

Recurrent Kimura's Disease: Report of One Case

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Kimura's disease (KD) is a rare, chronic inflammatory disease, and the etiology is not yet to be determined. The disease is usually found in young Asian men but rarely in Western population. Here, we report an 8-year-old boy with the initial presentation of right upper eyelid swelling. Laboratory tests disclosed peripheral eosinophilia and elevated IgE level. KD with lacrimal gland involvement was diagnosed. Surgical excision was performed several times and steroid was also used, however the disease recurred. Thereafter, steroid, cyclosporine-A and azathioprine were prescribed. The tumor mass resolved and the laboratory data returned to normal level. (Acta Paediatr Tw 2007; 48: 149-51)

Key words: Kimura's disease, eosinophilia, IgE, eyelid mass

INTRODUCTION

Kimura's disease (KD) is a rare, chronic inflammatory disease of unclear etiology.¹ The disease is usually found in young Asian men but rarely in Western population.^{2,3} The typical clinical features include tumor-like lesions in the soft tissue and lymph nodes of the head and neck or in the parotid glands. Upper limb involvement was also reported before. KD is often accompanied by regional lymphadenopathy, peripheral eosinophilia, and elevated serum IgE level. Histopathologic features are characterized by abnormal proliferation of small vessels, associated with prominent infiltration of eosinophils, plasma cells and scattered lymphoid follicles.^{4,5} The lesions in Kimura's disease are benign, and have been occasionally misdiagnosed as malignant tumors or other invasive diseases. Although the prognosis is usually good, renal involvement presenting with nephrotic syndrome is sometimes reported.⁶ In the present report, we describe a child of KD with the initial presentation of right upper eyelid swelling.

CASE REPORT

This 8-year-old boy was quite well until he was 5

years old. Right upper eyelid swelling has been noted since then. The mass was movable and non-tender. He visited a tertiary medical center and mass excision was performed. Orbital pseudotumor was diagnosed. After excision, oral steroid was prescribed for several months and then discontinued due to side effect of vomiting. Because the residual mass was still noted, the patient was then referred to National Taiwan University Hospital to receive tumor excision again. On histologic examination of the specimen, abundant eosinophil infiltration with eosinophilic abscess formation of the lacrimal glands was noted. In addition, lymphoid infiltrates in germinal centers and some thick-walled vessels were also present. Accordingly, KD with lacrimal gland involvement was diagnosed. After the surgery, a right upper eyelid mass recurred one year later. There were also a movable mass in the left upper arm with left axillary lymphadenopathy. Computed tomography disclosed enlarged right lateral rectus muscle with whole muscle bundle involvement. The globe was displaced (Fig. 1A). Laboratory examinations revealed peripheral eosinophilia (36% of total white blood cells (WBC)). Orbital tumor and left arm mass excision were performed. The histopathologic findings were, as previously described, eosinophil infiltration and follicular hyperplasia (Figs. 2A, 2B). Three months later, right upper eyelid swelling

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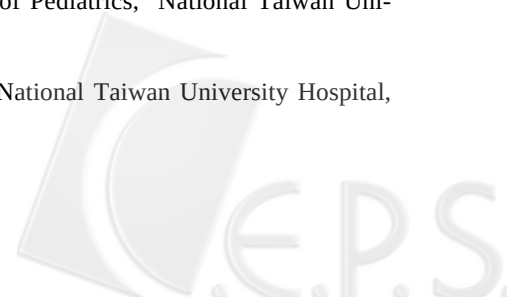
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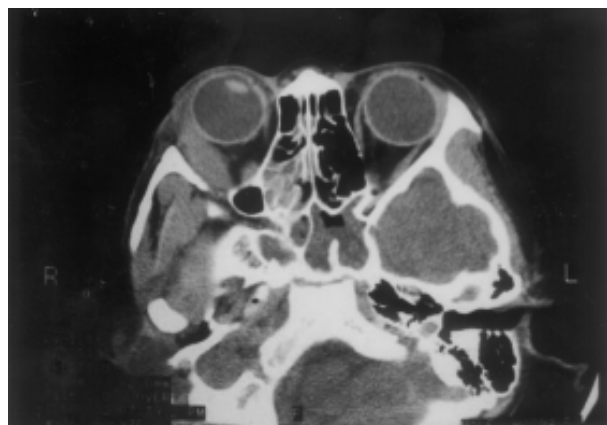


Fig. 1A. Computed tomography disclosed enlarged right lateral rectus muscle with whole muscle bundle involvement. The globe was displaced.

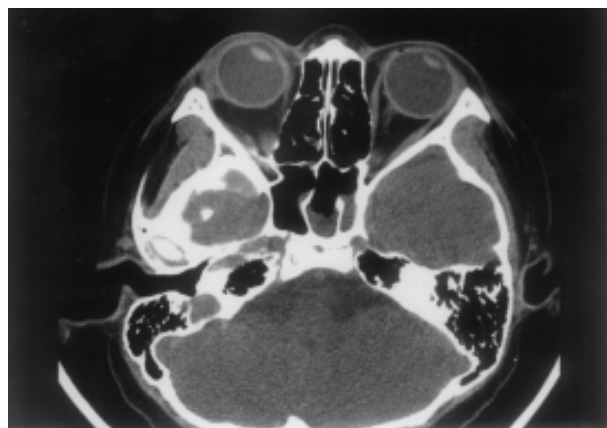


Fig. 1B. Resolution of the soft tissue mass around lateral rectus muscle in the right orbital cavity.

developed again with limitation of right eye movement. Eosinophilia (21% of total WBC) and raised IgE serum level (>2000 IU/dL) were noted. Computed tomography revealed increased soft tissues with moderate enhancement at the right orbital cavity around the lateral rectus muscle and right lacrimal gland. There were also multiple lymphadenopathies in the bilateral axillary portion and a nodular lesion (1 cm) at the medial aspect of the left middle humerus. Under the impression of recurrent KD, parenteral steroid (60 mg/day) was prescribed for 3 days and then tapered to lower dose. Because of the side effect of long-term usage of steroid, cyclosporine-A (2 mg/kg/day) and azathioprine (1.5 mg/kg/day) were added. Then we tapered down the dose of steroid as rapidly as possible. Two months after treatment, the lesions had resolved completely (Fig. 1B), with normal peripheral eosinophil count (1.8% of total WBC).

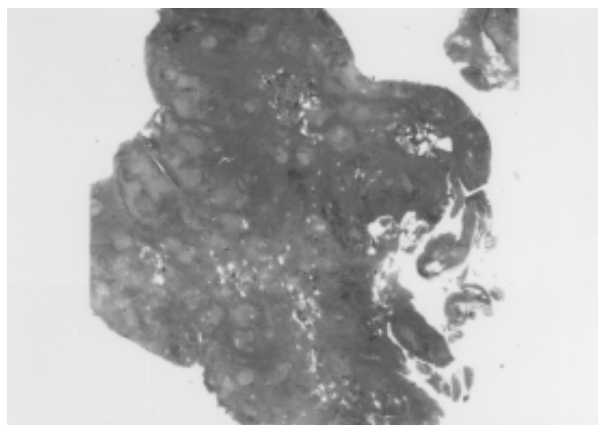


Fig. 2A. A lymph node with follicular hyperplasia. (40 $\times$)

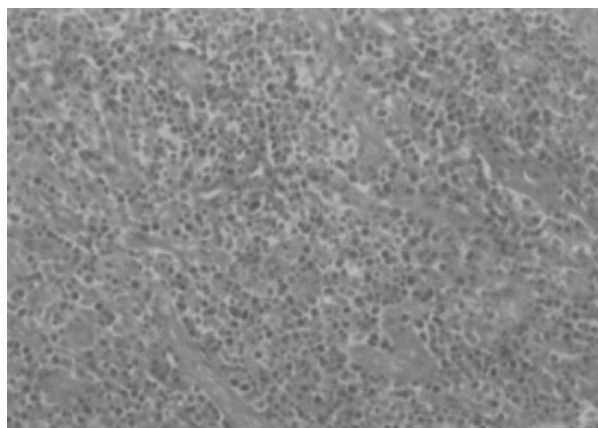


Fig. 2B. Abundant eosinophilic infiltrate and proliferative of postcapillary venules. (100 $\times$)

DISCUSSION

KD was first described in China in 1937 as “eosinophilic hyperplastic lymphogranuloma”,^{2,3} and then became well known as “Kimura’s disease” after a systematic description in 1948 by Kimura et al.^{1,4}

The etiology and pathogenesis of KD are not clear. The pathology of KD is characterized by prominent germinal centers in involved lymph nodes containing cellular, vascular, and fibrous components.^{4,7} The tumor-like masses usually contain predominant eosinophilic infiltrate, proliferative capillary manifesting as masses of canalized swollen endothelial cells, and fibrosis. Together with peripheral eosinophilia, it suggests that KD may represent a hypersensitivity reaction to unknown

stimuli. Type II T helper cell cytokines like IL-4, IL-5, and IL-13, have also been found to relate to KD. In addition, like our patient with allergic rhinitis and strong positive response to the allergens, most KD patients have co-morbidity of allergic disorders such as asthma, allergic rhinitis, and atopic dermatitis.^{8,9}

Until now, there is still no standard treatment for KD. Conservative surgical excision is considered the treatment of choice. Although KD is usually benign and self-limited, recurrence is not uncommon.^{10,11} Other therapeutic options, including radiation, systemic corticosteroids, cytotoxic agents, and cyclosporin-A, have all been reported with variable responses.^{12,13} Systemic steroid may be considered in cases with frequent relapses or complicated with nephrotic syndrome.⁴ Radiotherapy has been used to treat recurrent or persistent lesions.^{14,15} Cyclosporine-A has been reported to induce remission in a patient with KD with earlobe involvement.¹³ Besides, cetirizine has been reported to induce a complete remission within 2 months of treatment after gradual withdrawal of steroid.¹⁶

Our patient suffered from post-excision relapses three times. Under full-dose steroid treatment for three days, the lesion decreased in size and the limitation of movement of right eye improved. Because of the concern of side effects developed by large amount steroid and the regrowth of the lesion after discontinuing steroid treatment, we tapered steroid and added cyclosporine-A and azathioprine. The follow-up images disclosed complete remission of the disease. Importantly, this therapeutic strategy may be a choice of treatment for recurrent KD.

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