International Surgery Journal
Kuruvilla S et al. Int Surg J. 2016 May;3(2):947-949
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20161173

Case Report

Recurrent Kimura's disease successfully treated with steroids and cetirizine

Shalini Kuruvilla[1]*, Rojan Kuruvilla[2], Remesh Bhasi[3], Lilly M[1]

[1]Department of Pathology, [2]Department of Surgery, [3]Department of Rheumatology, Malabar Institute of Medical sciences, Kozhikode, Kerala, India

Received: 01 March 2016
Accepted: 12 April 2016

***Correspondence:**
Dr. Shalini Kuruvilla,
E-mail: shaliniku@gmail.com

ABSTRACT

Kimura's disease is a rare cause of a neck mass. A case of Kimura's disease presenting as a recurrent neck mass following previous excision is presented. Preoperative diagnosis may be possible if the eosinophilia is noted and a serum IGE is estimated. Surgery is the primary modality of treatment. Diagnosis is confirmed by histology. Steroids and Cetirizine are effective in the management of recurrences.

Keywords: Kimura's disease, Steroids, Cetirizine, Eosinophilia, IgE

INTRODUCTION

Kimura's disease is a rare chronic inflammatory disorder of unknown etiology that primarily affects young Asian males. The typical presentation is characterized by a triad of;

- Painless unilateral cervical adenopathy or subcutaneous masses predominantly in head or neck region
- Blood and tissue eosinophilia
- Elevated serum IgE levels

Kimura's disease has also been reported in the West, in non-Asians and in unusual sites such as axilla, groin, palate and epitrochlear region.[1,2] Though a confident diagnosis of Kimura's disease can be made by the pathologist, it is often misdiagnosed as angiolymphoid hyperplasia with eosinophilia or labeled descriptively as reactive lymphoid hyperplasia with eosinophilia. The primary treatment is surgical but recurrences can be effectively managed medically.

CASE REPORT

A 22 year old Indian male was referred to our surgical department with a recurrent right parotid swelling of three years. It was excised twice elsewhere and the histopathology was reported as lymphoepithelial lesion. The slides were not available for review. On physical examination a firm mass measuring 3x4 cm was palpable in the right parotid region lifting up the pinna. The rest of the physical examination was normal. Laboratory investigations showed a TC of 11290/ mm^3 and an eosinophilia of 29%.

A right superficial parotidectomy was done preserving the facial nerve. There was dense fibrosis surrounding the gland per operatively

At histopathology salivary gland tissue was identified at the periphery of the specimen. There was fibrosis and a dense inflammatory infiltrate in per parotid tissue. Intra parotid lymph nodes showed follicular hyperplasia (Figure 1) with eosinophilic micro abscess (Figure 2) formation within the cortex and in the inter follicular

regions. In many areas the eosinophilic infiltrate was subtler. Proliferation of high endothelial venules was seen. The salivary gland tissue was normal. Based on the histopathology clinical presentation and marked peripheral eosinophilia a diagnosis of Kimura's disease was made. A serum IgE was done which was markedly elevated (2500 IU/ml). Renal function and urine examination were normal.

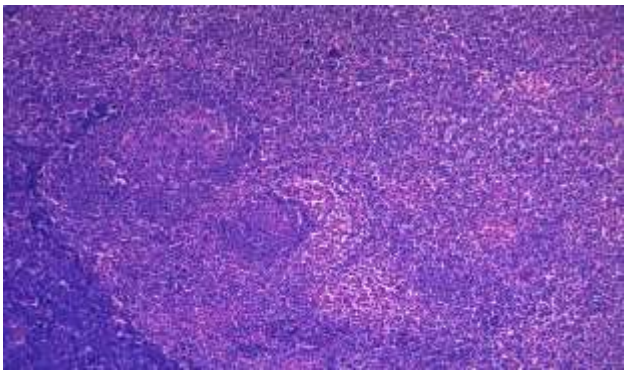

Figure 1: Lymph node showing follicular hyperplasia with eosinophilia (H&E stain x100).

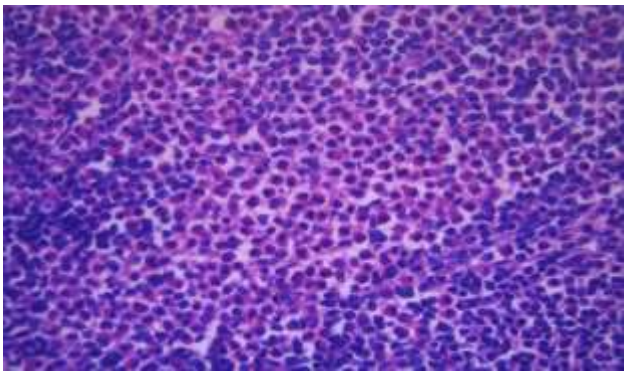

Figure 2: Inter follicular area showing eosinophilic micro abscesses (H&E stain x 400).

He remained disease free for two years and then presented with recurrence again in the same region. As surgery was not feasible the rheumatologist's opinion was sought and he was started on Tablet Methylprednisolone 16 mg TID with tapering doses over 3 months and Tablet Cetirizine 10 mg at night. Within two weeks the cervical mass totally disappeared. At four months follow up he was fine. However, he has been advised to continue Cetirizine to prevent recurrence.

DISCUSSION

There has been confusion between Kimura's disease (KD) and Angiolymphoid hyperplasia (ALHE) in the past years. They are two separate identities with distinctive clinical and histologic features though both present as soft tissue swellings in the head and neck region.

Kimura's disease occurs predominantly in young Asian males with eosinophilia and elevated serum IgE levels. ALHE on the other hand typically affects middle aged women without racial predilection. Eosinophilia and lymphadenopathy is rarely noted.[3]

Hui PK et al has outlined the histological features seen in KD. The constant features include preserved nodal architecture, florid germinal center hyperplasia, eosinophilic infiltration and postcapillary venule proliferation.[2]

By contrast ALHE is a circumscribed vascular neoplasm mostly confined to the subcutaneous plane. The vascular proliferation is florid and vaguely lobular with plump endothelial cells. An inflammatory infiltrate with a few eosinophils is present around medium sized arteries or veins. However, the presence of reactive germinal centers and eosinophilic microabscess formation or reactive lymphadenopathy is uncommon features as compared to Komura's disease.[1,2]

Proliferated vessels in the Kimura's disease lack the plump endothelial cells of ALHE

The differential diagnosis includes Hodgkin's lymphoma, Angioimmunoblastic T cell lymphoma, Langerhans's cell histiocytosis, Lymphadenopathy of drug reactions and parasitic lymphadenitis. The separation from neoplasm is not difficult when classic Reed Sternberg cells, atypical lymphocytes or Langerhans cells are identified. Positive history of drug use and detection of parasitic remnants can lead to the correct diagnosis in the latter two cases.[1,2]

Our case was initially diagnosed as lymphoepithelial lesion on two occasions. The florid lymphoid follicular hyperplasia probably led to this diagnosis. However, the presence of significant eosinophilic infiltrates should make one suspect Kimura's disease especially for masses in the cervical region. Raised serum IgE and a high peripheral eosinophil count would confirm the diagnosis

The clinical course of KD is benign though recurrences occur in 25%. The lesions of KD may precede or coincide with the development of renal complications. Nephrotic syndrome is the most common presentation.[4]

The etiology and pathogenesis are yet unknown. It is believed to be an allergic or autoimmune response to an unknown stimulus which results in the release of cytokines. Abundant expressions of eosinophil tropic cytokines such as IL4, IL5, and IL13 in peripheral blood mononuclear cells has been reported suggesting that these cytokines have a role to play in its pathogenesis.[5]

The primary treatment is surgical excision. Literature on optimal treatment of recurrent KD is limited to a few case reports. Corticosteroids and immunosuppressive agents have been tried out successfully. The dramatic response to cetirizine in our case may indicate that inhibition of

eosinophil activity is one of the pillars of treatment. Long term steroids have many side effects and Cetirizine can effectively replace it. Some authors support the use of Cyclosporine for maintaining remission. There are also case reports on successful treatment of KD with Leflunomide.[6-8]

Radiation maybe considered for recalcitrant and large tumours and in cases refractory to surgical and medical therapy.[9]

CONCLUSION

Kimura's disease is uncommon and should be suspected in cases of neck masses with eosinophilia. The importance of diagnosing KD is that patients need follow up both for renal involvement and recurrences. Recurrences can be managed medically without surgical excision.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

REFERENCES

1. Chen H, Thompson LDR, Nadine. Kimura disease: a clinicopathologic Study of 21 Cases. Am J Surg Pathol. 2004;28(4):505-13.
2. Hui PK, Chan JK, Ng CS, Kung IT, Gwi E. Lymphadenopathy of Kimura's disease. Am J Surg Pathol. 1989;13:177-86.
3. Chun SI, Ji HG. Kimura's disease and angiolymphoid hyperplasia with eosinophilia: Clinical and histopathologic differences. J Am Acad Dermatol. 1992;27:954-8.
4. Atar S, Oberman AS, Izak OB, Flatau E. Recurrent nephrotic syndrome associated with Kimura's disease in a young non-Oriental male. Nephron. 1994;68:259-61.
5. Katagiri K, Itami S, Hatono Y, Yamaguchi T, Takayasu S. In vivo expression of IL-4, IL-5, IL-13 and IFN-gamma mRNAs in peripheral blood mononuclear cells and effects of cyclosporin A in a patient with Kimura's disease. Br J Dermatol. 1997;137:972-7.
6. Chetrit EB, Amir G, Shalit M. Cetirizine: an effective agent in Kimura's disease. Arthritis Rheum. 2005;53(1):117-8.
7. Dai L, Wei XN, Zheng DH, Mo YQ, Pessler F, Zhang BY. Effective treatment of Kimura's disease with leflunomide in combination with glucocorticoids. Clin Rheumatol. 2011;30:859-65.
8. Shenoy VV, Joshi SR, Kotwal VS, Shedge RT, Ramraje NN, Lanjewar DN. Recurrent Kimura's disease: excellent response to cyclosporine. J Assoc Physicians India. 2006;54:153-5.
9. Kim GE, Kim WC, Yang WI, Kim SK, Oh WY, Suh HS, et al. Radiation treatment in patients with recurrent Kimura's disease. Int J Radiat Oncol Biol Phys. 1997;38:607-12.

Cite this article as: Kuruvilla S, Kuruvilla R, Bhasi R, Lilly M. Recurrent Kimura's disease successfully treated with steroids and cetirizine. Int Surg J 2016;3:947-9.