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Case Report

# Painless Mass on the Eyelid: Kimura Disease

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**Abstract**

Kimura disease (KD) is a rare chronic inflammatory disease of unknown etiology. It usually presents with regional lymphadenopathy in the head-and-neck region and swelling of the salivary glands and subcutaneous tissues. Periocular involvement in KD is rare. We report a case of KD on the eyelid as a rare cause of eyelid masses.

**Keywords:** Eyelid, kimura disease, painless mass

## Introduction

Kimura disease (KD) is a rare chronic inflammatory disorder of unknown etiology and involves subcutaneous tissue, salivary glands, and the lymph nodes of head-and-neck region (1). Although the pathophysiology of KD is not known exactly, allergic reactions, infections (parasites, viruses, and fungi), microbial toxins, atopy, and autoimmunity are shown as possible causes. The disease is usually diagnosed histopathologically with incisional or excisional biopsy. Here, we present the clinical features, diagnosis, treatment, and follow-up of a patient diagnosed with KD in the lower eyelid.

## Case Report

A 39-year-old Caucasian male patient was admitted to our clinic with a complaint of painless swelling on his right lower eyelid for 1.5 years, which had been growing steadily for the past year. The patient was healthy otherwise, and the history was unremarkable. Visual acuity was 20/20 and intraocular pressure was 15 mmHg bilaterally. There was no relative afferent pupillary defect in either eye and extraocular movements were full bilaterally. The slit-lamp and dilated fundus examination was normal in each eye. The Hertel exophthalmometer measurements were 18 mm and 17 mm in the right eye and left eye, respectively. On external examination, a palpable immobile mass on the right lower eyelid and a purplish discoloration of the overlying skin were detected (Fig. 1). On T1-weighted orbital magnetic resonance imaging (MRI), a 13×25×12 mm lesion localized to the subcutaneous adipose tissue and preseptal area of the right lower eyelid was detected, and it extended to the medial canthus partially (Fig. 2). The lesion had intense contrast enhancement. The patient underwent an excisional biopsy, and the tissue was sent for histopathological evaluation. In histopathological examination, dense lymphoid follicles with a prominent germinal center and inflammation rich in eosinophils were found, and the diagnosis of KD was made (Fig. 3). Subsequently, eosinophilia was observed in the complete blood count examination of the patient, and he was consulted with hematology. Serum IgE level was found to be

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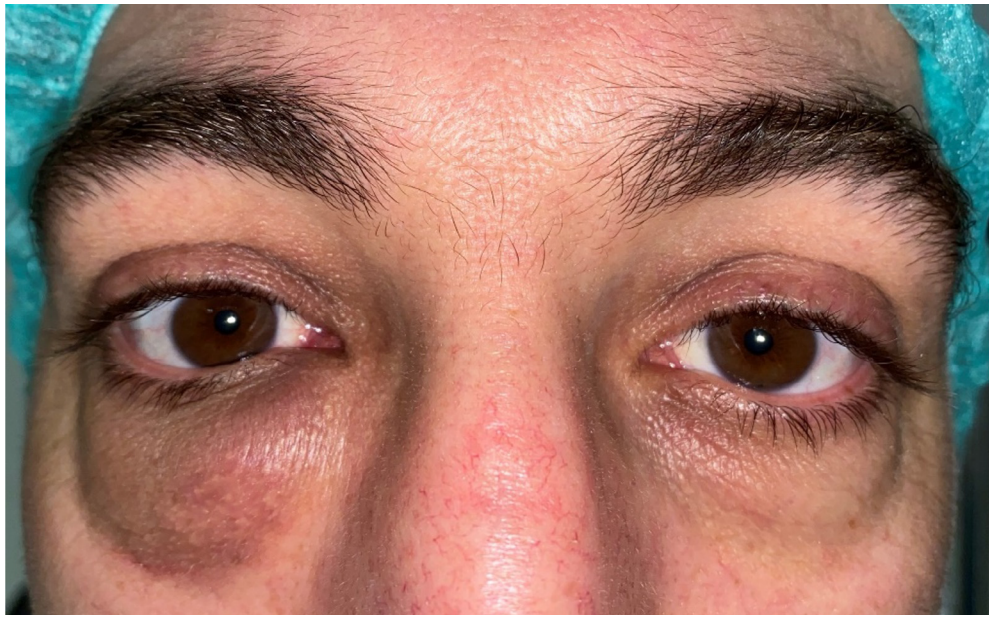

**Figure 1.** Swelling of the right lower eyelid.

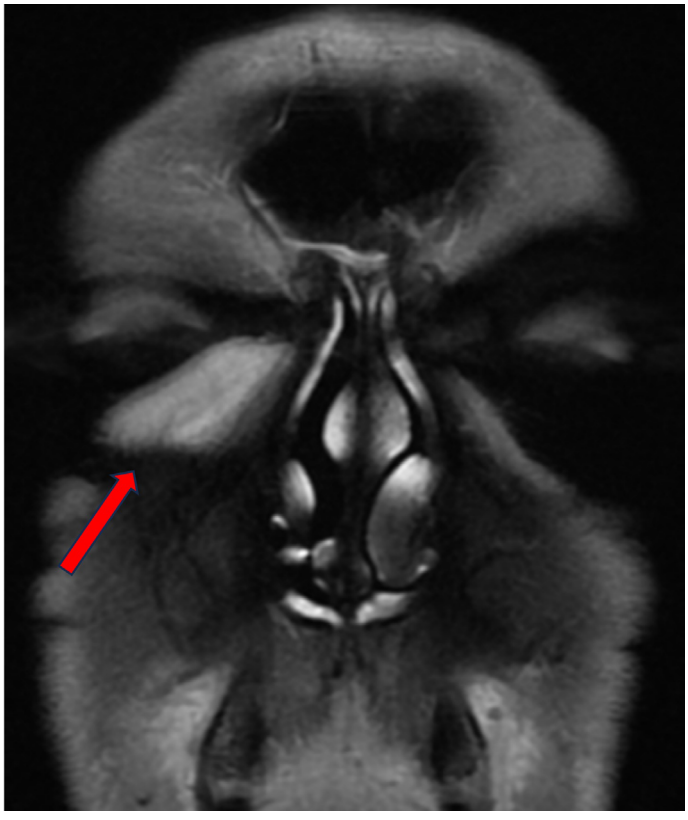

**Figure 2.** Coronal T1-weighted orbital magnetic resonance imaging, a mass of 13×25×12 mm localized to the subcutaneous adipose tissue and preseptal area of the right lower eyelid.

normal. The patient was followed up by the hematology department, and no immunosuppressive treatment was given. The patients were followed up for 24 months, and no recurrence was observed (Fig. 4).

## Discussion

Kimura disease is a rare chronic inflammatory disease. It was first contributed to the Chinese literature as "eosinophilic hyperplastic lymphogranuloma" by Kim and Szeto in 1937 and then introduced to the Japanese literature as Kimura's disease by Kimura in 1948 (2). It is an endemic disease for middle-aged Asians but sporadically detected in non-Asian ethnic groups. Although KD can be seen in all age groups, it is most commonly seen in the second or third decades. Men are more frequently affected than women (3:1). It often occurs as subcutaneous nodules in the head-and-neck region. Nodules are usually painless and larger than 3 cm and occur in the retroauricular, submandibular, mastoid, and cervical regions, and rarely in the inguinal, axillary region, and orbit. Orbital and periocular involvement is rare and usually affects the superior orbit, eyelid or lacrimal gland (3,4). Although the exact etiopathogenesis of KD is not known, it is suggested that the disease arises due to an allergic or autoimmune response. The coexistence of renal disease, which occurs most frequently with membranous glomerulonephritis and mesangial proliferative glomerulonephritis, is seen in 10%–60% of patients.

The diagnosis is made histopathologically and is characterized by lymphoid follicles with germinal centers distinguished by eosinophilic infiltrate and varying degrees of fibrosis. The clinical course is generally benign and self-limited.

Recently, Han et al. described a case of KD affecting the lacrimal gland, the ipsilateral orbita, and the contralateral earlobe (4). They also reviewed the literature and identified 37 cases of periorbital KD. The average age of patients was 39.1 years, and 81.1% of them were male. Bilateral involvement was only seen in 27% of the patients. Notably, 46% of cases of perior-

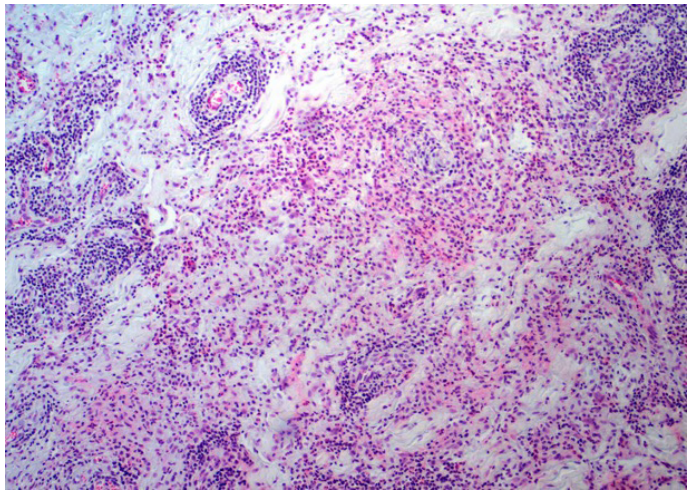

**Figure 3.** H&E×100-mixed inflammation with lymphoid follicles and scattered eosinophils.

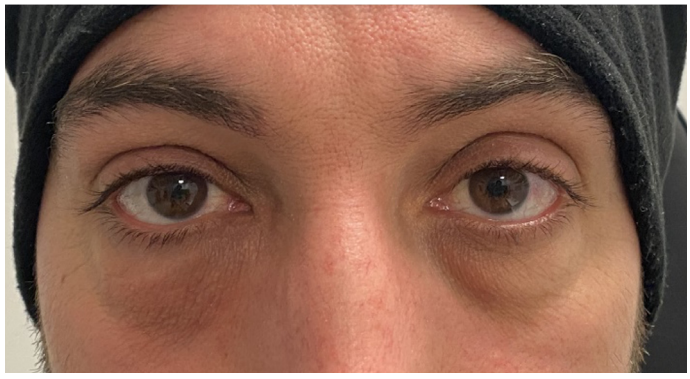

**Figure 4.** Follow-up at 24 months after surgical excision.

bital KD had additional extraorbital lesions. In our case, the lesion was only localized to the eyelid, and there was no orbital or systemic involvement. Orbital involvement is uncommon, and when it occurs, it typically affects the lacrimal gland, rarely involving the extraocular muscles. Orbital involvement of KD may be associated with decreased visual acuity, diplopia, limited eye movement, visual field defect, blepharoptosis, exophthalmos, and relative afferent pupillary defect. Goncalves et al. presented a case with bilateral, slowly progressive proptosis that was initially misdiagnosed as Graves' Ophthalmopathy (5). Following worsening of proptosis and the development of facial and temporal swelling, an MRI revealed enlargement of all recti muscles and diffuse orbital infiltration. Orbital biopsy was consistent with KD. Byeon et al. presented a case with bilateral epiphora and enlarged swelling of both inner canthi (6). Orbital MRI revealed contrast-enhanced soft tissues in both medial canthi, lacrimal sac and along nasolacrimal ducts. Histopathologic examination confirmed KD after incisional biopsy. This is a unique presentation of KD with invasion in the lacrimal sac and nasolacrimal duct.

Surgical excision is the most common treatment modality for ophthalmic KD. Buggage et al. suggested that surgical excision prevents recurrence (3). In the literature, while 80% of patients who initially received corticosteroids experienced a recurrence, only 20% of those who initially underwent surgery had a recurrence (4). In general, eosinophilia and an increase in peripheral blood IgE levels are seen as risk factors for recurrence. In our case, the mass was only localized to the eyelid clinically, and orbital involvement was not found in the radiological evaluation. In addition, serum IgE level was found to be normal. Therefore, only surgical excision was helpful without the need for additional medical treatment. Since there is anecdotal data, radiation therapy and steroid-sparing agents such as cyclosporine A, mycophenolate mofetil, and tacrolimus should be reserved for complex cases.

## Conclusion

KD is a rare chronic inflammatory condition that should be considered in the differential diagnosis of painless eyelid masses. The increasing number of ophthalmic KD patients will allow us to understand better the relationship between systemic diseases, underlying risk factors, treatment, and longer-term follow-up results.

### Disclosures

**Informed consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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