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Case Report

Surgery Section

Kimura's Disease: A Diagnostic and Therapeutic Dilemma

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ABSTRACT

Kimura's disease (KD) is a rare, benign disorder. It is a chronic inflammatory disease of unknown aetiology. It is endemic in Asia (China and Japan) and rare in the west. It has the highest predilection for the head and neck region. The histology shows a hyperplastic lymphoid tissue and an inflammatory infiltrate. The treatment options vary from surgical excision to corticosteroids to radiation therapy. The prognosis is very good and no malignant transformation has been reported so far.

Key Words: Acute Myocardial Infarction, Young adults, Risk factors, Smoking

INTRODUCTION

Kimura's disease was described in 1937 in the Chinese literature by HT Kimm and C Szeto. The definitive histological description was published by Kimura et al, in 1948 and henceforth, the disease has borne his name [1]. It is also known as eosinophilic lymphogranuloma and is a rare benign inflammatory disorder of unknown aetiology. It is endemic in Asia, especially in China and Japan, but is rarely seen in the west. It is typically seen in young males, especially in the second decade of their lives. It commonly presents as solitary or multiple subcutaneous nodules of variable sizes which grow upto 10 cm in size. The most common site of presentation is the head and neck region. It occurs most commonly in males, the male to female ratio being 3.5 to 7.1 [1].

Peripheral eosinophilia, regional lymphadenopathy and markedly elevated serum IgE levels are characteristic of Kimura's disease. Salivary gland involvement is typical of this disease. Renal involvement is the only systemic manifestation. The histology shows hyperplastic blood vessels which are lined by endothelial cells and numerous lymphoid follicles with a prominent germinal centre. The treatment of choice is still controversial, as the recurrence rate is high, irrespective of the type of treatment which is given.

CASE REPORT

Our patient was a 14-year old boy who had a swelling in the right parotid region, 5 years prior to his presentation at our hospital, i.e. at the age of 9 years, for which he underwent surgical excision 3 years back at another hospital. However, as per the history, the swelling promptly recurred immediately within a month, which gradually increased in size and 2 years later, he presented to our OPD for consultation. On examination, a large diffuse swelling was found to be present in the right parotid region, which had a nodular surface. A healed surgical scar was observed in its upper posterior aspect. The swelling was mobile and soft in consistency. All its borders could be well made out. It was partially compressible, but was not fluctuant. There was no evidence of facial nerve palsy. In view of the above findings, a clinical diagnosis of vascular malformation/ lymphangioma was made. On investigating his blood parameters revealed eosinophilia and raised ESR. A soft tissue scan revealed that the right parotid gland was compressed by multiple lymph nodes. Multiple lymph nodes were also noted in the submandibular and the cervical regions. The largest lymph node measured 2.2x 1.3 cms. There was subcutaneous oedema. The features were suggestive of Kimura's disease. FNAC revealed florid reactive lymphoid hyperplasia.

Since the above investigations did not help us in making a conclusive diagnosis, the histopathology slides of the previous excision were retrieved and sent for review to our pathologist. The review of the histopathology slides revealed hyperkeratotic epidermis. The dermis showed numerous lymphoid follicles with prominent germinal centres. Numerous eosinophils which were admixed with plasma cells were present in the intervening tissue. These findings clinched the diagnosis of Kimura's disease.

Once the diagnosis of recurrent Kimura's disease was made, the patient was started on corticosteroids and cetrizine. It was decided to treat the patient conservatively following surgical excision, as it was a recurrent swelling. The patient was followed up after a month of steroids and antihistamines and a 90% reduction was observed in the size of the swelling. The patient was continued on steroids and antihistamines and he requires further long term follow-up.

DISCUSSION

Kimura's disease, though described by Kim et al, was popularized by Kimura in 1947 [2]. It is endemic in Asia (especially in China and Japan) but is rare in the rest of the world. It is a benign inflammatory disorder with a predilection for the head and neck region. It involves the subcutaneous tissue, lymph nodes and the salivary glands. It presents as solitary or multiple sub-cutaneous nodules in the head and neck region. Epitrochlear, axillary, inguinal and popliteal localizations have been described. The eyes, ears, spermatic ducts and the nerves are rarely affected. Renal involvement, usually extramembranous glomerulonephritis, may be found.

It is a benign disease and there have been no reports of a malignant transformation so far. Though its aetiology is unknown, theories about its origin have been proposed, such as interference with the immune regulation and an atopic reaction to continu-

ous antigenic stimulation (candida albicans/ parasitic infestation/ neoplasm).

It commonly occurs in the second decade of life. Our patient presented at the age of 9 years, which is the youngest age which has been reported so for. The differential diagnosis of Kimura's disease include [3,4] reactive lymphadenopathy angioma/lymphangioma/ hemangioma, lymphoma, salivary gland tumours, nodal metastasis, (breast, colorectal, nasopharynx), Mickulikz's disease, etc. The most frequently highlighted differential diagnosis which has been observed in the literature is angiolymphoid hyperplasia with eosinophilla (ALHE) [5].

The diagnosis can be aided by USG/CT scan. These can also help in delineating the extent and in evaluating the progression of the disease. FNAC, although inconclusive and performed for the diagnosis of KD, it's role is largely in the diagnosis of the recurrent lesions and it may obviate the need for an open biopsy [6]. The confirmation of the diagnosis can only be done by histopathology, where the dermis and/or the subcutis shows hyperplasia of the small blood vessels which are lined by endothelial cells. These hyperplastic vessels are surrounded by a dense infiltrate of lymphocytes, plasma cells and histiocytes.

An optimal treatment for KD has not been well established. However, such a treatment should be aimed to preserve the cosmesis and function, while preventing recurrence and long term sequelae. The range of treatment options include conservative treatment, steroid therapy, radiotherapy, cryotherapy, laser fulguration and surgical excision. The value of achieving negative surgical margins after excision has not been studied.

The first line treatment options include surgical excision (recurrence of up to 25%) and topical steroids (these have a higher potency when they are used twice daily). Our patient had undergone surgical excision initially, but the disease recurred immediately. Hence, we put him on corticosteroids and antihistamines, to which he responded very well.

The second line options are topical tacrolimus twice daily, systemic steroid therapy (the recurrences are high after stopping the steroids), cyclosporine, antihistamines and local radiation therapy. Radiation therapy may be preferred for those lesions which are not amenable for surgery due to their large size or for those lesions which if operated upon, may result in unacceptable morbidity

CONCLUSION

In conclusion, it can be said that Kimura's disease is rare, benign, inflammatory disorder. Though it is rare, one has to be aware of it, as its recurrence rate is very high following any kind of treatment . A high index of suspicion is required to diagnose the disease. It should be kept in mind as a possible differential diagnosis for masses in the head and neck region, especially in the Asian population where it is endemic. The management of Kimura's disease is still controversial, with a high recurrence rate following either surgical excision or the medical line of treatment. In the paediatric age group, the medical line of treatment should be tried first due to the high recurrence rate in spite of surgical excision of the masses.

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