A Case of Kimura's Disease with an Unusual Location and Clinical Manifestation

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Kimura's disease is a chronic inflammatory condition of unknown cause. It usually appears as a painless nodule in the head and neck region often associated with regional lymphadenopathy, but there have rarely been reports of penile involvement or clinical presentation as an ulcerative plaque.

We present a case of Kimura's disease occurring on the penile shaft as an ulcerative plaque in a 44-year-old man. A vesicular eruption had occurred on his penile shaft followed by an ulcerative plaque for 2 months. He had suffered from recurrent regional lymphadenopathy for 15 years and he had also exhibited intermittent eosinophilia. Histopathologic findings revealed diffuse eosinophilic infiltration in the dermis of the skin and eosinophilic abscesses in the lymphoid follicles of the inguinal lymph node. After short-term systemic steroid therapy and local injection of triamcinolone acetonide, the ulcerative plaque was healed.


Key Words: Kimura's disease, Penile lesion, Ulcerative plaque.

Kimura's disease is an uncommon chronic inflammatory disease of unknown etiology which was described by Kimura in 1948. It is generally manifested as tumor-like nodules in the subcutaneous tissue of the head and neck.1,2,3

In western literature, cases with similar clinical and pathological features have been reported under the heading of angiolymphoid hyperplasia with eosinophilia.4,5,6,7 The relationship of both diseases has often been debated. Many reports have used the term synonymously4,5,8, although recently, many authors tend to think of them as separate entities.2,3,9,10 To the best of our knowledge, there have been only two reports of penile involvement and no reports presenting as an ulcerative plaque in English literature.

In Korea, Kimura's disease was first reported by Kim et al. in 1975.11 Since then, approximately 18 cases of Kimura's disease and angiolymphoid hyperplasia with eosinophilia have been reported, which are usually considered to be identical. In all of the reported cases, lesions were located in the head and neck area except two cases in the extremities and a case in the groin.12

We herein describe a case of Kimura's disease, presented as an ulcerative plaque on the penile shaft.

REPORT OF A CASE

A 44-year-old man was seen with an ulcerative plaque on his penile shaft. Three years ago, he found recurring vesicles on his penile shaft, which persisted for about 2 weeks on each occasion. Two months prior to admission, the vesicular eruption developed again, which progressively enlarged and ulcerated. He had also had painful swelling of both inguinal lymph nodes for 15 years. On the first examination, a walnut-sized ulcerative and oozing plaque on the penile shaft was discovered. It was covered with an easily detachable, yellowish mem-

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brane and surrounded by an erythematous elevated induration (Fig. 1). Lichenified, dry scaly patches were found on his scrotum. Multipleinguinal lymph nodes were palpable, freely movable and nontender. The complete blood count revealed intermittent eosinophilia (1850/mm$^3$ 20% of WBC initially; 360/mm$^3$ 4% of WBC, 10 days after; 3750/mm$^3$ 33.3% of WBC, 3 months after). The Ig E level measured by PRIST was more than 800 IU/dl and the test for anti-streptolysin O was weakly reactive. The other laboratory findings including the liver function test, renal function test, serologic test for syphilis, Coombs' test, antimicrosomal antibody, anti-ds DNA, EKG, and chest X-ray were within normal limits or were negative.

Histopathologically, massive and diffuse infiltration of eosinophils with some neutrophils and plasma cells were observed on the edematous base of the ulcer as well as in the whole dermis. There were fibrin deposits in the upper dermis. Endothelial cell swelling was observed but there was no evidence of vasculitis or atypical endothelial cells (Fig. 3).

A biopsy specimen from the inguinal lymph node also showed eosinophilic infiltration around lymphoid follicles, sometimes forming an eosinophilic abscess on several lymphoid follicles (Fig. 4). Short-term systemic steroid therapy with an initial dose of prednisolone 30mg a day followed by an intralosomal injection of triamcinolone acetonide made him almost free of the lesions (Fig. 2).

**DISCUSSION**

Kimura's disease occurs most often in the head and neck areas. The report of penile involvement in Kimura's disease has been uncertain because of the confusion regarding the diagnosis of Kimura's disease and angiolymphoid hyperplasia with eosinophilia. Olsen et al. had described a case of penile involvement in 116 patients, but it was not clear whether it was Kimura's disease or angiolymphoid hyperplasia with eosinophilia. Rao et al. reported a case of angiolymphoid hyperplasia with eosinophilia of the penis, although he treated the two diseases as the same one. In English and Korean literature, there have been no reports of Kimura's disease occurring on the penile shaft.

The morphological feature of the present case
also deserves mention. Usual cases of Kimura’s disease present as dome-shaped papules or subcutaneous masses. Manifestation as an ulcerative plaque seems to be unusual in most cases.

Our patient had recurrent lymphadenopathy for 15 years, before the skin lesion developed. We thought that it was also a part of Kimura’s disease, because histopathological findings of the lymph node were compatible with the disease and actually, recurrent lymphadenopathy can be the sole initial manifestation.\(^{13}\)

A biopsy specimen of our case showed numerous eosinophilic infiltration and swollen endothelial cells without atypism. In the lymph node, there were eosinophilic abscesses in several lymphoid follicles.

Differential diagnosis includes those diseases with eosinophilic infiltration such as insect bites, eosinophilic cellulitis, eosinophilic granuloma, eosinophilic leukemia and eosinophilic pustular folliculitis. However, any of these diseases can hardly show such heavy infiltration of eosinophils in the skin and lymph node, and so we could rule out the above.

Although our case did not show lymphoid follicles in the skin biopsy specimen, that is not essential for making the diagnosis. There have been reports of Kimura’s disease without lymphoid follicle formation in the skin.\(^1\) Fibrosis of the dermis is considered to develop late in the course of the disease.

Kimura’s disease has been treated by systemic and local steroid therapy as in our case. It can also be treated by surgical excision, X-ray therapy, electrosurgery or cryotherapy.\(^4,7\)

Although the etiology of Kimura’s disease is still not understood, an allergic origin has been suggested on the basis of eosinophilia, elevated serum Ig E levels, and its nephropathic associations.\(^{15}\) In the lymph node, intensive staining of the germinal center with anti-IgE was found.\(^{1,13}\) On immunologic blood examination, the proportion of Ig E-bearing lymphocytes was elevated above the normal range.\(^{17}\) The follicular structure in Kimura’s disease appeared immunohistochemically to be derived from a reactive inflammatory process\(^{10}\) and Isoda\(^9\) had detected eosinophil chemotactic activity in an extract of tumor tissue from a patient with Kimura’s disease.

There has been a lot of controversy regarding the relationship between Kimura’s disease and angiolymphoid hyperplasia with eosinophilia. In 1969, Wells and Whimster\(^4\) proposed that Kimura’s disease was part of the spectrum of angiolymphoid hyperplasia with eosinophilia and many reports in the literature adopted that viewpoint.\(^4,3,44\) But
other\(^2,^3\) insisted that the two diseases are distinct entities. They noted that clinically, Kimura's disease occurs most often in young males with the onset of lesions between adolescence and the fifth or sixth decade, whereas angiolymphoid hyperplasia with eosinophilia commonly affects middle-aged females. Furthermore, peripheral blood eosinophilia, elevated IgE levels and lymphadenopathy are observed in a majority of the patients with Kimura's disease but rarely or never in a patient with angiolymphoid hyperplasia with eosinophilia.

On histopathologic examination, Kimura's disease is characterized by dense fibrosis, lymphoid infiltration with numerous eosinophils in the subcutaneous tissue, salivary gland and lymph nodes, sometimes forming an eosinophilic abscess in the lymph node. In contrast, angiolymphoid hyperplasia with eosinophilia is seen as circumscribed proliferation of small blood vessels with plump endothelial cells accompanied by a mixed inflammatory infiltration in the dermis or subcutaneous tissue. Endothelial cells are described as plump, atypical or histiocytoid, resembling malignant vascular neoplasm.

Clinically, our case favors the diagnosis of Kimura's disease by lymphadenopathy, eosinophilia, elevated IgE levels, and most of all, histopathological findings of the skin and lymph node.

REFERENCES