Kimura Disease Histopathological Study-A Case Report

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Abstract

Kimura disease is a rare form of chronic inflammatory disorder involving subcutaneous tissue, predominantly in the head and neck region and frequently associated with regional lymphadenopathy. The etiology of Kimura disease is still unknown but may be due to impairment of immune system. Kimura disease should be suspected when the clinical finding of painless unilateral cervical adenopathy and hyperesinophilia presented. Definitive diagnosis arise by histopathology examination, with hematoxylin-eosin staining showed follicular hyperplasia with perifollicular fibrosis, dense eosinophil infiltrate, and proliferation of capillary vessel lined by normal, flat, spindle-shaped endothelial cells. We reported A 37 years old man with history of soft tissue tumor on facialis dextra region, lymphadenopathy colli dextra, and exopthalmic bulbus oculi dextra over the last 10 years. Laboratory assessment on hematologic review showed that eosinophil 47.9 (10^3/ul).

Keywords: Kimura disease, case report.

INTRODUCTION

Kimura disease is a rare chronic benign disorder young man of Asian race, usually affecting in western countries [1, 2]. Kimura’s disease is usually seen in young adults, with most patients being 20-40 years of age. Men are affected more commonly than women, with a 3:1 ratio [3]. The major physical manifestation of the disorder is slowly enlarging subcutaneous masses, often in the head and neck area and usually in association with peripheral blood and tissue eosinophilia, incombination with markedly increased serum Ig E concentrations [1]. The etiology of Kimura Disease is still unknown but may be due to impairment or interference with immune regulation, atopic reaction to a persistent antigenic stimulus by arthropod bites, virus, and neoplasm [3]. Peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate suggest that Kimura Disease might be a kind of hypersensitivity reaction.

CASE REPORT

37 years old man presented to oncologic surgery department with history of soft tissue tumor on facialis dextra region, lymphadenopathy colli dextra, and exopthalmic bulbous oculi dextra over the last 10 years and diagnose by soft tissue tumor facial dextra with differential diagnose tumor of fascial dextra suspek hemangioma. Laboratory assessment on hematologic review showed that eosinophil 47.9 (10^3/ul), the other value is on normal range. On radiologic expertise of AP lateral skull x-ray note with soft tissue density regio mandibulae dextra and on MSCT scan evaluation note with soft tissue tumor regio facialis dextra, lymphadenopathy regio colli dextra, exopthalmic bulbous oculi dextra and retension cyst sinus maxillaris sinistra. Tissue was accepted from oncologic surgery department and evaluated on Patologi Anatomi laboratory department, we accepted about four divers tissue that some coated with skin, size of tissue about 10x7x4cm, 7x5x2cm, 7x3x2cm, and 4x2x1cm, soft to firm in consistancy. Histopathology examination, stain with hematoxylin-eosin showed on dermis and subcutis, hyperplasia follicles of varying sizes with prominent germinal centre and perifollikular fibrosis [Fig1]. Eosinophilic micro abscesses seen in focal areas, folliculolysis by eosinophils was also seen [Fig 2, Fig 3]. Many blood vessels with flat endothelial cells were seen and lined by normal, flat, spindle-shaped endothelial cells [Fig 4]. On immunohistochemically staining for CD3 and CD20 showed T-cells surrounding well-formed lymphoid follicles with germinal centers containing B-cells [Fig.5 and 6].
**DISCUSSION**

Kimura disease is a chronic inflammatory disorder involving subcutaneous tissue and lymph nodes predominantly in the head and neck region frequently involving periauricular and submandibular region [4]. More common among Asians, predominant in young men aged between 20–40 years [4]. The clinical presentation is characterized by triad of painless unilateral cervical lymphadenopathy or subcutaneous swelling, eosinophilia (98%) and markedly elevated serum IgE levels. The onset is insidious, manifests as a painless enlarging nodular mass located deep in the subcutaneous tissues and in almost all cases it involves regional lymphnodes. The lesions are single in 60% of cases, occasionally an isolated enlarged lymphnode may be the only presentation. Some cases can present with generalized lymphadenopathy. Peripheral blood eosinophilia and elevated serum IgE levels are the constant features [5]. Definitive diagnosis can be obtained by histological examination of the excised lesion [6]. Histopathology shows markedly hyperplastic follicles with reactive germinal centre. Diffuse eosinophilia, eosinophilic microabscess and infiltration of germinal centre sometimes resulting in folliculolysis. Vascular hyperplasia with normal, flat endothelial cells. Immunohistochemistry staining for CD3 and CD20 showed T-cells surrounding well-formed lymphoid follicles with germinal centers containing B-cells [7, 9].
The etiology of Kimura Disease is still unknown. Although infectious etiologies (human herpesvirus-8, Epstein Barr virus) have been postulated, it is now believed to be related to an autoimmune or a delayed hypersensitivity reaction. An aberrant allergic response is further supported by the association of the disease with asthma, allergic rhinitis, atopic dermatitis, and peripheral hypereosinophilia, as well as raised serum IgE levels [10]. Dendritic cells of the skin and mucosa process allergens deposited on the mucosa and subsequently dendritic cell present antigens to T cells. T helper 2 cells release their mediators upon recognition of antigens presented by antigen-presenting cells. The Th2 cytokines IL4, IL13, and CD40L induce selective somatic recombination of immunoglobulin heavy chain regions in B cells before maturation into IgE-producing plasma cells. IL5 stimulates eosinophil growth and differentiation. Alternatively, IgE is produced by stimulating innate immunity cells to release IL4, IL5, and IL13 [11]. Ultrasound, MSCT and Magnetic Resonance Imaging (MRI) might be diagnostic and can help staging the extent and progression of the disease as well as the lymph node involvement [10]. Differential diagnosis includes mainly angiolymphoid hyperplasia with eosinophilia (ALHE). Kimura disease and ALHE were often confused in a number of early reports until Rosai et al. Eventually clarified this misconception, and thus Kimura Disease and ALHE were established as two distinct entities [10]. The vascular proliferation is most significant in ALHE, forming aggregates or lobules comprised of plump endothelial cells [8]. The other differential diagnosis of KD would include Kikuchi disease, Mikulicz’s disease and most importantly Hodgkin and non-Hodgkin lymphoma [10].

CONCLUSION
Kimura Disease in this case diagnosed based on histopathological findings of the excised lesion. Hyperplastic follicles with abundant eosinophil and angiolymphoid proliferation with normal endotel are characteristic findings and on immunohistochemistry staining for CD3 and CD20 showed T-cells surrounding well-formed lymphoid follicles with germinal centers containing B-cells. Above case was diagnosed as Kimura Disease base on histopathology and immunohistochemistry staining which showed the characteristic features.

REFERENCES
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