly enlarged hypoechoic oval shaped lymph nodes surrounding the lesion were noted, the largest being 2.1 x 1.5 cm. The skin overlying the swelling was not involved. The rest of the examination was unremarkable.

Core biopsy (done in July 2022) showed loose fibro collagenous tissue with intermittent lymphoid aggregates admixed with many eosinophils. Epitheloid granulomas or caseous necrosis were not seen. However, lympho-proliferative disorder could not be ruled out. Histopathological sections studied from lymph node showed partial effacement of architecture.

The patient was treated with surgical excision of the swelling. Excision biopsy sections revealed benign epidermis of skin. The dermis and underlying tissues revealed proliferation of enlarged but well-spaced reactive lymphoid follicles with intervening areas of hyalinization. [Fig 1] The lymphoid follicles, as well as hyalinized areas, showed numerous eosinophils and occasional eosinophil micro abscess formation. The hyalinized areas showed prominent vascular proliferation. Plasma cells and neutrophils were also seen. There was no evidence of obvious Reed-Sternberg cells or abnormal cells.

On immunohistochemistry (IHC), CD20 and CD3 were expressed in the "B" and "T" zones respectively. PAX5 was expressed in the "B" cells. MUM1 was expressed in plasma cells. CD15 and CD30 did not highlight any Reed Sternberg cells. S-100, CD1A and Langerin did not highlight any abnormal cells of Langerhans cell histiocytosis. Ki-67 was high in the follicles only. IHC ruled out lympho-proliferative disorder and Langerhan's cell histiocytosis. Although peripheral blood eosinophilia and raised serum immunoglobulin E levels are constant features of Kimura disease, the haematological parameters of the patient were within normal limits. Renal blood parameters were normal on investigation. Urine examination showed no proteinuria or other abnormalities.

Discussion:

Kimura disease, a chronic inflammatory disease, presents commonly as subcutaneous nodules in the head and neck. Often, unilateral and regional lymphadenopathy with or without involvement of salivary glands is also seen. [7, 8] There might be bilateral involvement in some cases. Apart from the commonly involved sites such as periauricular, groin, orbit and eyelids, sites like axilla, pharynx and palate have also been reported to be involved. [9] Although the disease can occur at any age, most patients are in the 2nd and 3rd decades of life. Male preponderance is seen, with a ratio of 3:1. [10]

The exact cause and pathogenesis of Kimura's disease are still unclear although it might be a self-limited allergic or autoimmune response. A disorder of type 2 T-helper cells, affecting the production of cytokines and causing eosinophil activation along with an IgE response, has been proposed. The immune reaction that is believed to be the root of Kimura's disease also predisposes the patient to allergic conditions like asthma, chronic urticaria, pruritus and rhinitis and is thought to be the probable cause of the renal involvement. [11] The constant histological features include, preserved nodal architecture, florid germinal centre hyperplasia, eosinophilic infiltration and postcapillary venule proliferation. Renal biopsy has shown various forms of renal pathology- membranous nephropathy being the most common.

Most patients have a prolonged course with gradual increase in the swelling while occasionally, spontaneous resolution is seen. [12] Diagnosis of KD is not easy and differential diagnosis includes angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin's disease, Kaposi sarcoma, eosinophilic granuloma, tuberculosis, epithelioid hemangioma, Castleman's disease, dermatopathic lymphadenopathy, lymphadenopathy of drug reactions, parasitic lymphadenitis, and many more. [13]

Treatment for Kimura disease includes surgical resection and regional or systemic steroid therapy. Cytotoxic therapy and radiation have also been utilized. The disease has an excellent prognosis although it may recur locally [14]. Sun *et al*. [15] reported that Imatinib—previously useful for treatment of hypereosinophilic syndrome, may work by selectively blocking protein tyrosine kinases. Hence, might be an effective drug for treatment of this disease.

Conclusion:

Relevance of this case is due to rarity of Kimura disease which mimics neoplastic conditions. Additionally, we can observe that the patient described above does not belong to the common age bracket of presentation and did not show any changes in blood eosinophil or IgE levels and no coexisting renal involvement, which are important discrepancies to be noted. Kimura disease should be considered in a differential diagnosis in patients presenting with head & neck swelling and lymphadenopathy and should be investigated accordingly.

Ethical Considerations

Compliance with ethical guidelines

Case report is written as per, "CARE GUIDELINES FOR CASE REPORT" in compliance with ethical guidelines.

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Conflict of interest

The authors declared no conflict of interest.

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