Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology 27 (2015) 287–290

Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmp

Case Report

Multiple nontender subcutaneous masses in the head and neck region: Kimura's disease

Wan-Ting Yang [1], Kuo-Yang Tsai *

Division of Oral and Maxillofacial Surgery, Changhua Christian Hospital, No. 235 Xuguang Road, Changhua City 500, Taiwan

ARTICLE INFO

Article history:
Received 25 November 2013
Received in revised form 3 March 2014
Accepted 19 March 2014
Available online 26 April 2014

Keywords:
Kimura's disease
Head and neck mass
Eosinophilia
Elevated IgE
Eosinophilic microabscess

ABSTRACT

Kimura's disease is a rare, chronic inflammatory disorder of unknown etiology, which is seen predominantly in East Asian male patients in the third decade of life. It is often presented as a clinical triad of slow-growing painless mass in the head and neck region with elevated IgE level and eosinophilia. We present such a case in a 32-year-old Taiwanese male, whose symptoms subsided with a course of high dose corticosteroids. As the etiology of Kimura's disease is unknown, the treatment varies from immunosuppressants to surgery.

1. Introduction

Kimura's disease is a chronic and benign inflammatory condition that affects mainly East Asian males. It is characterized by a triad of clinical signs, consisting of nontender subcutaneous masses in the head and neck region, elevated IgE level, and eosinophilia. It was first described in China by Kim and Szeto [1–4] in 1937 and then widely recognized when reported by Kimura et al. [1,2,4–7] in 1948. Kimura's disease was initially regarded as an advanced form of angiolymphoid hyperplasia with eosinophilia (AHLE), but Rosai et al. [2,8] recognized it as histopathologically different. We present such a case in our study, and focus on treatment modalities in our discussion.

2. Case report

Our patient is a 33-year-old single male, a mechanic for excavators, who initially visited our emergency department on January 31, 2011, and again at our oral and maxillofacial surgery outpatient department (OPD) on May 20, 2011, for management of pain associated with impacted bilateral lower third molars.

During physical examination, we noted subcutaneous masses over bilateral buccal and left submandibular region, swelling over bilateral epitrochlear region with overlying multiple erythematous papules, each about 2 mm along the largest diameter (Fig. 1), and a linear scar over right epitrochlear region, about 5 cm in length. The patient denied pain and rash over these swellings, but only had complaints of local heat.

When asked about his past medical history, the patient stated that he noted swelling over both arms since 2 years ago, followed by a swelling over his right face a year later. Hence, he underwent biopsy over right arm at Guang Tien Hospital on June 2010, but lost his copy of the pathology report. However, he remembered being diagnosed with eosinophilia and being admitted for treatment, where facial swelling subsided upon discharge. However, it was not long before his face swelled up again. The patient denied any past medical history or having any allergies, but travel history showed that he had been to Korea in 2009, and to Thailand in March 2010.

We arranged computed tomography (CT) scan and blood works, but the patient lost follow-up until November 9, 2011. By then, he had already undergone an incomplete course of antibiotics and corticosteroid therapy at a different hospital, where he was discharged against medical advice, due to lack of clinical improvement. Thus, he was admitted the next day in our hospital for further evaluation and management of facial swelling.

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

* Corresponding author. Tel.: +886 933 127 916.
E-mail addresses: wtyang24@gmail.com (W.-T. Yang), 72837@cch.org.tw (K.-Y. Tsai).
[1] Tel.: +886 911 855 324.

http://dx.doi.org/10.1016/j.ajoms.2014.03.007
2212-5558/

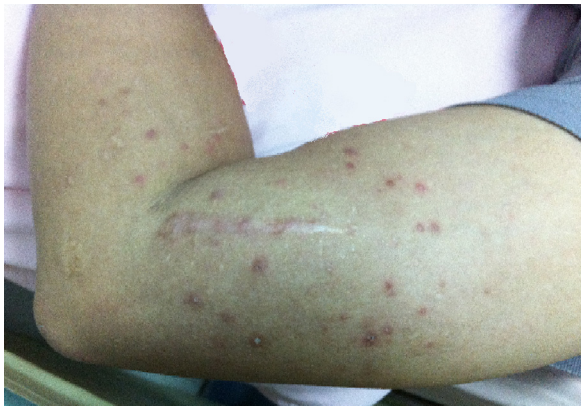

Fig. 1. Photograph of the patient's right arm, exhibiting mild swelling, multiple erythematous papules about 2 mm in along the largest diameter, and an old linear scar about 5 cm in length in the right epitrochlear region.

We suspected the nature of the head and neck lesions to be either of the following: oncologic, infectious, or autoimmune. Thus, we consulted all three departments for further evaluation, and significant findings are summarized in Tables 1 and 2. Head and nasopharynx CT scan, taken on May 30, 2011, showed a heterogeneous mass over right buccal region on the contrast enhanced image, about 3 cm × 2 cm in size (marked with an arrow in Fig. 2). A histopathological slide of the forearm skin specimen, enlarged at a magnification of 400×, showed eosinophilic infiltration (Fig. 3). Thus, according to the clinical triad of subcutaneous mass in the head and neck region with eosinophilia and elevated IgE level, the patient was diagnosed with Kimura's disease. He then underwent 4 days of corticosteroid therapy, and when the lesions subsided, he was discharged under stable conditions on November 20, 2011.

The patient's swelling was controlled at our OPD with oral prednisolone and levocetirizine, though their doses were adjusted frequently to avoid recurrence and Cushing's syndrome. The final regimen consists of 10 mg of prednisolone twice daily, 10 mg of levocetirizine once daily, and 50 mg of azathioprine once daily. His latest laboratory results on March 11, 2013 showed IgE level of 3020 IU/mL, and eosinophil count of 5.4%.

3. Discussion

Kimura's disease is a rare, chronic, inflammatory, and benign condition of unknown etiology characterized by a clinical triad of subcutaneous swellings in the head and neck region with eosinophilia and elevated IgE levels [1,2,4–6,9]. It was first described in Chinese literature in 1937 by Kim and Szeto [1–4], but was named after Kimura [1,2,4–7], who published his study in 1948. In the West, it was first reported by Wells and Whimster. However, they described it as an advanced form of angiolymphoid hyperplasia with eosinophilia (AHLE) [2,9]. It was later in 1982 that Rosai et al. recognized Kimura's disease as a histopathologically different condition [2,8].

Table 2
Significant laboratory findings.

Date	Study type	Value
June 3, 2011	White blood cell (WBC) count	$19.8 \times 10^3/\mu L$
	Eosinophil	66.1%
November 10, 2011	WBC count	$9.9 \times 10^3/\mu L$
	Eosinophil	31%
	Erythrocyte sedimentation rate (ESR)	35 mm/h
November 11, 2011	Urine red blood cells (RBC)	4.4/HPF, 24/μL
November 12, 2011	Eosinophil count	3700/μL
	IgE	>10,000.0 IU/mL
November 20, 2011	WBC count	$12.599 \times 10^3/\mu L$
	Eosinophil count	600/μL
	IgE	>10,000.0 IU/mL
	ESR	25 mm/h
	Urine RBC	3.3/HPF, 18/μL

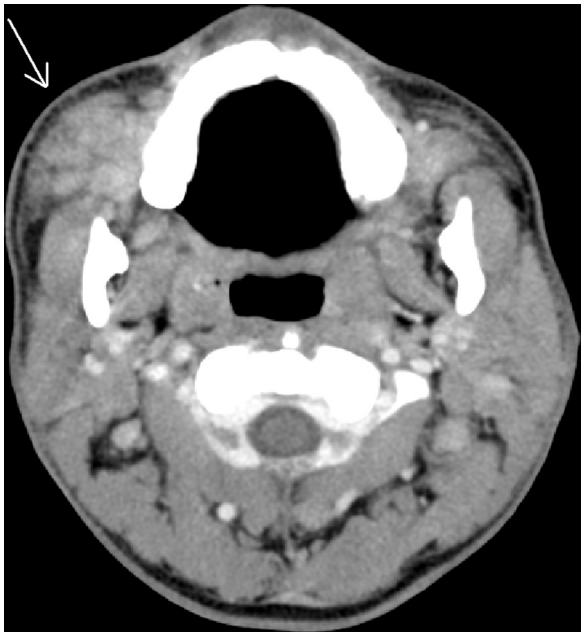

Fig. 2. A heterogeneous mass over right buccal region noted on a contrast enhanced CT scan, measuring about 3 cm × 2 cm in size, marked with an arrow.

Kimura's disease is a Th2-mediated disorder, in which there is a clonal proliferation of T cells [2,9], and an overexpression of interleukin (IL)-4, -5, and -13 [2,6,10,11]. IL-4 and -13 initiate and enhance IgE synthesis, while IL-5 enhances activation, proliferation, and differentiation of eosinophils. Thus, this translates into eosinophilia and elevated IgE levels clinically, and explains the atopic nature of the condition. The trigger for this

Table 1
Significant radiographic and histopathologic findings.

Date	Study type	Findings
May 20, 2011	Head and nasopharynx CT	Right facial swelling compatible with deep neck infection
November 12, 2011	Cytogenetics study	Only chromosomally normal metaphase cells are present
November 17, 2011	Biopsy of skin of forearm	Chronic inflammation with increased eosinophil infiltration
November 18, 2011	Head and nasopharynx MRI	Bilateral facial swelling and multiple bilateral cervical lymphadenopathy may be due to inflammatory process, systemic disease or lymphoma
November 23, 2011	Bone marrow biopsy	The bone marrow shows normal cellularity with decreased M/E ratio. Immunohistochemical stain reveals MPO(+), CD117(+), CD34(−), CD1a(−), CD3(−) and CD20 focal and scattered(+). Increased eosinophil and mast cells are noted