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Inguinal Lymphadenopathy in a Patient with Kimura Disease: A Rare Presentation of a Rare Disease

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Authors' contributions

This work was carried out in collaboration among all authors. Author TP conceptualized and wrote the first draft of the manuscript. Authors SP and DT obtained the patient data and consent. Author HP did the literature searches. Author TP and SP wrote the final manuscript. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Kimura disease is an extremely rare idiopathic condition. It is a chronic inflammatory disorder typically seen in Asian males in younger age groups. Its classical features include painless lymphadenopathy or subcutaneous swelling usually in head and neck, eosinophilia and raised levels of IgE. However, our case is of a 45 year old male who presented with right sided inguinal swelling with diagnostic dilemma that was later diagnosed as Kimura disease after invasive histopathological analysis. Here, we have described this case to shed some light on possibility of such presentation of this rare disease.

Keywords: Kimura disease; rare disease; surgery; lymphadenopathy; renal involvement; histopathology.

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1. INTRODUCTION

Kimura disease is a chronic inflammatory disorder with angiolymphatic proliferation, usually affecting young men of Asian race but is rare in western countries. It is often accompanied by nephrotic syndrome and is a rare, chronic inflammatory disorder of unknown cause. Chen et al. reported 21 histopathology specimens conducted in the United States by the US Armed Forces Institute of Pathology and concluded that, though rare, if clinically suspected, Kimura disease should be included in the differential diagnosis for people of any racial group. It is usually seen in young adults, with most patients being aged between 20 and 40 years; men are affected more commonly than women, with a 3:1 ratio. The etiology of this disease is still unknown but it may be due to impairment or interference with immune regulation, atopic reaction to a persistent antigenic stimulus by arthropod bites, virus, and neoplasm. The most interesting hypothesis suggests Candida acting as a source of persistent antigenaemia, although neither hyphae nor spores have been isolated. The disease is manifested by hyperplasia of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate suggest that KD might be a kind of hypersensitivity reaction. T-helper 2 (Th2) lymphocyte might play a role. Patients may present with a solitary enlarged painless lymph node or generalized lymphadenopathy (67% to 100%). It classically presents as a nontender subcutaneous swelling in head and neck region, predominantly in preauricular and submandibular area. It may be associated with lymphadenopathy (both local and distal), marked peripheral eosinophilia, and an elevated IgE level. Coexisting renal disease is common, with an incidence ranging from 10% to 60%, while 10% to 12% of patients may suffer from nephrotic syndrome characterized by clinically relevant proteinuria in 12% to 16% of cases. Renal impairment is probably due to immunocomplex-mediated damage or Th2-dominant immune response disorders [1].

Diagnosis of this disease is difficult and often warrants biopsy or excision of the mass for histopathological investigation. Here, we report an extremely rare case of a patient who presented with an inguinal swelling who was later diagnosed as Kimura disease.

2. CASE REPORT

2.1 Presentation

A 45 year old patient presented to our hospital with a complaint of right sided inguinal swelling since 3 years which was gradually progressive and insidious in onset with associated complaint of occasional itching and discomfort. On examination, an 8cm×5cm globular, firm, non-tender and relatively mobile mass was palpated in the right inguinal region. Skin over the swelling was normal and not adherent to the mass. There was no other significant finding on examination.

2.2 Investigations

Hematological examination revealed Hb 12.7 gm%, TLC 10,600 cells/cumm with 40% Eosinophils, 38% Neutrophils, 21% Lymphocytes and 1% Basophils. Platelets were 299,000/cumm. Serology reports for HIV, Hepatitis B and C were negative. Urine analysis was within normal range. Local part ultrasonography showed a 10cm×7cm ill defined heterogeneously hypoechoic lesion with internal vascularity suggestive of a conglomerated lymph node mass with possible neoplastic etiology. CT of the local part demonstrated lymph node enlargement encasing the femoral and profunda femoris arteries. Fine needle aspiration was done and smear was scantily cellular with lymphocytes in various stages of maturation and aggregates of histiocytes in background of blood cells that gave impression of reactive lymphadenitis.

2.3 Management

Excision biopsy was performed and two samples consisting of creamish 8cm×5cm×1.5cm homogenous soft tissue mass and 3 cm×1.5 cm×1 cm creamish soft tissue mass were obtained respectively. Histological study showed fibrofatty and fibroconnective tissue with focal lymphoid aggregates with vascular proliferation. There was marked presence of Eosinophils and neutrophils without evidence of necrosis. Final impression suggested a non-neoplastic lymphoid lesion with strong possibility of Kimura disease. Conservative management was done with active follow up for disease progression. He had been followed up for past 5 months and is on antihistamine medications without any new symptom or disease progression.

3. DISCUSSION

Kimura Disease (KD) is a rare chronic inflammatory disorder, which was first described in 1937 by Kim and Szeto in the Chinese literature as eosinophilic hyperplastic lymphogranuloma and has been known most often as Kimura's disease since its description by Kimura et al. in the Japanese literature in 1948. This disease is endemic in middle-aged Asian males and rarely seen sporadically in non-Asian population [1]. Multiple theories have been given but the etiology of this disease still remains unknown. Coexisting renal disease is common, with an incidence ranging from 10% to 60%, while 10% to 12% of patients may suffer from nephrotic syndrome characterized by clinically relevant proteinuria in 12% to 16% of cases [1,2]. Renal impairment is probably due to immunocomplex-mediated damage or to Th2-dominant immune response disorders.

Because of the rarity of the condition, Kimura disease poses difficulty in differentiating it clinically from relatively common conditions like Hodgkin's disease, tuberculosis, cylindroma, pyogenic granuloma, dermatofibrosarcoma protuberans and infectious lymph node enlargements until further investigations are done. Ultrasonography is initial investigation to be performed apart from routine blood investigations. Lymph nodes appear hypoechoic, round to oval in shape, solid and with usually normal surrounding tissues. It may be easily mistaken for diseases like tuberculosis or lymphoma. Thus, to exclude these conditions, histopathological analysis is required.

However differentiating it with from ALHE (Angiolymphoid hyperplasia with eosinophilia) can be very difficult. Clinically, both conditions present as soft tissue swellings arising in the head and neck region with prolonged indolent clinical course. Microscopically, both show eosinophilic infiltrates and vascular proliferations. But there are few characteristic and distinctive clinicopathologic features that differentiate the two entities. Kimura disease occurs predominantly in Asians, with a male predilection. Patients usually have peripheral eosinophilia and elevated serum IgE levels. The solitary lesions are mostly in the subcutaneous tissues, frequently associated with regional lymphadenopathy and salivary gland involvement. By contrast, ALHE occurs in all racial groups with a slight female predominance. Patients present with small, superficial dermal papulonodules, frequently erythematous, accompanied by bleeding, pruritus, and tumor growth. Regional lymphadenopathy, serum eosinophilia, and elevated IgE levels are rare [3]. Furtunately, this is not significant in clinical practice as the treatment strategy remains the same.

Treatment usually comprises of medical, surgical or radiological intervention. Surgery is useful for both diagnosis and treatment. It is particularly useful in lesions causing mass effect and compress surrounding tissues. It is usually preferred in single lesion, young patients and local recurrence. Medical treatment comprises of steroids that is useful in patients with renal involvement and multiple sites of lesions [4]. It helps in decreasing the size of the lesion along with its effect in reducing nephrotic syndrome symptoms. Sun *et al*., reported that Imatinib—previously to be useful for treatment of hypereosinophilic syndrome and may work by selectively blocking protein-tyrosine kinases—might be an effective drug for the treatment of the disease. Cytotoxic therapies are also sometimes used [5]. Finally, radiotherapy has been used to treat steroid resistant cases. Despite all these modalities available, our patient was treated surgically initially to reduce the size of lesion and then was followed up with antihistaminics for itching since past 5 months. There had been no new symptoms and the progression of the lesion is not occurring.

4. CONCLUSION

Kimura disease, a rare idiopathic chronic inflammatory disorder can be difficult to diagnose when the patient presents with unusual features like inguinal lymphadenopathy in our patient. This poses a challenge in differentiating Kimura disease from other relatively common pathologies of that area. It can be diagnosed only on the basis of histopathological examination and management is variable depending on its presentation. Hence, clinical suspicion has to be maintained to diagnose and treat this condition which can be developed by reporting cases like this and reviewing them. This becomes especially useful in the countries where the occurrence of such cases is very rare.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline patients consent and ethical

approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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