Case Report

Recurrent Kimura’s disease successfully treated with steroids and cetirizine

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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.¹² 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 112⁹/ mm³ 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.

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. 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. 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.

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. 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. 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 eosinophilic 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. 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

**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.
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.

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REFERENCES