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KIMURA'S DISEASE- A CASE REPORT

ENT

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ABSTRACT

Kimura's disease is a benign, slow-growing chronic inflammatory disorder. The etiology of kimura's disease is still unknown. It typically occurs at 20 to 30 years of age predominantly in Asian race. It mainly affects head and neck region with cervical lymphadenopathy. Kimura's is a triad of painless mass, eosinphilia, and raised IgE levels. Surgical excision is the treatment of choice. The surgery has minimal morbidity and good success rate. There has been no documented case of malignant transformation however recurrence rates after excision may be as high as 25%. We present a case of a 53 year old male patient with bilateral postauricular swelling since childhood.

KEYWORDS

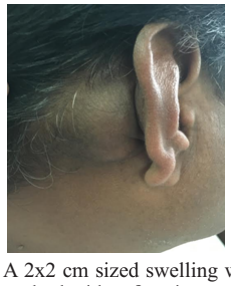
Lymphadenopathy, Eosinophilia

INTRODUCTION

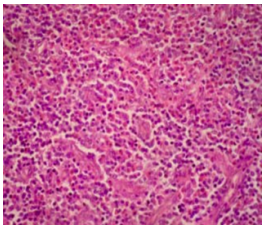
- Kimura's most commonly presents as painless unilateral lymphadenopathy or subdermal mass in the head and neck region.
- It is usually seen in young male adults during 3rd decade of life with median age being 28-32 years.
- It primarily affects the Asian descent and involves lymph nodes and salivary glands but patients of kimura's disease with nephrotic syndrome have also been reported.
- It is associated with raised serum IgE levels and eosinophilia.

A CASE REPORT

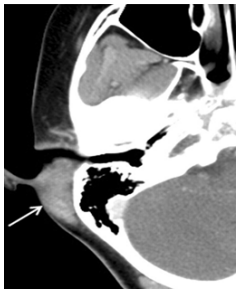
- A 53 year old hindu male patient presented to ENT department with chief complaint of Bilateral post auricular swelling since childhood, gradual in onset and painless. No complain of ear discharge, earache, decreased hearing or any other otologic symptoms. No history of fever, weight loss, anorexia, cough with expectoration, night sweats. Patient had no significant past history.



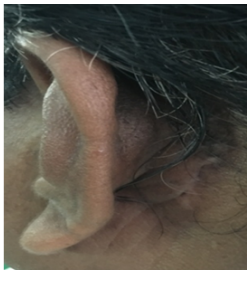
- On examination A 2x2 cm sized swelling was present over post auricular region on both sides, firm in consistency, Smooth and well defined margins, Non warm, Non tender, Non fluctuant, No other swellings were palpable. Ear, nose and throat examination was normal.
- CBC showed raised eosinophils (›10 in differential count) other parameters were normal. Serum IgE levels were highly elevated to 614 IU/ml[normal range is 11-162 IU/ml.] RFT was normal. Urinary protein levels were normal which excluded renal dysfunction. Chest X-ray was normal. USG local part - 30x10 mm enlarged node in left and 24x9mm enlarged node in right post auricular region.



- FNAC was performed which showed small mature lymphocytes, eosinophils, few histiocytes, plasma cells and neutrophils suggestive of 'CHRONIC NON SPECIFIC LYMPHADENITIS WITH PROMINENT EOSINOPHIL COMPONENT'.
- On CT scan of temporal bone 35x19x18 mm on right side and 28x19x24 mm on left side soft tissue density lesions were noted in post auricular region.



- No other abnormality seen.
- For treatment Bilateral Surgical excision was carried out and the biopsy was sent for histopathological examination. Section showed follicular hyperplasia with extensive infiltration by eosinophils and plasma cells with eosinophilic lysis which suggested diagnosis of Kimura's disease.



Discussion

- The disease presents as a swelling. It is usually unilateral soft tissue mass in head and neck. Frequently it involves lymph nodes and major salivary glands. Bilateral involvement is usually rare. Renal involvement is its only systemic manifestation found in 10% to 60%, while 10% to 12 % may suffer from nephrotic syndrome chracterised by clinically relevant proteinuria in 12% to 16% of patients. It Includes membranous glomerulopathy, mesangioprolif erative glomerulonephritis, minimal change disease and focal segmental glomerulosclerosis. Lymphadenopathy is associated with marked peripheral eosinophilia, and an elevated IgE level.

- The differential diagnosis includes inflammatory and neoplastic conditions Tuberculosis, Other infectious lymph node enlargements like, toxoplasmosis, Kaposi's sarcoma, Pyogenic tumours, lymphoma, Parotid tumours, Neurofibromatosis, Malignant tumours, Metastatic tumours.
- In rare cases, it has been reported to involve other sites, such as Oral cavity, Conjunctiva, Eyelid, Tympanic membrane, Skeletal muscle, Prostate, kidney, Peripheral nerves Epiglottis etc. The pathophysiology remains unknown.
- It has been hypothesized that an infection or toxin may trigger an autoimmune phenomenon or leads to a type I (immunoglobulin E-mediated) hypersensitivity reaction. Predominance of TH2 cells which produce eosinophilic cytokines, including interleukin (IL-4 and IL-5). Elevated granulocyte macrophage-stimulating factor, tumour necrosis factor-a, soluble IL-2 receptor, IL-5, IL-4 and IL-13 can also contribute. Kimura's disease can also be confused with angiolymphoid hyperplasia with eosinophilia [ALHE].
- Radiotherapy, intralesional steroids followed by oral steroids for longer duration has been tried but surgery remains the mainstay of therapy.

Conclusion

- Kimura's disease, although difficult to diagnosis clinically, should be considered in the differential diagnosis of patients who have a primary lymphadenopathy with eosinophilia with or without subcutaneous nodules. It should be investigated accordingly as the disease has an indolent course and good prognosis. Radiotherapy, intralesional steroids followed by oral steroids for longer duration has been tried but surgery remains the mainstay of therapy.

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