Kimura's Disease in the West-Case Report and Review of Literature

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Abstract: We report a 58 yr old lady, who had right temple swelling for 2 years, which on excision biopsy showed features of vasculitis. During the Rheumatology review, Clinical evaluation didn't reveal any features suggestive of Giant cell arteritis and inflammatory markers were normal. She had localised granulomatous changes without giant cells and pathologist gave the opinion as Kimura's disease. It has been described in East Asia especially in Japan as Juvenile Giant cell vasculitis (also known as Kimura's disease (KD). It was reported by Kimura *et al*. as chronic inflammatory disorder of unknown aetiology with granulomatous changes mimicking vasculitis but no giant cells. It is very rare in Caucasians and early diagnosis will result in appropriate treatment and sparing the immunomodulatory treatment. This case is presented to increase awareness of KD and to highlight the features which may aid the diagnosis.

Keywords: Kimura, Lymphadenopathy, Eosinophilia, Vasculitis, Granuloma.

BACKGROUND

A referral to rheumatologist with a swelling on the temple generally implies Giant cell arteritis, but we report a case of a less well known disorder, Kimura Disease (KD). KD is a chronic inflammatory disorder of unknown aetiology which generally presents with a characteristic triad of symptoms and signs. These are a painless slowly enlarging soft tissue mass, associated lymphadenopathy and mostly peripheral eosinophilia.

CASE SUMMARY

A 58 year old Caucasian woman, completely well in the past was referred for removal of a slow growing swelling on her right temple for cosmetic reasons. She has had a 1 cm swelling on her right temple for almost 2 years. Ultrasound suggested an aneurysm of the anterior branch of the right temporal artery and she underwent excision of the swelling. The histological appearances were suggestive of vasculitis and she was referred urgently to rheumatology for an assessment.

On review of her symptoms, there was no pain or tenderness over the temporal arteries. She denied any history of jaw claudication, visual or PMR symptoms and there were no features to suggest systemic vasculitis. There was no palpable lymphadenopathy. She didn't have any bruit and systemic examination was unremarkable. She was a smoker and had high cholesterol. All her investigations were normal including full blood count (neutrophils, eosinophils, lymphocytes, platelets and haemoglobin all were normal), electrolytes, liver functions tests, inflammatory markers- CRP and plasma viscosity and antinuclear antibody. Immunoglobulin subclass study was normal. Urine didn't reveal any hematoproteinuria. Chest roentgenogram was normal too.

In view of the negative history and normal serology, the histology report was reviewed. The description was of a medium sized muscular artery with intimal fibroplasias, luminal constriction, neovascularisation and destruction of the internal elastic lamina. This was associated with a sprinkling of eosinophils, plasma cells, and chronic inflammatory cells. No giant cells were seen. The aetiology of the arteritis was uncertain, but similar changes have been described in so-called "juvenile temporal arteritis or *Kimura disease."* (KD).

DISCUSSION

It is important to be aware of the temporal artery vasculitis which is increasingly more common. This patient was entirely asymptomatic which prompted us to review the biopsy with the pathologists.

This case is presented to increase the awareness of KD and to highlight features which may aid the diagnosis of this condition. Prior to biopsy, diagnosis of Kimura's disease was not considered in our patient because of its rarity in the West. With its predilection for the head and neck region and its variability in presentation, it can frequently be mistaken for other

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conditions. A high index of suspicion is required and KD should be considered in the differential diagnosis of head and neck masses. In general, KD is rare in Caucasians and there is often peripheral eosinophilia and elevated serum IgE. Neither was present in our patient, but the measurements were done at the time of regression. Hence, the levels during active disease were unknown.

Kimura's disease is a chronic inflammatory disorder of unknown aetiology, first reported by Chinese authors Kimm and Szeto in 1937 [1, 2], but the definitive description was published by Kimura *et al.* in Japan in 1948. KD is most prevalent in Asians, uncommon in Caucasians and rare in Blacks. Eighty-five per cent of cases occur in men, with six times as many males being affected than females. KD typically affects patients in their second to third decades of life, with the median age of 28 years in one series. It is often accompanied by markedly elevated serum immunoglobulin E (IgE) levels [3, 4]. There is a high incidence of renal involvement, notably proteinuria and nephrotic syndrome [5]. Arthropathy, Asthma [6] and raynauds have also been reported [7]. There have been suggestions about infections triggering the disease, some form an unusual atopic response or toxins as the precipitating event [8]. Elevated levels of interleukin (IL)-5, IL-4 and IL-13 have been found in peripheral blood mononuclear cells and IL-5 in lymph nodes of affected patients. Histologically, the lesions are characterized by reactive lymphoid follicles with eosinophilic infiltration, sometimes forming eosinophilic abscesses [4, 9, 10, 11]. The main differential diagnosis is angiolymphoid hyperplasia with eosinophilia (ALHE), giant cell arteritis, panarteritis nodosa, Takayasu arteritis, and isolated vasculitis of the vasa vasorum.

The treatment of choice for localized disease is surgical excision [1], though conservative treatment in the form of intralesional steroid injection or the use of radiotherapy [12] in mostly anatomically sensitive areas such as the periorbital area [13] has been found to successful. In two cases, pranlukast, a leukotriene receptor antagonist, (450 mg/day) was reported to be effective treatment for KD, without any adverse side effects. Despite steroid discontinuation, this patient remained disease-free at follow-up 6 months later.

PRACTICAL MESSAGE

Early diagnosis of KD could spare the patient unnecessary and potentially harmful diagnostic procedures in addition to extensive therapy and anxiety.

KEY MESSAGE

Kimura‟s disease is a rare differential diagnosis to consider in evaluating giant cell arteritis.

It is one of the pulmonary-renal syndromes with raised Eosinophils and IgE levels.

DISCLOSURE STATEMENT

The Authors have declared no conflict of interest.

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