

Case Report

Kimura's Disease: A Rare Cause of Chronic Lymphadenopathy in a Child

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Submitted: 14 May 2014
Accepted: 24 Jul 2014

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Abstract

Kimura's disease is an uncommon entity that affects adults, with a predilection for the Asian population. This may rarely be encountered in children, and the knowledge of this fact is essential to rule out the remote possibility of Kimura's disease in children with a slow-growing painless mass in the head and neck region. In this case report, we document this disease in an 8-year-old boy with a slow-growing swelling in the right posterior auricular region.

Keywords: child, Kimura disease, lymphadenopathy, eosinophilia, hyper IgE

Introduction

Predominantly observed in young Asian males, Kimura's disease is an uncommon benign slow-growing chronic inflammatory disorder of unknown definitive aetiology. As a triad of painless subcutaneous masses in the head and neck region, it is characterised by blood and tissue eosinophilia with clearly elevated serum immunoglobulin E (IgE) levels. Although several cases have been documented in adult populations, it has been rarely reported in children, as the incidence is common in males in the age group of 20–30 years (1). In this case report, we document this entity in an 8-year-old child who presented with painless right postauricular lymphadenopathy.

Case Report

An 8-year-old male child, a firstborn to non-consanguineous parents, presented to the paediatric outpatient department with multiple swellings behind the right ear of two years' duration. There was no accompanying history of fever, weight loss, or night sweating. He complained of itching over the swelling for the past 3 months, which made his parents seek medical attention. He underwent blood investigations, Mantoux test, and multiple courses of antibiotics.

All the results of the investigations were found to be within normal limits, except for a mild eosinophilia. Because the swelling did not show any rapid increase, the parents did not consult any physician during the 2 years period. He did not have any other complaints and was otherwise well throughout this period. His medical and family histories were unremarkable. He had normal development.

General physical examination revealed lymphadenopathy in the right posterior cervical region. There were three posterior auricular lymph nodes in the right side, with the largest measuring 2 × 1 cm. The nodes were firm, non-tender, and non-matted, without any changes over the surface of the skin. No other significant lymph nodes were palpable in the other areas of the body. The rest of the clinical examination results were unremarkable.

Investigations showed a haemoglobin level of 13.1 g%, white blood cell count of 10 530 ($\times 10^9/L$; Polymorphs 42%, Lymphocytes 39%, Eosinophil 11%, and Monocytes 5.8%) and platelet count of 3.38 ($\times 10^9/L$). The IgE level was 218 IU/mL. Peripheral blood film showed normochromic normocytic red blood cells. His chest radiograph was normal. The Mantoux test result was negative.

Routine urine and renal function test results were also normal. His liver functions, renal parameters, serum electrolytes, uric acid, and LDH were within normal limits.

The largest lymph node was excised and subjected to histopathological examination.

The histopathological examination of lymph nodes showed a preserved architecture, with follicles showing prominent germinal centres, dilated sinusoids with hyperplasia of postcapillary venules with abundant eosinophilic infiltrates, along with many plasma and mast cells. Some of the eosinophils were seen infiltrating the germinal centres. Few multinucleated giant cells of the Warthin-Finkeldey type were observed in the germinal centres. Histomorphology of the lymph node was suggestive of Kimura's disease (Figures 1 and 2).

Considering that the patient was asymptomatic without systemic involvement, he was observed without any intervention after a detailed counselling was provided to the family and the lesion disappeared completely. He has been followed up regularly once every 3 months, and after 1 year of follow-up, no signs of recurrence were observed.

Discussion

The common causes of chronic lymphadenopathy in children are viral (Epstein-

Barr virus, cytomegalovirus, and human immunodeficiency virus), bacterial (tuberculosis and cat scratch disease), malignancies (Hodgkin's and non-Hodgkin's lymphoma, leukaemia, histiocytosis, neuroblastoma, and rhabdomyosarcoma), autoimmune conditions (systemic lupus erythematosus, hemophagocytosis, and Kawasaki). The other rarer causes are Kimura's disease, Rosai-Dorfman disease, Kikuchi's disease, and Castleman's disease (2).

An eosinophilic inflammatory condition that is characterised by painless, gradually growing soft tissue mass and lymphadenopathy of the head and neck region best describes Kimura's disease. Bilateral association is a rare instance. Shankar et al. reported that the disease primarily affect Asian males aged between 20 and 30 years.

The disease typically involves periauricular lymph nodes, as noted in our case. However, axillary and inguinal lymph nodes, parotid and submandibular salivary glands, and rarely, oral mucosa are other sites of occurrence. Other unusual sites include the auricle, scalp, orbit, parotid, and gluteal region (3). Although first described in China in 1937 by Kim and Szeto (4), the name of the disorder was given in 1948, when the vascular component was observed by Kimura and others, and referred to as an "unusual granulation combined with hyperplastic changes in lymphoid tissue" (5).

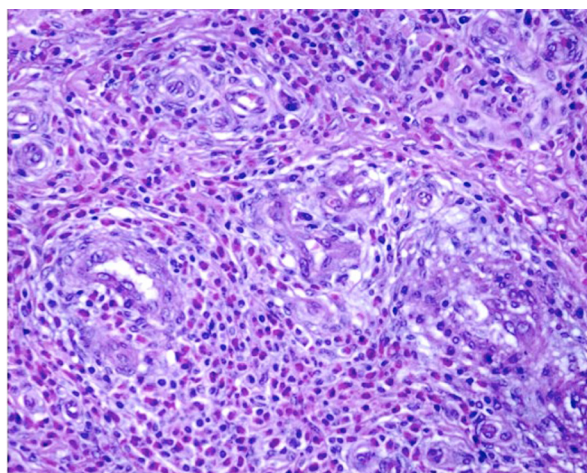


Figure 1: Lymph-node section showing infiltration by numerous eosinophils and vascular proliferation. Blood vessels are lined by prominent endothelial cells (hematoxylin and eosin stain, 200× magnification).

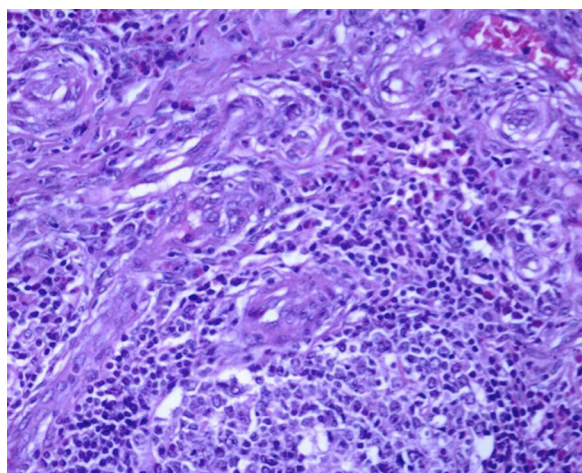


Figure 2: Prominent vascularisation observed in germinal centres of lymphoid follicles and interfollicular area showing filtration by many eosinophils (hematoxylin and eosin stain, 200× magnification).

Chen et al. (6), found only 7 children with Kimura's disease among all the children with head and neck masses from 1970 to 2002 in their institute. Vishwanatha et al. (7), reported only 18 children with Kimura's disease during the study period between 1998 to 2006 in India.

Although trauma, an allergic reaction, and an autoimmune process, have been implicated as the possible cause, the pathophysiological mechanism of Kimura's disease remains undetermined. Speculation of a viral or parasitic trigger that may alter T-cell immunoregulation resulting in the release of eosinophilic cytokines such as IL-4, IL-5, and IL-13 is trending. Immunohistochemical studies of the skin, lymph nodes, and peripheral blood have presented propagation of human leukocyte antigen DR CD4 cells. These release granulocyte macrophage colony stimulating factor and tumour necrosis factor, which in turn precipitate the high serum IgE level and marked eosinophilia (8). Other disorders such as eosinophilic granuloma, malignant lymphoma, salivary gland tumours, and Mikulicz's disease can be mirrored by Kimura's disease. Fine-needle aspiration cytology may not be conclusive, and excision biopsy is the diagnostic standard to avoid unnecessary and harmful interventions in a child with chronic lymphadenopathy. Preserved nodal architecture, follicular hyperplasia with reactive germinal centres, well-formed mantle zones, dense eosinophilic infiltrates, eosinophilic microabscess with central necrosis, Warthin-Finkeldey polykaryocytes, and proliferation of postcapillary venules in paracortex are the most common histological features of Kimura's disease (9).

A high prevalence of nephropathy has been reported to be associated with Kimura's disease. Matsuda et al. (10), reported 16% of cases, of which 78% had nephritic syndrome. Proteinuria appeared simultaneously with or later than the onset of skin lesions in most of the patients. Therefore, to exclude concomitant renal dysfunction and/or nephrotic syndrome, blood urea nitrogen, creatinine, and urinary protein levels ought to be obtained. In a series studied by Qunibi et al (11), 86 cases were reported to have renal involvement with membranous nephropathy followed by mesangial proliferative glomerulonephritis and minimal change disease. In our case, a thorough renal evaluation did not reveal any involvement both at the time of diagnosis or during follow-up.

The clinical course of Kimura's disease is generally benign and self-limited. Most cases

have a prolonged course with slow enlargement of the masses. While spontaneous involution is rare, malignant transformation has not been reported to date (1). In a prospective study by Vishwanatha (7), of 18 Kimura's disease patients for 9 years, only 3 had a recurrence and 15 were symptom-free by the end of 1 year.

Surgical biopsy is the most frequent diagnostic procedure, and excision at an initial presentation may be curative. Nevertheless, reappearance is a usual feature. In frequent relapses or cases complicated by nephrotic syndrome, systemic steroids may be indicated. If the Kimura's disease lesions are neither symptomatic nor disfiguring, observation is acceptable. In cases refractory to surgical and medical therapies, radiation is considered. Local irradiation (25–30 cGy) has been found effective for recurrent lesions not responding to either modality. Effective treatment with cyclosporine, azathioprine, pentoxifylline, and imatinib has also been reported (12).

Conclusion

Although rare, Kimura's disease should be regarded as a rare possibility of chronic lymphadenopathy in children, especially in Asian and developing countries, where tuberculosis is always suspected and empirical treatment is initiated without a proper diagnosis. We, therefore, document this case to create awareness among paediatricians about the rare occurrence of this condition in a child, which can be diagnosed conclusively by histopathological examination of the excised lesion.

Acknowledgement

None.

Conflict of Interest

None.

Funds

None.

Author's Contributions

Drafting of the article: LMS, BK

Critical revision of the article for the important intellectual content: VPN, SR, JXS

Final approval of the article: JXS

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