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Co-occurrence of Kimura's Disease and Keratoconus: A Case Report

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Abstract:
Kimura disease, often referred to as eosinophilic granuloma, is an uncommon and long-lasting condition that is mostly seen in people of Asian ancestry. Early signs include increased levels of IgE in the blood, nodules under the skin, and swelling of the lymph nodes. Keratoconus is a disorder where both corneas get steeper and vision is reduced. It is not genetically connected to Kimura disease, although both illnesses are more common among Asian people. Performing a histopathological investigation of the mass is essential in order to get an accurate diagnosis. This case reports a 24-year-old male patient living in Saudi Arabia who had a growth on his face that extended above and below the medial canthus of his eye. The patient also had a gradual decrease in vision in both eyes. The patient received a comprehensive biopsy procedure and topographical examinations. The biopsy findings indicated the presence of angiolymphoid hyperplasia with eosinophilia, and no accompanying lymphadenopathy was seen. The diagnosis of Keratoconus was verified by the examination of the topography. Considering these results, the main therapeutic option is surgical removal of the tumor. This case report offers valuable insights into the diagnostic criteria, differential diagnosis, and management of Kimura disease, specifically in relation to keratoconus. Additionally, it contributes to the existing literature by presenting the first documented case of Kimura disease with concurrent keratoconus.

Keywords: Kimura disease, Eosinophilic granuloma, Eyelid mass, Unilateral Keratoconus, lymphatic vessels proliferation.

Introduction

Kimura Disease (KD) is a chronic inflammatory condition characterized by abnormal proliferation of blood and lymphatic vessels [1]. It predominantly manifests in the head and neck area of Asian males during their second to fourth decades of life. Although most cases are observed in Asia, sporadic occurrences have been reported in Europe and America [2-3]. This disorder is often accompanied by nephrotic syndrome and is classified as a rare and persistent inflammatory ailment with an uncertain underlying cause. The male-to-female prevalence ratio ranges between
3.5:1 and 9:1 [4]. KD was initially described in a Chinese study in 1937 [5], initially referred to as "eosinophilic proliferative lymphogranuloma." In 1948, Kimura in Japan provided the definitive histological description, leading to the disease being named after him [6].

During clinical examination, painless swelling or nodules beneath the skin in the head and neck area are typically observed. These manifestations are often accompanied by regional lymphadenopathy. Diagnosing this disease can be challenging due to its similarity to both benign and malignant conditions, as well as its low incidence [7].

The exact cause of KD remains unknown, but it is speculated that it may be associated with impairment or disruption of immune regulation. One hypothesis proposes that KD could be triggered by an atopic reaction to a persistent antigenic stimulus, such as arthropod bites, viral infections, or neoplasms. A particularly

intriguing theory suggests that Candida could act as a potential source of persistent antigenemia, although no hyphae or spores have been isolated thus far. The disease is characterized by excessive growth of lymphoid follicles and vascular endothelium. The presence of peripheral eosinophilia and eosinophils within the inflammatory infiltrate indicates that KD might involve a form of hypersensitivity reaction. It is speculated that T-helper 2 (Th2) lymphocytes may play a role in this process [8].

To the best of our knowledge, there is no reported association between Kimura disease and keratoconus in the existing literature. Therefore, this case report presents a unique and noteworthy occurrence as it is the first documented case of Kimura disease and keratoconus coexisting in a patient. This finding highlights the importance of further exploration and investigation into the potential relationship between these two conditions.

Materials And Methods

Case Presentation:

During the previous two years, a male soldier, aged 24, had gradual vision loss and a mass in the medial region of his left lid. The evaluation revealed that the patient's left eye's visual acuity was lower (20/100) than his right eye's (20/40). There was no afferent pupillary impairment, and all extraocular motions and pupil reactions were present. Both sides displayed floppy-eyelid syndrome and giant papillary responses. On the left upper lid, the 5x2 cm extension above and below the medial canthal tendon was solid, painless, and immobile. Munson's sign was positive in the left eye, and both eyes' corneas were clear.

Initial Screening:

The anterior chamber examination revealed quiet, deep chambers, while the lens and iris were within normal ranges. The dilated fundus examination revealed an excellently shaped optic disc and a normal, flat retina. There was nothing palpable in the lymph nodes. With the possible exception of an increased eosinophil count, laboratory tests exhibited a total blood count within normal ranges. Screening for tuberculosis (PPD) came out negative. There was also evidence of inflammation, as the C-reactive protein levels were high (1.34 mg/dl). CMV IgG showed weak positivity. The IgE levels, as well as the tests for liver and kidney function, were all normal. There were no parasites in the blood or stool samples. There were normal IgG4 levels. A chest X-ray revealed no anomalies.

Medical Procedure:

After all the initial screening came negative the patient was examined for lesion removal. Patient was transferred to the operating room, and the lesion was surgically removed under local anesthesia. Biopsy was performed on this lesion for verification.

Results

The medial canthus of the left eye was found to have a diffuse and infiltrative soft tissue disorder, according to CT imaging. A protrusion of the left eye socket was observed. The left frontal sinus was completely occupied, and there was inflammation and swelling of the mucous membrane in both ethmoid sinuses. Figure 1 demonstrate the CT imaging which revealed a diffuse infiltrative soft tissue disorder affecting the inner corners of the eyes on both sides, as well as enlarged lacrimal glands on both sides.

Corneal topography analysis is shown in figure 2. Fig. 2(a) is the topography of right eye which matches the normal corneal topography whereas fig. 2(b) validate corneal ectasia in left eye.

A biopsy of left eyelid lesion is shown in figure 3 as a histological investigation which presented fibrosis that was dividing hyperplastic germinal centers. Evident eosinophilic infiltrates, proliferating blood vessels, and inflammatory cells were seen underneath the subcutaneous tissue.

Discussion:

The fact that Kimura's illness and keratoconus are both present at the same time raises significant concerns about the underlying causes of these two disorders. A rare and chronic inflammatory illness known as

Kimura Disease (KD) was first referred to as "eosinophilic hyperplastic lymphogranuloma" in Chinese literature by Kim and Szeto in 1937. KD is a condition that affects a small percentage of the population. After Kimura et al. published a full description of the condition in 1948 in Japanese literature, it eventually became known as Kimura's disease and earned global notoriety because of this.

Kimura's illness is thought to be caused by an allergic or autoimmune reaction that is self-limiting and is initiated by an undiscovered persistent antigenic stimulation. However, the precise processes that underlie this disease are yet unknown. The current view is that Kimura's disease (KD) is related to either an autoimmune response or a delayed hypersensitivity reaction. This is despite the fact that various recommendations have been made about possible viral origins. According to the association between the existence of an aberrant allergic response and illnesses such as asthma, allergic rhinitis, conjunctivitis, atopic dermatitis, peripheral hypereosinophilia, and high levels of serum IgE, the presence of an abnormal allergic response is further supported[9-10]. The actual etiology of keratoconus is not yet completely recognized by medical professionals. There are a number of hypotheses that have been put forth by various researchers. These hypotheses include developmental alterations in corneal growth, degradation of elastic fibers inside the cornea, and the possibility of illness or nutritional inadequacies. However, the relationship that is noted the most commonly is with the practice of wiping one's eyes repeatedly.The numbers [11-12]. There is presently no published case report that describes the coexistence of keratoconus with Kimura's disease; nevertheless, the relationship of floppy eyelid syndrome has been recorded in two instances [10, 13]. This is despite the fact that multiple cases of Kimura's disease affecting the orbit have been documented in the literature.

In rare cases, Kimura's illness might manifest itself in the eye. The first documented instance of orbital involvement was reported by Nakai et al. [14] in the year 1966. In 2014, Li et al. [15] published a case of a Chinese male person who was 47 years old and had been diagnosed with orbital Kimura's disease (KD). This particular form of the illness impacted the lacrimal gland and had characteristics that were comparable to those of IgG4-related orbitopathy. Furthermore, Goncalves et al. [16] recorded occurrences in 2016 when KD appeared as bilateral extraocular muscle hypertrophy. These examples were described. The same recurring case of KD that was limited to the inner canthus and did not follow lymphadenopathy was reported in another study that was published in 2017 by Francis et al. [17]. In addition, Chakraborti et al. [18] presented discoveries in 2019 about KD that included several orbital structures and lymph nodes in the area. Agarwal et al. [19] recently revealed that Kimura's illness was present in both of the patient's orbits. The many symptoms of KD that might occur inside the orbital area are brought to light by these examples.

The purpose of this case report is to shed light on the exceptional incidence of Kimura's illness in conjunction with keratoconus, which has not been documented in the existing body of research. The diagnosis was verified by the histological examination of the mass, which revealed angiolymphoid hyperplasia with eosinophilia. Additionally, corneal topography indicated the existence of unilateral corneal ectasia. Concerning the underlying causes of these two states and the possible linkages between them, the presence of these two circumstances raises some fascinating concerns. Excision of the tumor using surgical means is often regarded as the main therapeutic option.

Conclusion:

The comprehensive evaluation and subsequent surgical intervention for a 24-year-old male soldier with gradual vision loss and a mass in the medial region of his left lid revealed a complex ocular condition characterized by asymmetrical visual acuity, floppy-eyelid syndrome, and giant papillary responses, ultimately diagnosed as a diffuse and infiltrative soft tissue disorder with corneal ectasia.

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