Kimura disease a case report

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Abstract:
Kimura disease (KD) is a rare, benign, chronic inflammatory disease with unknown etiology. Its manifestation is protean. Kimura disease has a predilection for the head and neck area, and typically presents as tumor-like lesions that could be easily misdiagnosed. We review our experience with our case.

Keyword: Kimura disease, lymph adenopathy, Immunoglobulin E, eosinophilia.

INTRODUCTION Kimura disease is a chronic inflammatory disorder prevalent in Asians. It involves subcutaneous tissues and lymph nodes predominantly in the head and neck region and is characterized by angiolymphoid proliferation and eosinophilia. Otherwise it is known as eosinophilic lymphogranuloma. The first known report of Kimura disease was from China in 1937, when Kimm and Szeto1 identified seven cases of the condition. It is endemic in Asia. It is described in 1948 by kimura2 and noted a change in the surrounding blood vessels and referred to it as "unusual granulation" combined with hyper plastic changes in lymphoid tissue.

CASE REPORT
18 year old male Mr. Bharanidharan presented to hospital with complaints of two swellings one behind and other below and front of the right ear for past 3 years which is slow growing, painless, and not associated with any facial weakness. FNAC from the lesion behind the ear is suggestive of Kimura disease, and FNAC from the lesion in the region of parotid is suggestive of Lymph cyst or Warthin’s tumor. Peripheral smear shows eosinophilia (20%). Serum Immunoglobulin E level is 5000 IU/ml, blood urea, and serum creatinine level is normal. Ultra sonogram neck reveals right parotid enlargement with right post auricular lymph nodes. Case proceeded with right superficial parotidectomy and right post auricular lymphadenectomy. Post operative period uneventful. On follow up there is no recurrence till one year.
Figure 1: Fine needle aspiration cytology Low power view shows abundancy of lymphocytes and eosinophils

Figure 2: Pre operative picture

Figure 3: Intra operative picture

Figure 4: Post operative picture

Figure 5: Specimen of lymph node

Figure 6: Specimen of Parotid gland

Figure 7: Histopathology low power view Feature shows capsule, fibrous strands, lymphocytes, eosinophils, and vascular proliferation
DISCUSSION ETIOLOGY:
The cause of Kimura disease remains unknown. Reasons like an allergic reaction or an alteration of immune regulation are suspected. Other theories like persistent antigenic stimulation following arthropod bites and parasitic or candidial infection have also been proposed. To date, none of these theories have been substantiated.

PATHOPHYSIOLOGY: The pathophysiology of Kimura Disease remains unknown, although an allergic reaction, trauma, and an autoimmune process have all been implicated as the possible cause. The disease is manifested by an abnormal proliferation of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate suggest that Kimura Disease may be a hypersensitivity reaction. Some evidence has indicated that TH2 lymphocytes may also play a role, but further investigation is needed.

CLINICAL FEATURES: It is essentially located deep in subcutaneous tissues and in almost all cases involves the regional lymph nodes. It occurs in young adult males between 27 and 40 years. Male to female ratio of occurrence is 3:1. It is insidious in onset, enlarging nodular masses in the head and neck areas, most frequently infra or retro auricular region, which may simulate a neoplasm. The lesions are single in 60% of cases and multiple in the rest of cases, occasionally it is symmetric. Salivary glands particularly parotid, oral cavity, axilla, groin, and limbs are occasionally involved. It may be associated with nephrotic syndrome. Peripheral eosinophilia and elevated serum Immunoglobulin E are constant features. The skin and lymph node lesions do not ulcerate, and although recurrence is common, the disease has a benign course.

LABORATORY FEATURES:
Complete blood count with differential count almost always reveals peripheral eosinophilia in persons with Kimura disease (98%). The number of eosinophils was closely correlated to the sizes of the neck masses in one series. Serum immunoglobulin E levels are often elevated in persons with Kimura disease. Blood urea nitrogen, creatinine, and urinary protein levels should be obtained to exclude concomitant renal dysfunction and/or nephrotic syndrome. Serum eosinophil cationic protein levels parallel the course of the disease.

HISTOLOGY: Histopathology of Kimura disease shows lymphoid infiltrates with formation of follicular and germinal centers accompanied by plasma cells, mast cells, and particularly large amount of eosinophils in subcutaneous tissues. Thin walled capillaries are also present. Lymph nodes are enlarged to 1-4 cm in diameter and frequently adherent to one another; show markedly hyper plastic follicles with reactive germinal centers and a well defined peripheral mantle. Diffuse eosinophilia, eosinophilic micro abscesses and infiltration of germinal centers, some time result in folliculolysis.
Polykaryocytes of the Warthin-Finkeldey type characterized by overlapping, grape-like arrangement of nuclei are common, often with in the germinal centers. In areas of eosinophilia crystalline structures and sometimes been seen in cytoplasm of histiocytes or extruded in stroma as Charcot-Leyden crystals. Vascular hyperplasia mostly post capillary venules more pronounced in the mantle zone of germinal centers. Sclerosis develops in older lesion with vascular atrophy, sinus obliteration but eosinophilia still persists. IMMUNOHISTOCHEMISTRY: Immunoglobulin E deposits in germinal centers, also Immunoglobulin G, Immunoglobulin M deposits are demonstrated. Vascular endothelial cells strongly stain with Ulexeuropaeus agglutinin (UEA-1). The polykaryocytes are Warthin-Finkeldey type and features are probably of helper T-cell origin. CYTOPATOLOGY: FNAC shows dissociated and clustered endothelial cells, eosinophils, lymphocytes, Warthin-Finkeldey giant cells. Excision biopsy is recommended to confirm the diagnosis. DIFFERENTIAL DIAGNOSIS: Angiolymphoid hyperplasia with eosinophilia (ALHE) which is largely restricted to dermis. ALHE is marked by a proliferation of blood vessels with distinctive large endothelial cells accompanied by a characteristic inflammatory infiltrate that includes eosinophils.

Hodgkin’s disease with mixed cellularity is differentiated by Reed-Stenberg cell which is diagnostic and with absent germinal centers and Immunoglobulin E deposits.

Castleman disease which lacks eosinophilia. It has involuted hyalinized rather than hyper plastic germinal centers.

Dermatopathic lymphadenopathy which has hemosiderin, melanin, and lipid deposits

Cylindroma

Kaposi sarcoma

Mickulicz disease

Dermatofibrosarcoma protuberance

Pyogenic granuloma

Drug reaction

Parasitic infestation

RADIOLOGY Although not diagnostic, imaging studies can play a major role in aiding the diagnosis of KD and distinguishing the disease from other conditions. It is also useful in delineating the extent and progression of the disease. Radiological features, particularly on CT and MR imaging, have been reported to be useful in the diagnosis of KD. Tissues involved in KD, for example the parotid gland and lymph nodes, show intense enhancement on CT, reflecting the vascular nature of the lesions13-15. The borders are usually ill-defined and there is usually adjacent enhancing cervical lymphadenopathy. On MR imaging, these lesions demonstrate intermediate to high signal intensities on T1-weighted images and hyper intense signals on T2-weighted images.
TREATMENT:
The optimal treatment for KD is not well established. However, treatment should aim to preserve cosmeses and function while preventing recurrences and long-term sequelae. The range of treatment options includes conservative treatment, steroid therapy, radiotherapy, cryotherapy, laser fulguration, and surgical excision. Other therapeutic options, including antihistamines like cetirizine, cytotoxic agents, cyclosporin and pentoxifyline, have been used with variable results. At initial presentation, surgical excision is the choice for both diagnosis and therapy. The value of achieving negative surgical margins for local control in excision has not been studied. Nevertheless, the treatment outcome after excision is variable and recurrence is common. In cases treated with surgical excision alone, the recurrence can be as high as 25%. Localized recurrences can often be managed by surgical excision. However, if recurrence is frequent or there is symptomatic nephrotic syndrome, systemic steroids should be started. Steroid dosages should be initiated at high doses and then tapered to effect. Unfortunately, there is a tendency for lesions to recur when steroid therapy is stopped. For recurrent cases or lesions not amenable to surgery due to size or unacceptable resultant morbidity, radiotherapy can be considered. Low-dose local irradiation (about 25 to 30 Gy) has been reported to yield good control and obviates the need for long-term corticosteroids. However, in addition to the side effects of radiotherapy, there is also concern regarding secondary malignancies in the irradiated field.

CONCLUSION:
A high index of suspicion is required and Kimura disease should be considered in the differential diagnoses of head and neck masses especially if it is associated with adjacent cervical lymphadenopathy and peripheral blood eosinophilia.

REFERENCE:


