Research Paper

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Kimura's Disease: Case Report and Review of the Literature.

Medical Science

KEYWORDS : Kimura's disease(KD), eosinophilia, Ig E

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ABSTRACT

Kimura's disease (KD) is a chronic inflammatory soft-tissue disorder of unknown cause. It is a rare disease mostly reported from East and South East Asia. The typical presentation is in a young Asian male with nontender subcutaneous swellings in the head and neck region, lymphadenopathy and eczema . Investigations reveal raised Ig E, peripheral eosinophilia and frequently deranged renal functions usually nephrotic syndrome.It can impose a challenging diagnosis both clinically and pathologically .Treatment options range from conservative observation for asymptomatic patients toCyclosporine, Intravenous immunoglobulin (IVIG),Imatinib , steroid therapy, radiotherapy and surgical excision for symptomatic patients.In this article we present a case report of a rare disease with review of literature and recent trends in the management.

Introduction

Kimura disease is a rare, chronic inflammatory disorder that usually involves the subcutaneous tissues and lymph nodes in the head and neck region of young Asian males [1-5]. Till the mid of twentieth century terms Kimura's Disease and Angiolymphoid hyperplasia with eosinophilia (ALHE)were used interchangeably. Controversy existed regarding the pathophysiology of the disease. It is now hypothesized that an infection or toxin may trigger an autoimmune phenomenon or lead to a type I (immunoglobulin E [IgE]–mediated) hypersensitivity reaction. The usual clinical presentation is in a young Asian male with painless mass or masses in the head and neck region, with occasional pruritus of the overlying skin. With few cases reported in literature of this disease which presents as a clinical and pathological diagnostic dilemma,a typical case of this rare post auricular Kimura's Disease is reported.

Case Report

A 35 year old male presented to our department complaining of slowly enlarging mass in left post auricular region for past 8 years . He complained of itching at the area for which he took levocitrizine.Itching subsided with anti histaminics but recurred as soon as the drug was stopped.History revealed similar lesion at left post auricular region 10 years back for which he was operated.His medical history was unremarkable. A general physical examination was normal and local examination revealed 2cm *1 cm swelling in the left post auricular region. The swelling was single, smooth, skin coloured, regular margin ,smooth surface with a overlying surgical scar mark.

The swelling was non tender, normal temperature with firm consistency and was not attached to the underlying structures.

Total Ig E serum was done and values were 4603.2 IU/ml (normal values are <1.5-378.0).

Fine needle aspiration cytology was performed which showedpredominantly non specific reactive hyperplasia with abundant eosinophils,occasional multinucleated giant cells. These features suggested of eosinophilicgranuloma(kimuras disease?).

Ultrasound of the lesion showed multiloculated cystic lesion and multiple lymph nodes benign in character,largest of size 0.9 x 0.7 cm.

Excision biopsy was performed from left post auricular sub cutaneous swellings. The sections showfibrocollagenousstroma,sc anty fatty tissue and dense inflammatory infiltrate,composed of lymphocytes,plasma cells and focal few eosinophils. Focal lymphoid aggregate formation,focal crushing artefacts and focal myxoid changes noted.

There is no evidence of granuloma,parasite,malignancy.The histologic features were those of KD(Figure 1).

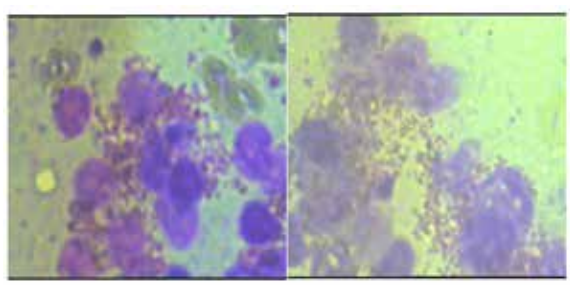

FIGURE 1: Histopathological slides

The patient was followed up for six months. No recurrence of the swelling noted.

Discussion

Kimura's Disese was first described in 1937 in China (Kim and Szeto[6] reported 7 cases of eosinophilic hyperplastic lymphogranuloma). The common term Kimura disease was coined in 1948; Kimura et al[1] used it in describing "an unusual granulation combined with hyperplastic changes of lymphatic tissue." Kimura's Disease being a rare entityfew cases (mainly among young Asian men[3,4]) have been reported under variousnames.Most cases have been reported in Asians with males more commonly affected than females , with a 3.5:1 to 9:1 male-to-female ratio in most series reported. The disease usually presents as nontender subcutaneous nodules and masses in the head and neck, especially in the parotid and submandibular regions. These lesions are often associated with lymphadenopathy[3]. KD may involve uncommon sites like orbit[7] (including the eyelids, conjunctiva, and lacrimal glands), paranasal sinuses, epiglottis, tympanic membrane, parotid gland, and parapharyngeal space[8,9],inguinal lymph nodes[10] and pulmonary hilar mass[10].

Several findings are characteristically associated with KD. Local examination reveal regional lymphadenopathy which was also detected ultrasonographically in our case.

Blood examination reveal peripheral blood eosinophilia[11] and raised Ig E levels[12]. In our reported case we find both raised Eosinophils as well as markedly raised Ig E. Renal involvement is noted in significant number of cases of KD. In a study conducted

in Japan in patients with KD, 13 (7%) also had nephrotic syndrome, and 8 (5%) also had proteinuria[13].

Imaging studies like CT Scan and MRI show variable appearance. One of the largest case series to date notes the characteristic findings to be multiple ill-defined, enhancing lesions around the parotid gland, with associated lymphadenopathy[14].

Histologically, KD is characterized by dense fibrosis, lymphoid infiltration with reactive follicles, and a mixed inflammatory cell infiltrate with numerous eosinophils[15], all of which can develop in subcutaneous tissue, salivary glands, and lymph nodes. Some authors view KD as having 3 components—cellular, fibrocollagenous, and vascular[16]. The conspicuous

feature of the cellular component isdistinct lymphoid follicles, consisting mainly oflymphocytes. The fibrocollagenous component is formed by the infiltrate with numerous eosinophils, and eosinophilicmicroabscesses are common; mast cells and plasma cells also abound; fibrosis is a constant, even in young lesions. The vascular component consists of proliferating and swollen endothelial cells, but these do not have atypical nuclei or abundant eosinophilic cytoplasm; salivary glands are frequently involved (they experience

parenchymal atrophy and fibrosis); regional lymph nodes, usually enlarged, experience follicular hyperplasia with increases in eosinophils with or without fibrosis.

The etiology of KD remains dicey. The disease is almost always associated with peripheral eosinophilia and raised IgE. Occasional association with renal disease ;mesangial proliferation, eosinophilic infiltration and podocyte confusion are prominent histological features in one of the studies[17]. The above findings suggest an allergic or autoimmune process.

Various treatment modalities have been tried like oral corticosteroids, Cyclosporine[18,19], Intravenous immunoglobulin (IVIG)[20] ,oral pentoxifylline[21] . Radiotherapy has occasionally been used to treat recurrent or persistent Kimura disease lesions[22].

Radiotherapy, systemic steroid therapy and conservative management have all proved to be unsatisfactory treatments for KD. After surgical resection and variable follow-up time, the KD lesion tends to recur in as much as one fourth of patients[3].

In the view of the above facts, most experts treating a KD lesion advocate surgical resection.[23]

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