

A Case of Kimura's Disease Presenting as a Rhinophyma-like Configuration

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Kimura's disease is a benign, uncommon, chronic inflammatory condition that usually presents with painless subcutaneous nodules or plaques. Head and neck are the most frequently involved sites in Kimura's disease. Mandible is the most commonly involved, followed by neck, cheek, scalp and forehead. Other possible sites are oral cavity, inguinal area and extremities, but there have been no reports involving the nose, especially the one that looks like a rhinophyma. We describe a case of Kimura's disease presenting like a rhinophyma. (Ann Dermatol 15(2) 85~88, 2003).

Key Words : Kimura's disease, Rhinophyma

Kimura's disease is an idiopathic chronic inflammatory disorder that predominantly affects young men of Asian descent. The disease is usually characterized by asymptomatic solitary or multiple subcutaneous nodules or plaques on the head and neck. Although the disease is predominantly found in Asian populations,¹ there are occasional cases reported among Caucasians and other ethnics. We describe a case of Kimura's disease which clinically looks like a rhinophyma.

CASE REPORT

A 71-year-old Korean man presented with a rapidly growing mass without a distinct border on the nose that had increased in size over a 3-month pe-

riod. The surface of the mass was pitted and showed telangiectasia, but there was no erythema. He denied any pain or discharge from the site. Examination revealed a 2.0 × 2.5cm-sized skin-colored mass located on the tip of the nose (Fig. 1). No additional nodules, papulopustules, comedones, nor episodes of flushing were noted. He had taken oral antibiotics for 3 months, prescribed by his primary physician who had considered the lesion as a rosacea, but there was no clinical improvement. Laboratory results showed 3.5% of eosinophil count in the peripheral blood, and the bony structures of the nose showed no remark on the plain X-ray. Magnetic resonance imaging (MRI) had showed a 2.0 × 2.3cm-sized well-circumscribed subcutaneous mass.

The patient underwent an excisional biopsy of the lesion and had an unremarkable postoperative course (Fig. 2). Microscopically, histologic sections revealed many lymphoid follicles with germinal centers and proliferated small vessels (Fig. 3) embedded in a dense infiltration composed of small lymphocytes, plasma cells, histiocytes, and eosinophils (Fig. 4). It was consistent with Kimura's disease. There has been no sign of recurrence after a

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follow-up period of 5 months.

DISCUSSION

Kimura's disease mainly affects young Asian men, although it has been reported in other areas and ethnic groups. The most common presentation is discrete nodules or localized swelling with or without pain and pruritus. The lesion primarily involves the subcutaneous tissue and sometimes the salivary glands and lymph nodes. Kimura's disease occurs insidiously and the amount of swelling at the affected site gradually increases over a period of months or years. There is no evidence of malignant transformation. Peripheral blood eosinophilia and increased level of serum IgE are the characteristics of this disease.

Since Kimura *et al.* defined this disorder 50 years ago,² approximately over 30 cases have been reported in Korea. Head and neck are the most frequently involved sites in Kimura's disease. Within the sites, mandible is the most commonly involved, followed by neck, cheek, scalp and forehead. Other possible sites are oral cavity, inguinal area and extremities, but it seems that there have been no reports involving the nose, especially the one that looks like a rhinophyma.

Rhinophyma consists of hypertrophic, hyperemic and large nodular masses. The bulbous nose develops over many years as a result of progressive increase in connective tissue, sebaceous gland hyperplasia, ectatic veins, and chronic deep inflammation. In its glandular form, the surface is pitted with deeply indented and mildly distorted follicular

orifices. Humps and sulci occur. In the fibroangiomaticous form, the nose is copper-red to dark red, greatly enlarged, edematous, and covered by a network of large, ectatic veins³. Prior to the biopsy, we had not considered the diagnosis of Kimura's disease in our patient because it clinically resembled a rhinophyma.

The pathogenesis of Kimura's disease is still unknown, but the cause is believed to be either immune mediated or infective (parasitic or fungal). The deposition of immune complexes seen on immunofluorescence lends credence to the notion that this condition occurs secondary to immunological injury⁴. It is likely that environmental or exogenous agents may incite an immune response⁵. Our patient had been in good health without any trauma on the nose.

Kimura's disease is associated with a high incidence of renal disease, and the nephrological manifestations include a diverse spectrum of histological lesions. Proteinuria was reported in 12%-16% of patients and 60%-78% developed nephrotic syndrome. Various renal pathologies such as membranous nephropathy, minimal change disease, mesangioproliferative glomerulonephritis, IgM nephropathy, and IgA nephropathy have been associated with Kimura's disease⁶. Beside renal diseases, bronchial asthma, serous retinal detachment, and chorioretinal atrophy may be associated. In this case, we could not find any evidence of nephropathy.

Kimura's disease can be confused with angiolymphoid hyperplasia with eosinophilia (ALHE). These two diseases have some common histologic features, such as eosinophilia and vascular proliferation. However, ALHE is found in an older, predominantly female population with smaller, circumscribed, intradermal, or more superficial lesions. The prominent feature of ALHE is vascular proliferation, but lymphoid follicle formation is more marked in Kimura's disease. The plump (cuboidal to dome-shaped) endothelial cell characteristic of ALHE probably remains the key to separation of these entities^{7,8}. In this case, histologic sections revealed prominent formation of lymphoid follicles with germinal centers and proliferated small vessels (Fig. 3) embedded in a dense inflammatory infiltration. The endothelial cells that line the blood vessels were usually flat, sometimes swollen. Mitoses were rare. Therefore, we diag-

nosed it Kimura's disease.

The treatment of Kimura's disease is not well established. Surgery has a pivotal role in providing the diagnosis and for excising large, cosmetically unacceptable masses. Kimura's disease responds well to oral steroids, even though steroid withdrawal can result in recurrence on the lesion and a certain proportion of lesions are refractory to this treatment modality. Radiation is reserved for recalcitrant and large lesions. Cryotherapy, electrodesiccation, and laser fulguration have been tried with variable outcomes^{9,10}. Our patient underwent excisional biopsy and there have been no signs of recurrence.

We present this case to increase the awareness of Kimura's disease and to discriminate this unusual manifestation from other diseases, such as a rhinophyma and subcutaneous tumors of the nose.

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