Clinical Case Report

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Kimura disease of buccal region in a pediatric patient with nephrotic syndrome

A case report

Dong Hoon Lee, MD[a], Ga-Eon Kim, MD[b], Eunmi Yang, MD[c], Tae Mi Yoon, MD[a], Joon Kyoo Lee, MD[a,*], Sang Chul Lim, MD[a]

Abstract

Rationale: Kimura disease is a rare benign, chronic inflammatory disorder that typically presents with slowly enlarging, nontender, subcutaneous swellings in the head and neck region. The occurrence of Kimura disease in the oral cavity is extremely rare.

Patient concerns: A 16-year-old boy presented with a complaint of a right painless buccal mass of 3 years' duration.

Diagnosis: The patient had been diagnosed with nephrotic syndrome and treated with corticosteroid at the age of 5 years.

Outcomes: We report an extremely rare case of Kimura disease of the buccal region in a 16-year-old boy with nephrotic syndrome.

Lesson: We controlled Kimura disease and nephrotic syndrome in this patient by using a combination of surgical resection of the buccal mass and systemic steroid therapy.

Abbreviations: CT = computed tomography, MRI = magnetic resonance imaging.

Keywords: buccal, Kimura disease, nephrotic syndrome, pediatric, surgery

KEY POINTS

Kimura disease is a rare benign, chronic inflammatory disorder that typically presents with slowly enlarging, nontender, subcutaneous swellings in the head and neck region.

We report an extremely rare case of Kimura disease of the buccal region in a pediatric patient with nephrotic syndrome.

We controlled Kimura disease and nephrotic syndrome in this patient by using a combination of surgical resection of the buccal mass and systemic steroid therapy.

1. Introduction

Kimura disease is a rare benign, chronic inflammatory disorder that typically presents with slowly enlarging, nontender, subcutaneous swellings in the head and neck region.[1–7] The majority of these lesions occur in the parotid glands, submandibular glands, or lymph nodes of the neck.[1,4] There are only a few reports of Kimura disease in the oral cavity.[1,4,7] Herein, we report an extremely rare case of Kimura disease in the buccal region in a 16-year boy with nephrotic syndrome.

2. Case report

A 16-year-old boy presented with a complaint of a right buccal mass of 3 years' duration. The buccal mass was painless, but it had slowly increased in size. There was no previous infection, trauma, or contributory medical history, such as allergic disease. The patient had been diagnosed with nephrotic syndrome and treated with corticosteroid at the age of 5 years.

Upon physical examination, a firm, 3.0 cm sized mass was palpated in the right buccal region. Neck computed tomography (CT) scans demonstrated a 2.2 × 3 cm sized, ill-defined, nodular enhancing lesion with perilesional soft-tissue infiltration in the right buccal space (Fig. 1). The lesion had increased in size compared to that on the previous CT scan performed 3 years ago (Fig. 1). Fine needle aspiration cytology of the right buccal mass was performed; however, we experienced failure to achieve an adequate cell harvest because of the hardness of the mass.

For ensuring adequate diagnosis of the right buccal mass, we performed right buccal mass removal through an intraoral approach under general anesthesia. Intraoperative exploration revealed a hard tumor which was attached to the surrounding tissues. The mass was excised completely without any damage to the surrounding structures. Histological examination of the specimen revealed Kimura disease (Fig. 2). Following the

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[a] Department of Otolaryngology-Head and Neck Surgery, [b] Department of Pathology, [c] Department of Pediatrics, Chonnam National University Medical School and Chonnam National University Hwasun Hospital, Hwasun, South Korea.

[*] Correspondence: Joon Kyoo Lee, Department of Otolaryngology-Head and Neck Surgery, Chonnam National University Medical School and Chonnam National University Hwasun Hospital, 160 Ilsimri, Hwasun, Jeonnam 519-809, South Korea (e-mail: joonkyoo@jnu.ac.kr).

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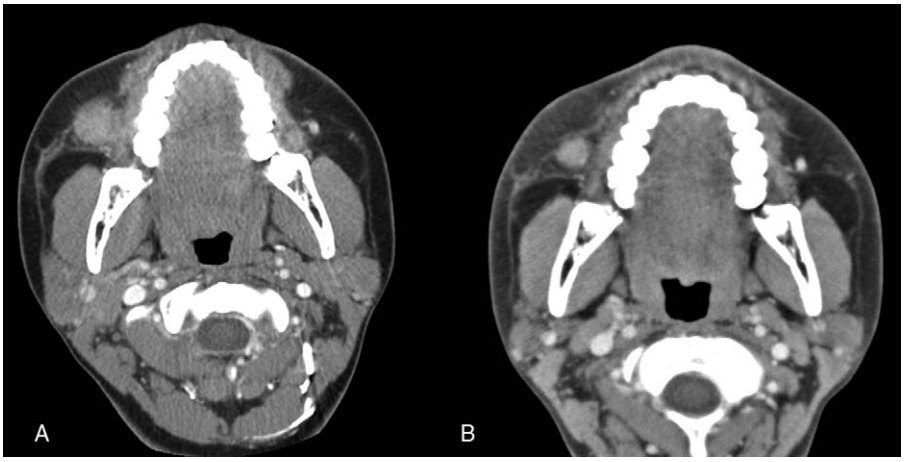

Figure 1. (A) Neck computed tomography (CT) scan demonstrates a 2.2 × 3 cm sized, ill-defined, nodular enhancing lesion with perilesional soft-tissue infiltration in the right buccal space. (B) Three years ago, a CT scan had demonstrated a 1.3 cm sized enhancing mass in the same region.

diagnosis of Kimura disease, the patient has been treated in the pediatric department using corticosteroid to control Kimura disease and nephrotic syndrome. There has been no evidence of recurrence during the 1-year follow-up.

3. Discussion

Kimura disease is a rare, chronic inflammatory disorder of unknown etiology.[1–7] It usually presents in young Asian men with painless soft-tissue masses in the head and neck region.[1–7] However, the occurrence of Kimura disease in the oral cavity, as seen in this patient, is extremely rare.[1,4,7]

There are some reports of patients who have Kimura disease accompanied by nephrotic syndrome.[2–4,6,8–10] The incidence of Kimura disease with coexisting renal disease ranges from 10% to 60%, with two-thirds of patients presenting with nephrotic syndrome.[2,6] However, the pathogenesis of Kimura disease and nephrotic syndrome is unknown.[2–4] Some authors have explained that renal impairment may be probably due to immunocomplex-mediated damage, such as interleukin, cytokines, or T-helper immune response.[3,9,10] In addition, renal symptoms can develop both before and after symptoms of Kimura disease.[3] In this case, the buccal mass appeared after the diagnosis of nephrotic syndrome was made 8 years ago.

The diagnosis of Kimura disease is confirmed by surgical biopsy, which is characterized by angiolymphoid hyperplasia with eosinophilic infiltration.[2,6] Radiologic examinations, such as ultrasound, CT, and magnetic resonance imaging, are helpful to identify the extent and progression of disease, and to provide access to surgical management.[2] In this case, we performed a CT scan several times during 3 years. However, we could not think of the possibility of Kimura disease before surgery. The differential diagnosis of Kimura disease includes Hodgkin disease, lymphoma, leukemia, eosinophilic granuloma, and angiolymphoid hyperplasia with eosinophilia.[2,5]

The optimal management strategy for Kimura disease has not yet been established.[1–9] Treatment of Kimura disease includes surgical excision, regional or systemic steroid therapy, and radiotherapy.[1–9] Surgical excision is considered the 1st-choice treatment for Kimura disease, but relapses are frequent.[2,4] The role of surgical excision in controlling renal manifestations in patients with Kimura disease has not yet been proved.[9] Corticosteroids have been shown to reduce the size of the lesion, but the disease tends to recur after withdrawal of the medicine. Recently, an immunosuppressant agent, cyclosporine, has been reported to be effective in achieving disease remission in patients with Kimura disease.[4,5,7] Radiotherapy has been used to treat recurrent or persistent lesions.[4,5] In this case, we performed complete surgical excision of the lesion. Then, we controlled both

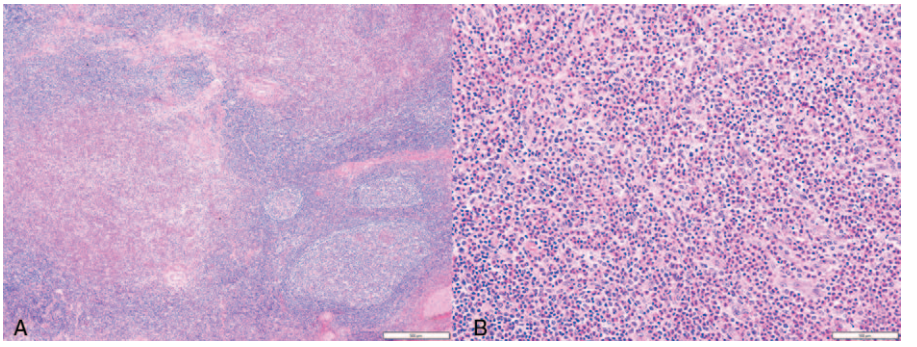

Figure 2. Histopathological images. (A) Follicular hyperplasia with reactive germinal centers and intense eosinophilic infiltration with eosinophilic microabscesses are present (H&E stain, ×40). (B) Predominant eosinophils and proliferation of thin-walled capillaries are present (H&E stain, ×200).

Kimura disease and nephrotic syndrome with corticosteroids. Malignant transformation of Kimura disease does not occur; however, recurrence of the disease frequently occurs in 25% to 40% of patients after surgical, radiation, or steroid therapy.[5,7]

In conclusion, we report an extremely rare case of Kimura disease of the buccal region in a pediatric patient with nephrotic syndrome. In this case, the nephrotic syndrome predated the symptoms of Kimura disease by 8 years. We controlled Kimura disease and nephrotic syndrome in this patient by using a combination of surgical resection of the buccal mass and systemic steroid therapy.

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