Kimura's disease involving bilateral lacrimal glands and extraocular muscles along with ipsilateral face: A unique case report

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A 23 year female presented with bilateral recurrent swelling of eyelids along with ptosis and proptosis for last 3 years. She also had swellings over the right cheek, parotid gland, and retro auricular area along with regional lymphadenopathy. Systemic laboratory workup revealed raised serum IgE and a high peripheral eosinophil count. Computed tomography and magnetic resonance imaging showed bilateral enlargement of extraocular muscles, lacrimal glands, and ipsilateral parotid gland. Excision biopsy of the retro-auricular lymph node was suggestive of Kimura's disease (KD). The patient responded well to systemic corticosteroid. KD rarely affects orbit, but it should be included in the differential diagnosis of inflammatory diseases of the orbit. To our knowledge, this is the first reported case of KD from India involving the orbit, lacrimal gland, extraocular muscles, parotid gland and buccal area.

Key words: Bilateral lacrimal gland swelling, eosinophilia, extraocular muscles enlargement, Kimura's disease

Kimura's disease (KD) was first reported in 1937 in China.[1] It primarily affects young Asian male.[2] KD is a rare chronic inflammatory disease of unknown etiology. It is believed to be an immune-mediated disease and T-helper cells (TH2) are suspected to play an important role.[3] The exact prevalence of KD is not known. It is characterized by painless unilateral cervical lymphadenopathy or subcutaneous masses predominantly in head and neck region.[4] The lacrimal glands or the eyelids are usually affected in the orbital involvement of KD. Involvement of unusual sites such as axilla, groin, palate, and epitrochlear region is also reported.[5]

KD is associated with blood and tissue eosinophilia and elevated serum IgE levels.[6] The definitive diagnosis requires surgical excision followed by histopathological examination (HPE).We report a young female patient with KD who presented with bilateral orbital involvement, unilateral parotid gland swelling along with regional lymphadenopathy.

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Case Report

A 23-year-old female presented with swelling and drooping of both upper lids along with bulging of eyes for the last 3 years. She also complained of swelling over the right cheek and retro-auricular area for the last 2 years. The swellings were gradually increasing and were associated with itching and pigmentation of overlying skin. Her medical history was non-contributory. She was treated with oral steroid by a private physician 2 years back without any definitive diagnosis. The swellings partially resolved with medications but started increasing after the discontinuation of therapy.

Her visual acuity was 20/20 in both eyes. There was bilateral swelling of upper lids with blepharoptosis [Fig. 1]. The swellings were non-tender, soft, and nodular in feeling. There was no restriction of ocular movements. Proptosis was 20 mm and 18 mm in RE and LE, respectively. Anterior and posterior segments were within normal limit. Schirmer's test values were 12 mm and 13 mm in right and left eye, respectively. General examination found diffuse non-tender, soft swelling extending over the right cheek, and parotid region [Fig. 2]. There was diffuse brownish pigmentation over the eyelids and right cheek. A localized non-tender swelling (2.5 cm × 2.5 cm × 1 cm) was found at right retro-auricular region [Fig. 3]. There was no other systemic involvement found except right-sided diffuse cervical and submandibular lymphadenopathy.

Systemic laboratory works up revealed Hb 10.15 gm%, TLC 9600/cu mm with eosinophilia (E-30%), ESR 24 mm/h, TSH- 0.89, T3-1.84, T4-11.69, serum IgE- 262.64 (normal 0–200), serum urea, creatinine, and uric acid normal. Chest X-ray and skin tuberculin tests were normal and Kveim test was negative. Ultrasonography whole abdomen was normal. Contrast MRI of orbit revealed bilateral enlargement of lacrimal glands, extraocular muscles with sparing of tendons, diffuse apical orbital mass, swelling of eyelids, and orbicularis muscle, along with swelling of right-sided temporalis muscle [Fig. 4a-c].

Excision biopsy of right retro-auricular lymph node revealed follicular hyperplasia with the proliferation of post-capillary venules and para cortical sclerosis along with plenty of tiny eosinophilic collections/abscess, suggestive of Kimura's disease [Fig. 5a and b]. The patient was treated with oral prednisolone 40 mg once daily for 1 month, then on tapering dose. At 1-month follow-up, there was marked decrease in size of all the swellings along with the disappearance of

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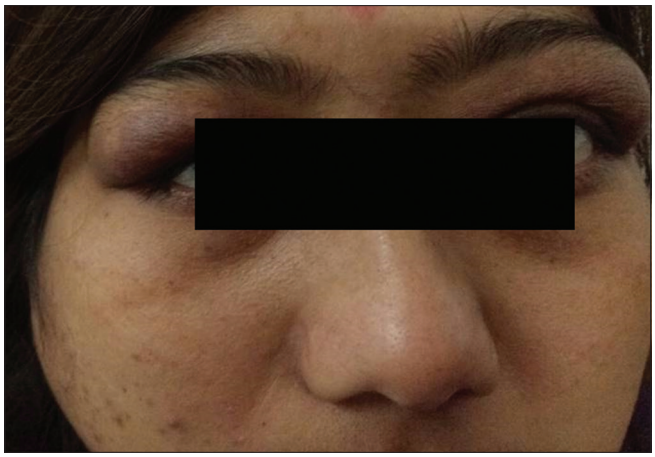

Figure 1: Showing bilateral swelling of the upper lid with blepharoptosis

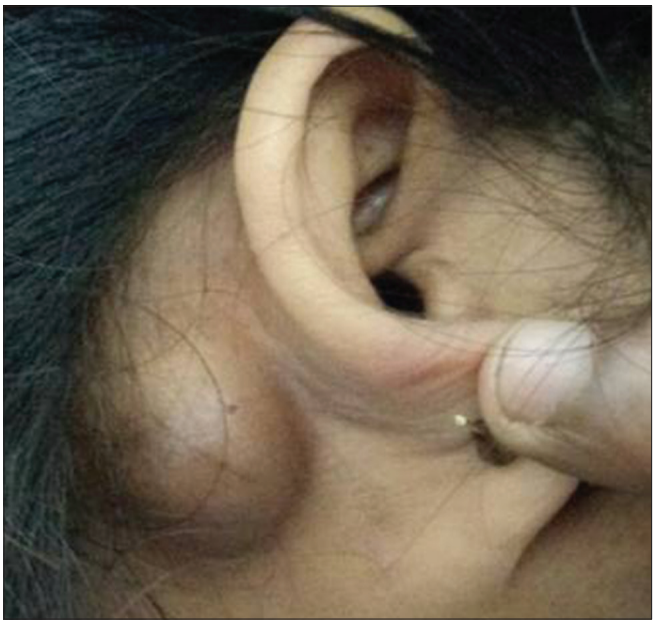

Figure 3: Showing swelling at the right retro auricular region

hyperpigmentation of overlying skin [Fig. 6]. At 2 month, her symptoms and signs decreased further. The patient is on low dose oral corticosteroid since last 2 years. We advised her blepharoplasty for cosmetic purpose, but she refused.

Discussion

KD is believed to be an allergic or autoimmune response to an unknown stimulus, which results in the release of cytokines, which might have a role to play in its pathogenesis. Patients with orbital involvement may present with exophthalmos, eyelid swelling, palpable mass, ocular dysmotility, ptosis, lacrimation, pruritus, pain, or headache. The superior orbit is the most common location of the disease.[7] KD may mimic inflammations of the orbit, neoplasia, dysthyroid orbitopathy, Hodgkin's lymphoma, angioimmunoblastic T cell lymphoma, Langerhans cell histiocytosis, Castelman's disease, eosinophilic granulomatosis with polyangitis, and angiolymphoid hyperplasia (ALHE). Parotid gland enlargement may be

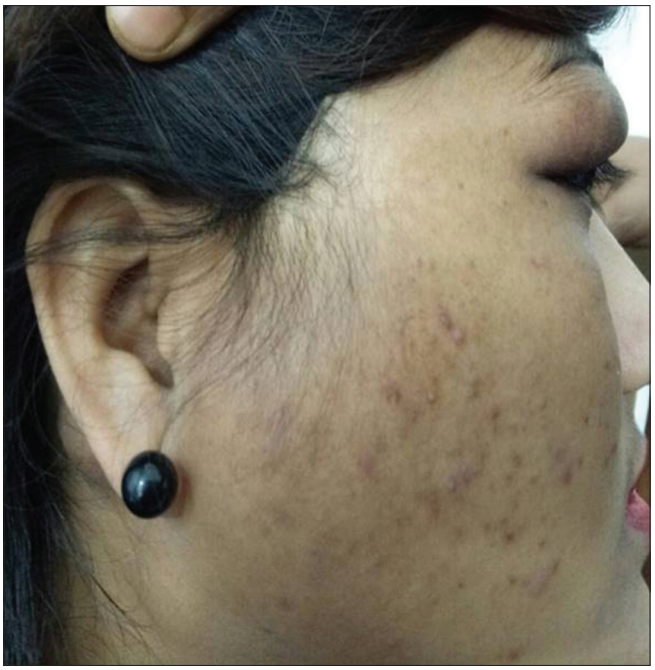

Figure 2: Showing diffuse swelling of right cheek and parotid region

misdiagnosed with common lesions of the parotid, like Mikulicz's disease, neoplastic lesions, ALHE, etc. ALHE usually affects middle-aged woman. It is associated with eosinophilia, but rarely lymphadenopathy with no increase of IgE.[8] In our case, there was elevated IgE and absence of vascular proliferation of endothelial cells in HPE (feature of ALHE). The lesions of KD may be associated with renal complications like nephrotic syndrome.[9]

Observation has been recommended for asymptomatic lesions. Medical treatment has been advised using cetirizine, corticosteroids, cyclosporine, and retinoid with a variable degree of success. However, recurrence on cessation is a problem. Intravenous immunoglobulin has been reported to give good remission.[10] Surgery has been performed for primary, isolated lesion. Radiotherapy has been occasionally used for recalcitrant and large tumors and in cases refractory to surgical and medical therapy and in young patients.[11] Although spontaneous resolution has been reported, most patients have a prolonged course with slow enlargement of the masses. There is no potential that the lesions will become malignant.[12] Our patient is coming for follow-up regularly, she is symptom-free, and there is no intolerance to steroid till date.

Conclusion

Kimura's disease, although rare, must be kept in mind in the differential diagnosis of lacrimal gland and extraocular muscles enlargement; especially when there is marked eosinophilia. A correct diagnosis is important because of its good response to nonsurgical treatment and high rate of recurrence.

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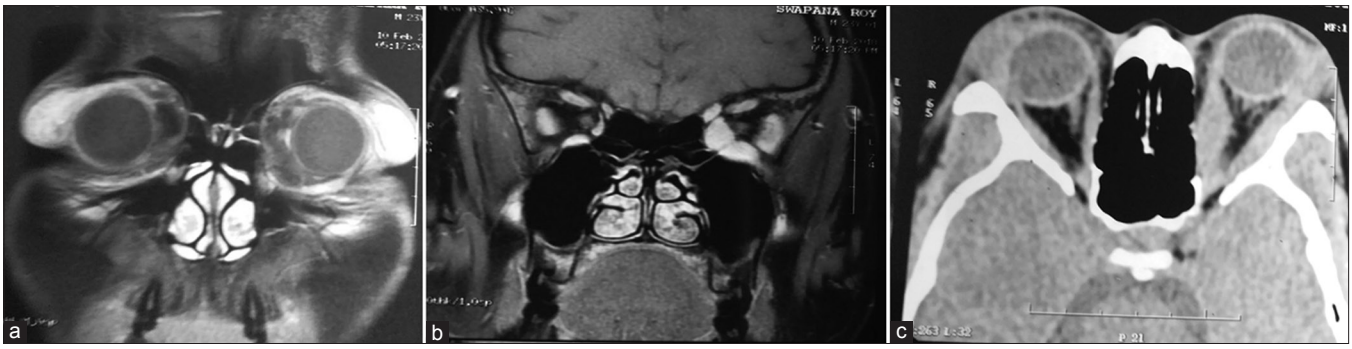

Figure 4: (a) Contrast MRI scan orbit (coronal view) showing bilateral enlarged lacrimal gland. (b) Showing diffuse thickening of all extraocular muscles along with enhancement of right temporalis and masseter muscles. (c) Axial view CT showing thickening of all extraocular muscles with sparing of tendons along with orbital infiltration (RE>LE)

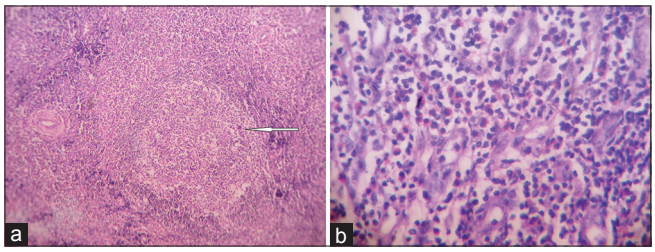

Figure 5: (a) Histopathological examination of postauricular lymph node showing follicular hyperplasia and proliferation of post capillary venules (H and E stain × 100). (b) Showing plenty of tiny mature eosinophilic collections/abscesses (H and E stain × 400)

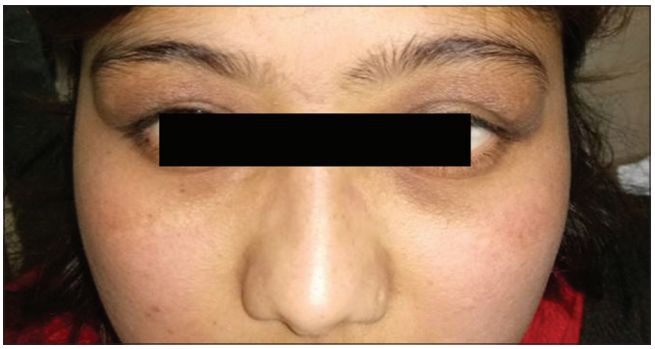

Figure 6: Showing remission of swelling of upper lids and face with a decrease in pigmentation of overlying skin

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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