

Kimura's Disease in a Female Patient: Case Report and Review of the Literature

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Submitted: 10 Sep 2018; **Accepted:** 17 Sep 2018; **Published:** 01 Oct 2018**Abstract**

Kimura's disease is a rare chronic inflammatory disease of uncertain cause, manifesting commonly as a painless swelling of the subcutaneous tissue in the head and neck region with predilection for peri auricular areas. A significant increase in serum levels of immunoglobulin E and eosinophilia in the peripheral blood and in tissues has been associated with this disorder. Many cases of nephropathy in patients with Kimura's Disease had been reported in the literature. Excisional biopsy for histopathological evaluation is the only way to confirm the diagnosis of Kimura's Disease. We report a case of Kimura's disease in a 26-year- old female patient who presented to our clinic with painless right retro-auricular swellings.

Keywords: Kimura's Disease; Nephropathy; Lymphadenopathy; Neck; Eosinophilia.

Introduction

Kimura's Disease (KD) is a benign, uncommon chronic inflammatory condition of uncertain cause. It has a predilection for males mostly in the third decade of their life [1]. Most commonly, KD presents as painless, unilateral Lymphadenopathy or subcutaneous masses in the head and neck region. The origin of KD was in China when Kimm and Szeto reported the first case in 1937 where it was known as "eosinophilia hyperplastic lymph granuloma" [2]. The current name of the disorder was received in 1948, when Kimura et al noted the vascular Component of the disease and referred to it as an "unusual granulation combined with hyperplastic changes in lymphoid tissue" [3].

The exact prevalence of Kimura disease is not known. Most cases of this rare entity are reported in East and Southeast Asia, with a small number of cases reported in Europe, with hardly 120 cases reported worldwide [4,5].

In most series reported, male-to-female ratio was 3.5:1 to 9:1, with the exception of one series in which the male-to-female ratio was 19:1, The mean age of affected individuals being 31-years- in one case series [6,7].

Kimura's Disease is generally limited to the skin, lymph nodes, and salivary glands. However, many cases of nephropathy in patients with Kimura's Disease had been reported in the literature [8].

Treatment of KD is not well established and still controversial. However, early diagnosis is critical so we can keep the patient away from unnecessary, and probably harmful, diagnostic procedures.

We report the case of a 26 -year- old female who complained of a

right retro-auricular swellings. The diagnosis of Kimura's Disease was established by histopathological evaluation of an excisional biopsy.

Case Report

A 26 -year- old female married patient, non-smoker, with body weight of 54 kg, was doing well till one year and a half ago when she presented to our outpatient clinic; in Al-Makassed Hospital, Jerusalem- with a chief complaint of a multiple right retro- auricular painless swellings, three in number, occasionally associated with pruritus, increasing gradually in the size, just very close to a single swelling appeared 6 months prior to these lesions which was excised under local anesthesia in a private clinic with no complications as the patient stated. However, no reports are available. She did not notice other swellings in her body as well.

There was no history of anorexia, unintentional weight loss, chronic cough, prolonged fever, or low back pain excluding, to some extent, potential malignancy and Tuberculosis (TB). There was no functional impairment of any kind. Surgical history of a nodule in the same area excised 6 months prior to presentation. Her medical and family histories were noncontributory. She was not taking drugs, and she had no drug allergies.

The physical examination revealed firm, non-tender, fixed three masses in the right retro-auricular area with a size ranging from 1-2 cm. They had a smooth surface with regular defined edges and no skin changes. The skin over the swelling could not be pinched. There was no hepatosplenomegaly and no other palpable regional lymph nodes. Swellings of similar kind could not be found elsewhere.

Pertinent laboratory values included Complete Blood Count (CBC), Urinalysis, and Kidney Function Tests (Creatinine and Blood Urea Nitrogen) and were within normal ranges. Abdominal CT scan showing the liver was done and excluded evidence of underlying

malignancy.

The three nodules were totally excised under local anesthesia and set for histopathological examination which showed “lymph nodes with preserved architectures, but with follicular hyperplasia, prominent eosinophilia infiltration, and proliferation of post capillary venules suggestive of Kimura’s disease.”

The plan of management was a course of prednisolone (20mg once daily) and azathioprine (50mg twice daily) for six months. The patient responded well to the previous management and follow up through twenty-one months after that was satisfactory with no recurrence.

Discussion

Kimura’s Disease (KD) is a benign chronic inflammatory disease mainly affecting males in the third decade of life with the mean age of onset being 31-years- in one case series [1,7]. The exact etiology of this rare entity remains uncertain although some authors suggested possible risk factors that may play a role in development of this benign condition, including: allergic reactions, infections, and autoimmune reactions [9-12].

Although KD is not an uncommon condition in East and Southeast Asia countries with most cases being reported in these areas; it is quite rare in the western countries [4].

It usually presents as a painless subcutaneous single or multiple nodules in the head and neck area with a predilection for the peri-auricular region. Although the lumps grow slowly, the patient remains otherwise asymptomatic. Pruritus may occur occasionally as we noticed in our patient. Many cases of nephropathy in patients with Kimura’s Disease had been reported in the literature [8]. Different groups of renal diseases can present in patients with KD are as membranous glomerulonephritis, minimal change glomerulonephritis, diffuse proliferative glomerulonephritis, meningeal proliferative glomerulonephritis, and nephritic syndrome [13-15]. However, other Complications like Raynaud’s phenomenon and ulcerative colitis have been reported [16,17]. Laboratory findings consistent with KD include peripheral eosinophilia and elevated serum IgE levels, and the definite diagnosis is only made by histopathological evaluation from excisional biopsy.

The most prominent histological features that can differentiate KD from other similar entities presented in the head and neck region are preserved architecture of the lymph nodes, follicular hyperplasia (Figure 1), prominent eosinophilia infiltration (Figure 2), and proliferation of post capillary venules.

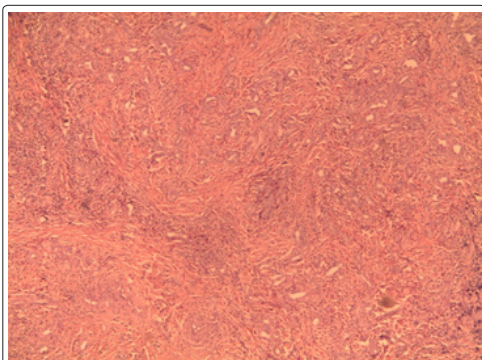


Figure 1: Micrograph of preserved architecture of lymph nodes

with follicular hyperplasia.

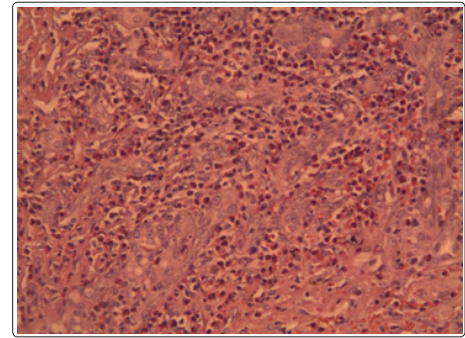


Figure 2: High-power photomicrograph demonstrating increased eosinophils in the paracortex.

KD should be strictly differentiated from a very similar rare condition; Angiolymphoid hyperplasia with eosinophilia (ALHE). They were thought, at certain point of time, to be the same disease [18].

The two conditions possess close similarities making them difficult to be distinguished except by Excisional biopsy, these features include; preference of the head and neck region to grow in as a painless subcutaneous nodule, carry a percentage of recurrence despite adequate treatment, and the presence of lymphoid infiltration with eosinophil’s and vascular proliferation [7,18].

However, ALHE usually affects women in the 3rd-4th decades of life with no racial propensity and presents as multiple dermal nodular eruptions which occur with a long duration. Peripheral blood eosinophilia and increased serum IgE levels, which are prominent features in KD, are rare in ALHE [18,19].

The only way by which KD and ALHE can be distinguished is through a histopathological evaluation which reveals blood vessels with a “histiocytoid” and “epithelioid” characterizing ALHE. This feature, however, is absent in KD [10].

Other differential diagnoses to be considered in case of enlarged lymph node comprise: reactive lymphadenopathy, nodal metastasis, and lymphoma. However, reactive lymphadenopathy was excluded due to the prominent eosinophilia and vascularization shown on the histological picture, nodal metastasis was ruled out due to absence of tumor cells, and lymphoma as the lymph node biopsy was not suggestive of lymphoma in this case and the patient did not have any symptom consistent with malignancy [13].

Management of KD is still controversial. However, aim of the treatment is directed toward reducing the risk of recurrence, long term complications, and preserving the function and cosmetics. Conservative observation may be adequate for asymptomatic patients. Therapeutic modalities like: surgical excision, radiotherapy, or steroid therapy can be used for symptomatic cases [11]. Steroid therapies are effective in mitigation of the lesions of KD but a recurrence is possible when the medication is stopped [20]. However, for lesions returning after treatment by either surgery or steroid therapeutic modalities, local irradiation can be considered with a satisfactory outcome [21].

In our present study, the female patient developed multiple classical

features of KD, including painless, subcutaneous masses, pruritus, with eosinophilia and elevated serum IgE. Blood sample was taken and revealed no abnormalities in the hepatic and the renal functions.

A regimen of Prednisone (20mg once daily) and Azathioprine (50mg twice daily) were given for six months. The patient responded to the previous combination very well with no recurrence till his moment of submitting the case. To our knowledge, no one has reported before remission For Kimura's lesions for such a very long period of time (21 months).

In summary, KD is a chronic inflammatory disease seen commonly in young Asian males and presents as a single or multiple subcutaneous nodules. Diagnosis is only possible by evidence of high levels of IgE and peripheral blood eosinophilia and histopathological evaluation. Treatment aims to reduce the risk of recurrence and to preserve the function. Kimura's disease has no potential for malignancy.

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