

Case Report

Kimura's disease: a diagnostic and therapeutic enigma

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ABSTRACT

Kimura's disease is a rare chronic inflammatory disorder present in 2nd and 3rd decade. It has a predilection for head and neck region presenting as a slowly growing painless swelling. It is usually accompanied by peripheral eosinophilia and elevated serum IgE and hence it was initially thought to be of allergic origin. Histologically the lesions are characterized by reactive hyperplasia of lymph nodes, eosinophilic infiltration and increase in postcapillary venules. Authors have reported a male patient with a slowly growing right sided neck swelling which is recurring even after course of steroids and excision done twice at an interval of 6 months. Kimura's disease although a benign Lymphoid disorder but the incidence of recurrence despite taking treatment is a cause of much concern for the patient.

Keywords: Clinical dilemma, Histopathology, Kimura's disease, Lymphadenopathy

INTRODUCTION

Kimura's disease is a diagnostic and therapeutic challenge. It is a rare benign clinical disorder of unknown etiology characterized by painless subcutaneous swellings commonly affecting head and neck region. Axilla, popliteal region, orbit, groin and forearm are other less commonly involved sites.¹

Although it may mimic a neoplastic process but peripheral blood eosinophilia, increased serum IgE concentration and histopathological findings suggest an immunological etiology to an unknown stimulus. Other postulated theories are allergic reaction, stimulation following arthropod bites, parasitic or candidal infection. Asian population is more often affected with a male preponderance in a ratio of 3:1 in 2nd and 3rd decade.^{1,2} Even though benign in nature its incidence of recurrence and devastating complications sometimes leading to disfigurement is a cause of much concern for the patient.

Long term steroids, radiotherapy, cryotherapy, cytotoxic therapy and excision have been tried with different success rates.³

We describe a case of Kimura disease presenting with a recurring slowly growing neck swelling with waxing and waning course.

CASE REPORT

A 24-year-old male presented to medicine department with multiple painless swellings (Lymph nodes) on right side of neck for the last 15 days. The swellings were gradually increasing in size. There was no history of loss of appetite, weight loss, fever, cough or any other upper respiratory tract infection symptoms. He had no other complaints related to the kidneys, liver or heart.

Routine investigation revealed Hemoglobin 13.5gms/dl, normal for age and sex, TLC was raised

15,000cells/cumm, Differential Count was Polymorphs=67%, Lymphocytes = 18%, Eosinophils = 15%, Monocytes =00%; Absolute Eosinophil Count = 2250 cells/cumm; Erythrocyte Sedimentation Rate (Westgren Method) was 22mm /1st hour, Mantoux test was negative. Urinalysis, Serum electrolytes, BUN, S. Creatinine, Blood Glucose, S. Calcium, S. Protein, Liver Enzyme levels were within normal limits. HIV, HbsAg, HCV were non-reactive.

IgE level was raised 20,200µg/litre (Normal less than 430µg/litre). X-ray Chest and whole abdomen Ultrasonography was normal. A provisional diagnosis of Lymphoma was made.

FNAC was done, it revealed a rich polymorphous population of lymphoid cells with numerous eosinophils (Figure 1). Reed Sternberg cells or any other evidence of Lymphoma was not observed. A diagnosis of eosinophilic lymphadenitis with possibility of Kimura disease/ Angiolymphoid hyperplasia was suggested. A biopsy was recommended.

Gross examination revealed single globular tissue measuring 3.4x2.8x1cm. Outer surface was nodular with few dilated blood vessels. Capsule was identified. Cut Surface was greyish white with brownish and hemorrhagic areas.

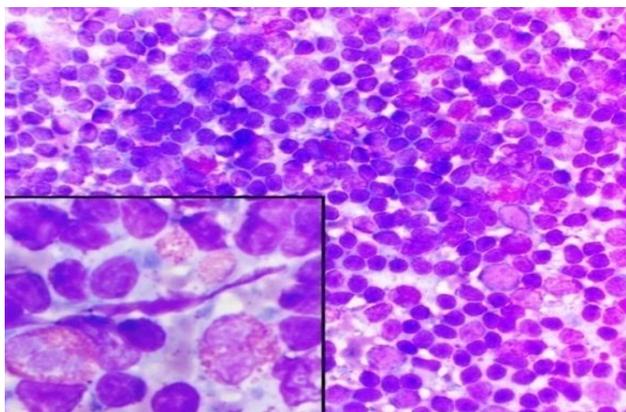


Figure 1: FNAC smear showing eosinophilia Leishman -Giemsa, X 40 (Inset X100).

Microscopic Examination revealed structure of Lymph node which was partially effaced. Large lymphoid follicles were present with prominent germinal centres (Figure 2). Interfollicular area showed dense collagenized tissue with numerous eosinophils, plasma cells, mast cells and occasional dendritic cells.

At places eosinophilic abscesses (Figure 3) were also noted. Hyalinated blood vessels were seen in paracortical area alongwith proliferation of venules (Figure 3). Van Gieson stain showed collagenized thick and thin fibres mostly in paracortical area (Figure 4).

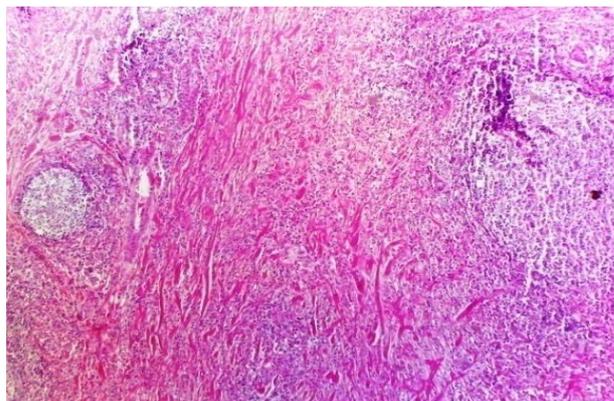


Figure 2: Large lymphoid follicles with prominent Germinal centre (Hematoxylin and Eosin, X 40).

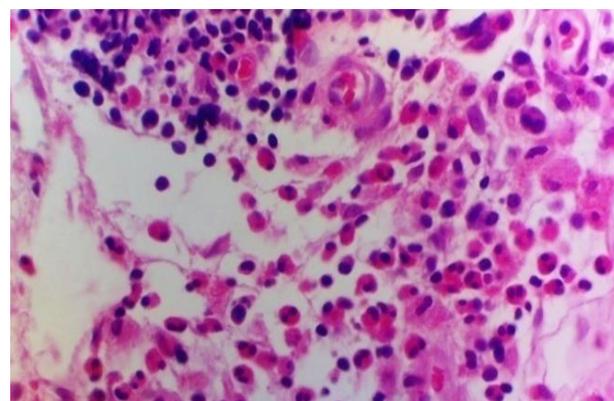


Figure 3: Eosinophilic abscess and postcapillary venules (H and E, X 40)

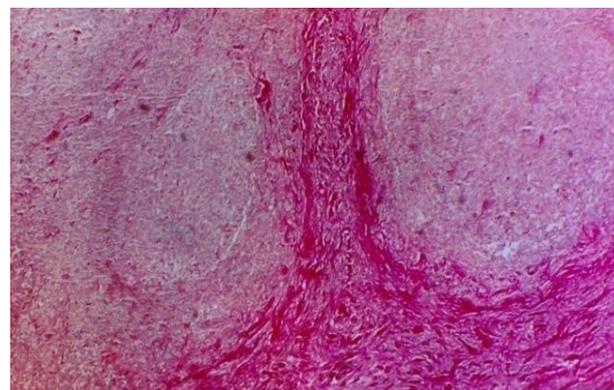


Figure 4: Thick collagen bundles in paracortical area (Van Geison x40).

Treatment with Prednisilone was started for a couple of months but since the swelling did not regress to any significant degree he took a third opinion from a different Medical institute. All his routine investigations along with systemic examination was done. Apart from peripheral eosinophilia there were no other significant findings. A repeat FNAC followed by excisional biopsy was again performed. There also a diagnosis of Kimura's disease was made, and he was put on prednisolone

1mg/Kg body weight along with cyclosporine 50mg/day. He took treatment for about 6 months after which although his swellings slightly decreased in size but there was pigmentation all over the body and steroid induced cataract related changes also developed. He stopped the treatment for 2 months but again the swellings started increasing in size with persistence of pigmentation. (Figure 3). He again came to our institute for further treatment. At present he is on prednisolone 1mg/day and leflunomide 20mg/day for 3 months. However, still the swelling has not completely regressed.

DISCUSSION

Kimura disease was first described in China, but it was not mentioned until Kimura et al, mentioned it in Japanese language literature in 1948.¹ It is a rare benign chronic inflammatory disease, presenting as a slowly growing soft tissue swelling associated with lymphoid and peripheral blood eosinophilia. Lymph nodes of head and neck are most commonly involved mimicking neoplastic lymphoproliferative disorder.⁴ Other sites of involvement are parotid glands, epitrochlear, axillary and inguinal lymph nodes.⁵ Rare sites of KD are orbits, spermatic cord, kidneys and nerves.⁶ KD usually occurs in a young Asian male between 2nd and 3rd decade. Lymph node enlargement is slow over a period of months to years.^{2,3}

The present case describes an Asian male aged 24 year presenting with a slowly growing painless enlarged cervical swellings causing disfigurement. Similar swelling has been reported by Claudia et al.³

Aetiopathogenesis of KD is unknown. Various theories have been postulated like Allergic reactions, infections, autoimmune reaction. Devastation complications like Kidney involvement, asthma, atopic dermatitis, allergic rhinitis and urticaria occur in patients.⁷

Since this entity is relatively rare especially in the western countries it poses problems in the diagnosis. On peripheral blood examination, the eosinophilia and elevated IgE levels prompts the clinicians to consider an allergic condition. FNAC aspirates with large number of eosinophils and polykaryotic giant cells may lead one to a differential diagnosis of Hodgkin lymphoma but absence of Reed -Sternberg cells rules out Hodgkin lymphoma.³ KD of long duration has been found to have associated organ involvement also, specially the kidney.⁶ The various renal lesions encountered are minimal change disease, mesangio-proliferative glomerulonephritis, focal segmental glomerulosclerosis, membranous and IgA nephropathy.⁷ However since the history of our patient is of comparatively short duration he had no such complications.⁸

The constant histologic features that are seen in this disease are lymphoid infiltrate with formation of hyperplastic follicles and florid germinal centres, well

defined mantle zone, dense eosinophilic infiltrate, even forming eosinophilic abscesses and increased postcapillary venules. Polykaryocytes (Warthin-Finkeldey) can be seen.³

Some authors propose that KD and ALHE are different stages of the same disease process.⁹ Although histologically both have a proliferative vascular nature with eosinophilic infiltrate but peripheral blood does not reveal eosinophilia in ALHE cases.¹⁰ Other distinguishing features are that ALHE is seen more often in Caucasian females involving the superficial skin forming papules, hypertrophic endothelial cells protruding into the vascular lumina occlude the lumen of the vessels.¹¹ Lymphadenopathy is not a frequent finding in ALHE.¹²

The differential diagnosis of Kimura disease includes Hodgkins Lymphoma characterized RS cells but deposits of IgE and hyperplastic germinal centres are not found. Castleman disease lacks eosinophilia and has involuted hyalinized germinal centres.¹²

In dermatopathic lymphadenopathy deposits of hemosiderin, melanin and lipids will be seen along-with follicular hyperplasia and sclerosis. Drug reactions might give a similar picture, but history of drug intake will exclude the diagnosis. There are other disorders too like epitheloid hemangioma, low grade angiosarcoma, pseudopyogenic granuloma and eosinophilic granuloma of the soft tissues which are thought to be variants of Kimura.¹²

In this case the FNAC and biopsy findings of prominent germinal centre of the involved lymph nodes with dense eosinophilic infiltrate in a background of lymphocytes and plasma cells and increased postcapillary venules in the paracortex are characteristic findings of KD.¹²

Treatment of KD is controversial. Patient underwent excision biopsy of cervical swelling and was started on oral steroid therapy. He took this treatment for 6 months after which he developed pigmentation of skin all over the body along with cataract, although the swellings subsided for some time and recurred again at same site after one year with persistence of Pigmentation. This shows that although KD is a benign condition but despite taking treatment it is not completely curable. The patients probably get refractory to steroid therapy.¹⁰ In younger patients radiotherapy has been tried to prevent relapse and reduce long term side effects of steroid therapy.¹⁰

CONCLUSION

Kimura's disease although a benign Lymphoid disorder but its waxing and waning clinical course with the incidence of recurrence despite taking treatment and getting excision done is a cause of much concern for the patient. For the treating clinician to it is exasperating to pacify the patient. There seems to be no absolute cure for this disease.

In this case although the swellings as such was not painful nor was it in any way pressing on the adjacent organs but was causing disfigurement and cosmetic trauma to the patient.

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Ethical approval: Not required

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