Case Report

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Orbital Involvement in Kimura's Disease Presenting as Diffuse Bilateral Extraocular Muscle Enlargement: A Case Report

미만성 양측성 외안근 비대의 안와침범소견을 보인 기무라병: 증례 보고

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Kimura's disease (KD) is a rare, chronic inflammatory disorder characterized by angiolymphoid hyperplasia, peripheral eosinophilia, and elevated serum immunoglobulin E levels. It primarily affects young Asian males and typically involves the head and neck region, especially near the mandible and postauricular regions. Orbital involvement is unusual and extraocular muscle (EOM) involvement is exceedingly rare, with only a few cases reported in the literature. The present report describes a case of surgically confirmed KD in a 16-year-old male, involving the bilateral EOM, lacrimal gland, and left parotid gland.

Index terms Kimura's Disease; Exophthalmos; Extraocular Muscles Enlargement; Bilateral Lacrimal Gland Swelling

INTRODUCTION

Kimura's disease (KD) is an uncommon chronic inflammatory disorder characterized by subcutaneous mass-like lesions, predominantly located in the head and neck region. Lesions are frequently found in the parotid and submandibular areas, often accompanied by lymphadenopathy. Less prevalent locations in the head and neck include the paranasal sinuses, orbits (including the eyelids, conjunctiva, and lac-

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rimal glands), epiglottis, tympanic membrane, and parapharyngeal space (1). Orbital involvement is infrequent and typically affects the eyelid or lacrimal gland. We present a rare case of KD involving the bilateral extraocular muscles (EOMs) and lacrimal gland, with detailed imaging findings.

CASE REPORT

A 16-year-old male patient visited the outpatient clinic with complaints of bilateral proptosis and a palpable mass in the left postauricular area which have presented for over 3 years. The patient stated that the mass gradually increased in size. The patient had no underlying medical conditions and denied any other history of previous disease or family history of disease.

Exophthalmometry measurements were 21 mm for the right eye and 22 mm for the left eye. In laboratory testing, thyroid function was normal and therefore thyroid-associated orbitopathy was ruled out. Autoimmune-related laboratory tests were conducted to detect immunoglobulin G (IgG)-related sclerosing disease and Sjögren's syndrome, yielding negative results for IgG4, anti-Ro, La, and Jo-1 antibodies. However, there was a notable elevation in serum IgE levels, reaching 1457 IU/mL (normal range, ≤378 IU/mL), accompanied by peripheral eosinophilia (29%).

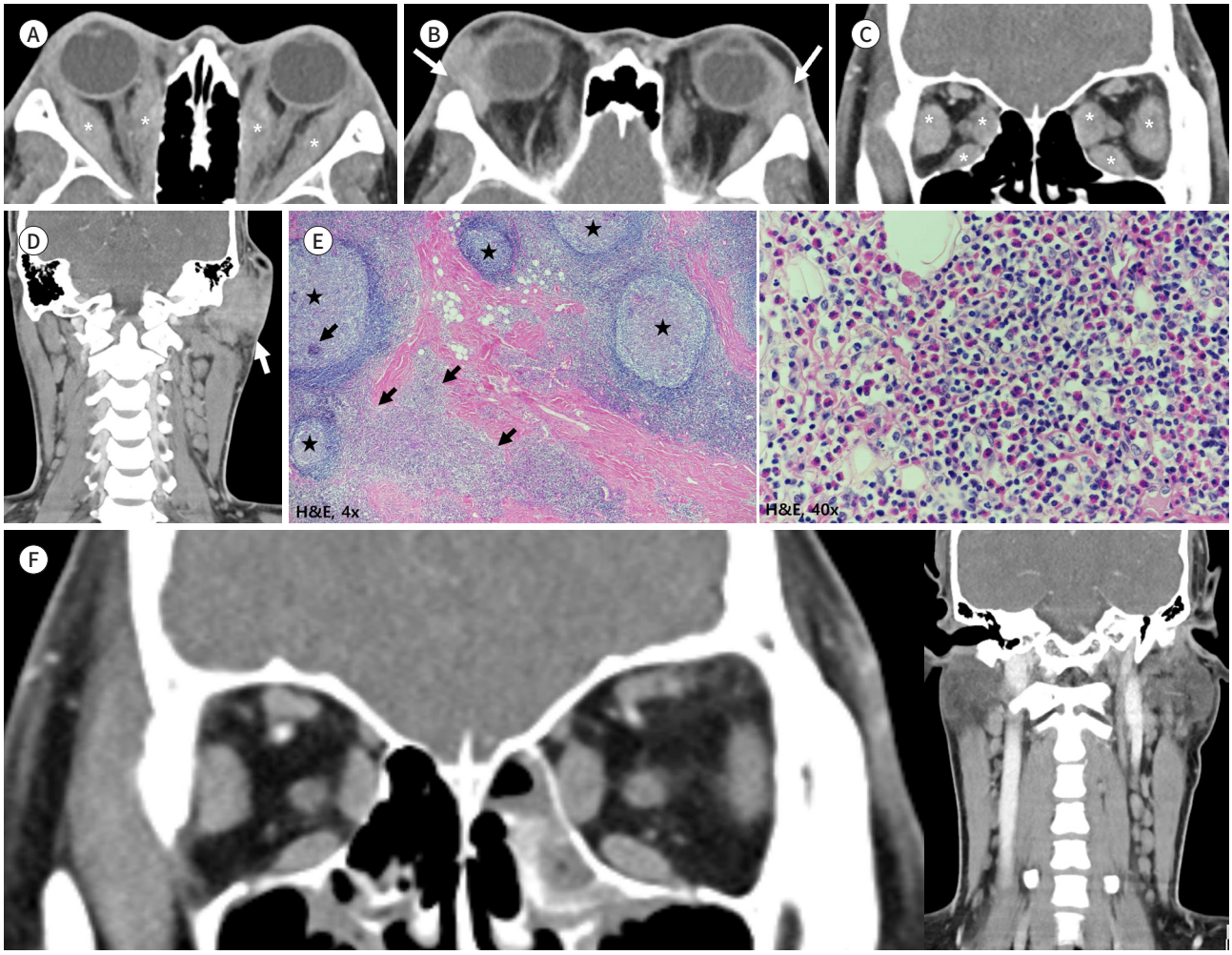
Contrast-enhanced neck CT demonstrated diffuse fusiform enlargement of the lateral, medial, and inferior rectus muscles bilaterally including the anterior tendon; both lacrimal glands were symmetrically enlarged (Fig. 1A-C). Diffuse bilateral preseptal and postseptal edema and mild fat infiltration were also observed. Additionally, there was a homogeneous mass-like enhancement with ill-defined margins measuring up to 5 cm, located in the superficial lobe of the left parotid gland and extending into the surrounding subcutaneous fat layer (Fig. 1D). There were multiple small lymph nodes with homogeneous enhancement measuring up to 1 cm in long axis diameter, with mild perinodal infiltration. These were predominantly observed in the periparotid area and left cervical levels I–V (Fig. 1D).

These findings, including diffuse symmetric enlargement of bilateral EOM and lacrimal glands, a left parotid mass associated with regional lymphadenopathy, and peripheral eosinophilia in a young Asian male, suggested KD. Subsequently, the patient underwent excisional biopsy of the left parotid mass and left level II lymph nodes. Microscopic examination showed reactive lymphoid hyperplasia and significant eosinophilic infiltration, forming an eosinophilic microabscess, leading to a diagnosis of KD (Fig. 1E).

Steroid treatment was administered for 2 weeks. Blood tests revealed an eosinophil count of 0.2% after the completion of steroid treatment. A follow-up neck CT performed after 2 months demonstrated decreases in the sizes of the enlarged EOM and lacrimal glands on both sides, with no detectable residual mass in the parotid gland after superficial partial parotidectomy (Fig. 1F). The previously observed left-sided regional cervical lymphadenopathy also improved. Follow-up to date showed no recurrence of symptoms and signs.

This study was approved by the Institutional Review Board, which waived the requirement for informed patient consent (IRB No. 2023-10-015).

Fig. 1. Kimura's disease in a 16-year-old male presenting with bilateral exophthalmos and a painless left postauricular palpable mass.
A-C. Axial (A, B) and coronal (C) images of contrast-enhanced neck CT show bilateral fusiform enlargement of both lateral, medial, and inferior rectus muscles (*) and diffuse symmetrically swollen bilateral lacrimal glands (arrows).
D. A coronal image shows an ill-defined, homogenously enhancing infiltrative mass in the left parotid gland (arrow), extending to the subcutaneous layer of the periauricular area. Regional reactive lymphadenopathy is observed in left cervical level IB, II–V.
E. Histopathological examination of the left parotid mass demonstrates the aggregation of hyperplastic follicles with germinal centers (left, *), eosinophilic microabscess formation, and prominent eosinophilic infiltration (left, arrows).
F. Coronal images of contrast-enhanced CT after steroid treatment show improved diffuse enlargement of both lateral, medial, and inferior rectus muscles in both orbits. Regional lymphadenopathy in left cervical levels II–V and left parotid and periauricular soft tissue lesions also improved.
H&E = hematoxylin and eosin



DISCUSSION

KD is a chronic inflammatory disorder characterized by the proliferation of folliculoid structures with the infiltration of eosinophils, plasma cells, lymphocytes, and mast cells (1). KD is often observed in young Asian males, presenting as subcutaneous masses, particularly in the head and neck. It commonly occurs in the parotid and submandibular regions. It can be associated with regional lymphadenopathy (42%–100%) and bilateral involvement is observed in some cases. Less frequent sites include the paranasal sinuses, orbits, epiglottis, tym-

panic membrane, and parapharyngeal space. Rarely, KD extends beyond the head and neck to the axilla, groin, popliteal region, and forearm. Most patients have a prolonged course with slow enlargement of masses; however, the clinical course is generally benign and self-limited (1, 2).

Orbital involvement in KD is relatively rare (3), predominantly affecting the eyelid or lacrimal gland, with superior orbit involvement being more common (4, 5). Patients presenting with orbital involvement exhibit various symptoms, such as exophthalmos, eyelid swelling, palpable mass, ocular dysmotility, ptosis, lacrimation, pruritus, pain, or headache (6). Among cases of KD with EOM involvement, unilateral involvement is mainly reported (5-7), with very few cases of bilateral EOM involvement (2, 4, 5).

The exact cause and pathogenesis of KD remain unclear, although it might be a self-limited allergic or autoimmune response triggered by an unknown persistent antigenic stimulus. Studies have also shown that the proliferation of CD4+ T cells and resultant overproduction of cytokines trigger lymphoid follicle formation and elevated IgE (2). Consequently, clinical manifestations often include peripheral blood eosinophilia (ranging from 10% to 70%) and elevated serum IgE levels (800–35000 IU/mL), which may fluctuate throughout the course of the disease.

It is difficult to diagnose KD when it affects the EOM bilaterally. EOM involvement in KD can manifest as diffuse enlargement and mild homogeneous enhancement, as documented in several reports (2, 4, 5) and observed in our case. Differential diagnoses include other orbital inflammatory diseases, including thyroid-associated orbitopathy, idiopathic orbital myositis, and IgG4-related disease, and certain neoplasms, such as lymphoma and Kaposi's sarcoma (8). Nevertheless, the identification of a subcutaneous component within the salivary mass, along with the presence of lymphadenopathy and laboratory findings (such as peripheral eosinophilia and elevated serum IgE levels), considering ethnicity and sex, assists in the exclusion of other conditions. However, in cases where a salivary mass and peripheral eosinophilia are absent and differentiation from other orbital inflammatory diseases and tumors is required, histopathologic confirmation is ultimately required.

In conclusion, this study represents a rare case of KD with bilateral EOM and lacrimal gland involvement. Although the orbit is an uncommon location, it is crucial to consider KD as a differential diagnosis in patients with slowly progressive orbital inflammatory disease, even in cases of predominant EOM involvement.

Author Contributions

Conceptualization, L.K.J., L.H.Y.; investigation, L.H.Y.; project administration, L.H.Y.; resources, L.H.Y., C.S.J.; supervision, L.H.Y., C.S.J., L.M.K., K.Y.H.; validation, L.H.Y.; writing—original draft, L.K.J., L.H.Y.; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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REFERENCES

1. Park SW, Kim HJ, Sung KJ, Lee JH, Park IS. Kimura disease: CT and MR imaging findings. *AJNR Am J Neuroradiol* 2012;33:784-788
2. Chakraborti C, Saha AK, Bhattacharjee A, Lakra R. Kimura's disease involving bilateral lacrimal glands and extraocular muscles along with ipsilateral face: a unique case report. *Indian J Ophthalmol* 2019;67:2107-2109
3. Agrawal S, Sen S, Sabu S, Modaboyina S, Bajaj MS, Das D. Bilateral orbital Kimura's disease: a case report and brief review of literature. *Indian J Ophthalmol* 2022;70:2737-2738
4. Goncalves AC, Moritz RB, Aldred VL, Monteiro ML. Bilateral extraocular muscles enlargement from Kimura's disease of the orbit. *Indian J Ophthalmol* 2016;64:538-540
5. Dhingra H, Nagpal R, Baliyan A, Alva SR. Kimura disease: case report and brief review of literature. *Med Pharm Rep* 2019;92:195-199
6. Lee SJ, Song JH, Kim SD. Kimura's disease involving the ipsilateral face and extraocular muscles. *Korean J Ophthalmol* 2009;23:219-223
7. Chung DH, Kim BJ, Kim YD. [Kimura's disease involving the eyelid and orbit]. *J Korean Ophthalmol Soc* 2002;43:1789-1796. Korean
8. Rana K, Juniat V, Patel S, Selva D. Extraocular muscle enlargement. *Graefes Arch Clin Exp Ophthalmol* 2022;260:3419-3435

미만성 양측성 외안근 비대의 안와침범소견을 보인 기무라병: 증례 보고

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기무라병은 주로 젊은 동양인 남성에서 발생하는 혈관림프증 증식, 말초 호산구 증가증, 혈청 면역글로불린 E 상승을 특징으로 하는 드문 만성 염증성 질환으로, 대부분 두경부에, 특히 하악과 후이개 주변에 발생한다. 안와에 발생하는 기무라병은 드문데, 특히 외안근 침범으로 나타나는 경우는 매우 드물고 문헌에 소수의 증례 보고만이 있다. 이에 저자들은 양측 외안근과 눈물샘 및 좌측 이하선 종대로 내원한 16세 남자에서 수술로 확진된 기무라병의 영상소견을 기술하고자 한다.

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