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Unilateral Orbital Kimura's Disease in a 79 Year-Old Patient

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We report a rare case of an elderly patient with Kimura's disease. A 79-year-old man had a subcutaneous mass on his left upper eyelid. The eyelid was swollen, and there was mild dermatochalasis of the left eye. Excisional biopsy and blepharoplasty were performed, and pathology identified the mass as Kimura's disease. To the best of our knowledge, this is the oldest such patient in Taiwan. Kimura's disease should be suspected in any age group if a painless unilateral adenopathy, hypereosinophilia and peripheral hyperimmunoglobulin E are present.

Key words: Kimura's disease, hypereosinophilia, painless adenopathy, hyperimmunoglobulin E

INTRODUCTION

Kimura's disease is a rare entity of uncertain etiology that occurs mainly in Asians. This chronic condition primarily involves the subcutaneous tissue, parotid glands and lymph nodes. Orbital cases are infrequent and may cause a devastating visual outcome if not properly treated. Among reported cases, patients with orbital Kimura's disease tend to be older (30-50 years) than the typical teenage presentation. The present case is unique in that the patient was a 79-year-old man. There are few cases reported in this age group in the literature. Based on this case, we have reevaluated the diagnostic criteria for Kimura's disease.

CASE REPORT

The patient, a 79-year-old-man, presented with a palpable mass that had been growing insidiously over his left upper orbit for one year. He had experienced pruritus with accelerated growth of the mass over the previous six months. Examination revealed a round firm mass, measuring $1.5 \times 2 \times 0.5$ cm^3 on the front part of the eyelid (Fig. 1A). The overlying skin was stretched but with no lesions. No other abnormalities were noted on ocular examination. The results of systemic examinations were unremarkable. Laboratory results indicated an eosinophil count of 22%, a serum immunoglobulin (Ig) E level of 110 IU/mL (normal 0-87 IU/mL) and proteinuria of 10mg/dL. Computed tomography scanning showed an orbital mass that involved the upper eyelid (Fig. 1B). An excisional biopsy of the lacrimal gland was performed. Scattered lymphoid follicles with germinal centers, surrounded by lymphocytes and plasma cells, were evident on pathology sections. There are also many eosinophils in the interfollicular areas, with many high endothelial venules (see Figs 2A and B). Immunohistochemistry revealed no myogenous or oncogenous origin. Based on the peripheral hypereosinophilia, elevated serum IgE and pathology results, we made a clinical diagnosis of Kimura's disease. The patient was treated with 60 mg/day of prednisolone for one week. No lesion or swelling of the left upper eyelid was apparent at the five-month follow up.

DISCUSSION

Kimura's disease usually appears in the head and neck region. Other possible sites of involvement are the orbits, paranasal sinuses and kidneys[1]. Renal involvement can manifest as concomitant proteinuria and nephritic syndrome.

Microscopically, the nodes involved show marked hyperplasia of the germinal centers. These germinal centers are often well vascularized and contain polykaryocytes, interstitial fibrosis and deposits of proteinaceous material. There is also extensive infiltration by mature eosinophils, with the occasional formation of eosinophilic abscesses[2].

There is controversy in the literature as to whether Kimura's disease and angiolymphoid hyperplasia with eosinophilia (ALHE) are the same entity. Patients with

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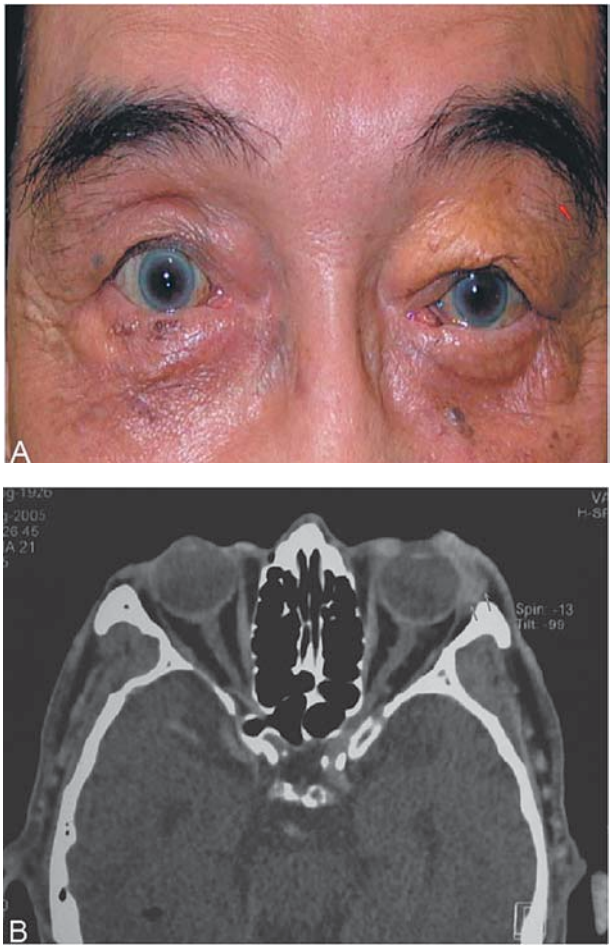

Fig. 1 A. The lesion was a single round, firm mass covering an area of approximately 2 cm in diameter on the upper part of the left orbit (arrow). B. Orbital computed topography, coronal section; note the enhancing mass lateral to the left orbit (arrow).

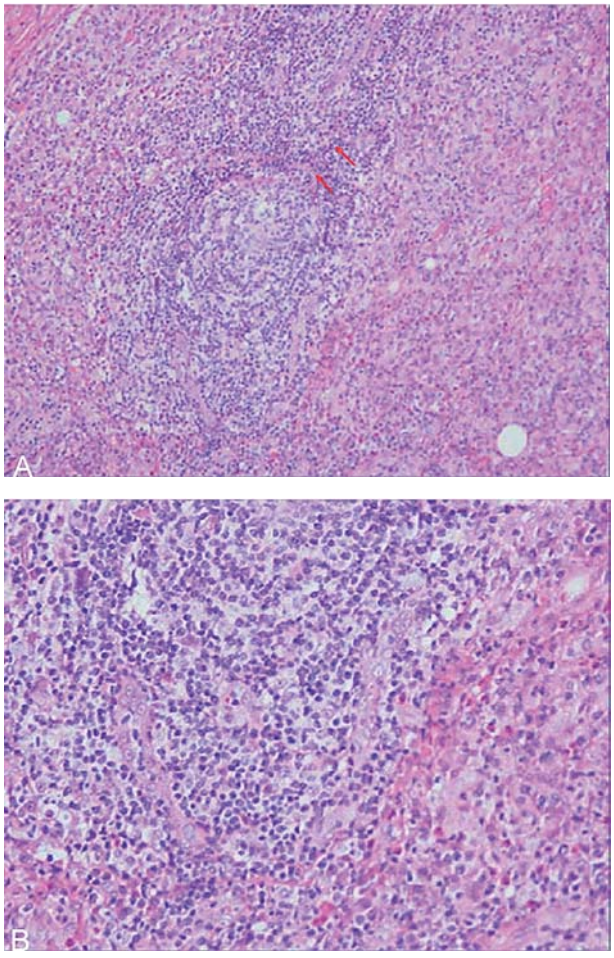

Fig. 2 A & B Lymph node involvement in this case of Kimura's disease. There is follicular hyperplasia and massive perinodal infiltration composed mainly of eosinophils (arrow). (Hematoxylin & eosin staining, 36% reduction; original magnification 10a±A and 25±B).

ALHE tend to be older than those with Kimura's disease: in the third or fourth decade of life. There is elevated serum IgE in patients with Kimura's disease but not in those with ALHE[2]. Thus, serum IgE level is of diagnostic value in differentiating these diseases.

Periocular Kimura's disease may have devastating outcomes, with deformed appearance and visual loss resulting from exposure keratitis[3]. Therapies for ocular and adnexal Kimura's disease include observation, excision, steroids, radiation, or a combination of these. Steroids are particularly effective in treating lesions when renal involvement is present, but there is a tendency for the disease to recur after their withdrawal. Surgery has a role in providing a diagnosis and in the excision of large cosmetically unacceptable masses[3]. Radiation is prescribed for lesions refractory to steroids or when surgery is not possible.

Kimura's disease can occur at any age, but the peak age of onset is in the second and third decades, ranging from 7-59 years[4]. Patients with orbital Kimura's disease are usually older (30-50 years) than the typical (teenage) patient[5,6]. It rarely occurs in elderly patients. Therefore, we report this case to increase awareness of this condition, as the disorder should be suspected when the clinical triad of a painless adenopathy, hypereosinophilia and hyper-IgE is present.

REFERENCES

1. Rajpoot DK, Pahl M, Clark J. Nephrotic syndrome associated with Kimura disease. Pediatr Nephrol 2000; 14:486-488.
2. Hui PK, Chan JKC, Ng CS, Kung ITM, Gwi E. Lymphadenopathy of Kimura's disease. Am J Surg Pathol 1989;13:177-186.
3. Moroz I, Rosen N, Rosner M. Bilateral Kimura's disease with devastating visual outcome. Eye 1998;12: 102-103.
4. Ronald R, Buggage C, Spraul W, Wojno TH, Grossniklaus HE. Kimura's disease of the orbit and ocular adnexa. Surv Ophthalmol 1999;44:79-91.
5. Chang B, Ryan D, Kennedy S, O'Connor G. Orbital Kimura's disease in a white child. Br J Ophthalmol 1999;83:1198-1199.
6. Milton AG, Narayan BP, Rajendra JS, Joshua R, Lakshmy V, Young JC. Kimura's disease: a case report and literature review. J Surg Oncol 1999;70:190-193.