ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 8, Issue, 08, pp.36107-36109, August, 2016

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

KIMURA'S DISEASE AS UNILATERAL LID MASS – A RARE CASE REPORT AND REVIEW OF LITERATURE

Dr. Ramesh Chander Nagpal, *Dr. Anuradha Raj, Dr. Nadia Shirazi and Dr. Harsh Bahadur

Department of Ophthalmology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Swami Ram Nagar, Jolly Grant, Dehradun, Uttarakhand, India

ARTICLE INFO

Article History:
Received 12th May, 2016
Received in revised form
23rd June, 2016
Accepted 18th July, 2016
Published online 20th August, 2016

Key words:

Extraocular muscle, Eyelid swelling,
Kimura's disease.

ABSTRACT

Kimura's disease (KD) is a chronic inflammatory disorder with angiolymphatic proliferation, predominantly affecting young men of Asian race but is rare in other races. The etiology of KD is still unknown. We report a case of 23 year old male with Kimura's disease involving right upper lid along with eosinophilia.

Citation: Dr. Ramesh Chander Nagpal, Dr. Anuradha Raj, Dr. Nadia Shirazi and Dr. Harsh Bahadur, 2016. "Kimura's disease as unilateral lid mass – A rare Case report and Review of Literature", *International Journal of Current Research*, 8, (08), 36107-36109.

INTRODUCTION

Kimura's disease (KD) was first described by Kimura (Kimura *et al*., 1948). It is one of the rare diseases that predominantly occurs in Asians. KD is idiopathic chronic inflammatory disease characterized by recurrent subcutaneous mass lesions predominantly in the head and neck region of young oriental males. This condition is a benign disorder and may clinically simulate a neoplasm. KD has also been described to involve the chest wall, inguinal region, antecubital fossae, thighs, kidneys, and median nerves (Kung *et al*., 1984). But orbital, eyelid and lacrimal gland involvement in KD is extremely rare (Lee *et al*., 2009). Peripheral blood eosinophilia with an elevated IgE level is a common finding. Patients with orbital KD are usually older (30-50 years) than the typically teenaged KD patient (Buggage *et al*., 1999). Although orbital KD is a benign lesion but it may cause a devastating visual outcome (Moroz *et al*., 1998).

Case report

A 23 years old male presented to outpatient department of ophthalmology, Himalayan Institute of Medical sciences with complain of swelling over right upper lid for last two years which was painless in nature and progressively increased in size since last 2 weeks. On examination unaided visual acuity was 20/20 for both eyes. Hirshberg was central and extraocular movements were full, free and painless bilaterally without any diplopia. No proptosis or exophthalmos was noted. There was a firm, painless palpable mass under the skin of the right upper lid beneath eyebrow associated with puffiness of lid without any signs of inflammation or any associated punctum. There was pseudoptosis of right eye with narrow vertical palpebral aperture of 7 mm. There was hyperpigmentaion of the skin of right upper lid. The mass was mobile, firm and nontender in nature. The lacrimal gland was normal in size. No lagophthalmos was seen. Corneal sensations were normal. Bells phenomenon was good. Lid retraction of the left eye without any lid lag was seen (Figure 1a). Intraocular pressure was 12 and 14 for right and left eyes respectively. Anterior segment and posterior segment examination was unremarkable. The patient was otherwise healthy and normally built without any systemic disease. There was history of having bilateral, symmetric postauricular masses on both sides for about 4 years which increased in size for last 2 years. He got surgical excision of those masses elsewhere under the impression of lymphadeopathy 6 months back. There were big surgical scars on both sides of postauricular area. No mass lesion was palpable in the parotid or submandibular region. No history of any systemic disease could be elicited. Laboratory

Corresponding author: Dr. Anuradha Raj,
Department of Ophthalmology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Swami Ram Nagar, Jolly Grant, Dehradun, Uttarakhand, India.

investigations included elevated erythrocyte sedimentation rate and negative serology for hepatitis B and C and HIV. Chest radiograph was normal. The haemoglobin, platelet counts, thyroid function tests, serum IgE levels, serum calcium and albumin renal fuction tests were normal. Differential count showed eosinophilia (20%). Physical examination revealed no lymphadenopathy or organomegaly. Differential diagnosis includes inflammatory and neoplastic conditions, tuberculosis, cylindroma, Kaposi's sarcoma, pyogenic granuloma, dermatofibrosarcoma protuberans and other infectious lymph node enlargements for example toxoplasmosis. Ultrasound abdomen revealed both average sized normal kidneys. CT chest and MRI excluded evidence of underlying malignancy or lymphadenopathy. Surgical excisional biopsy was done for right upper lid under local anaesthesia and mass was found to be infilterating orbicularis oculi above the tarsal plate. No capsule was found around the mass. The excisional plane was not well defined. The mass excised was sent for histopathological examination. Diagnosis of KD was established on histopathological examination. Patient was started on oral steroids postoperatively. Suture removal was done on 10th post operative day (Figure 1b).

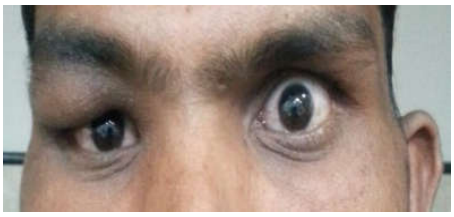

Figure 1a. Photograph of patient showing R/E pseudoptosis with hyperpigmented lid and L/E showing lid retraction

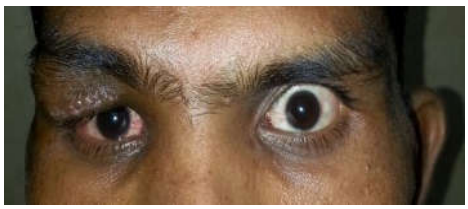

Figure 1b. Photograph showing R/E postoperative lid scar after suture removal

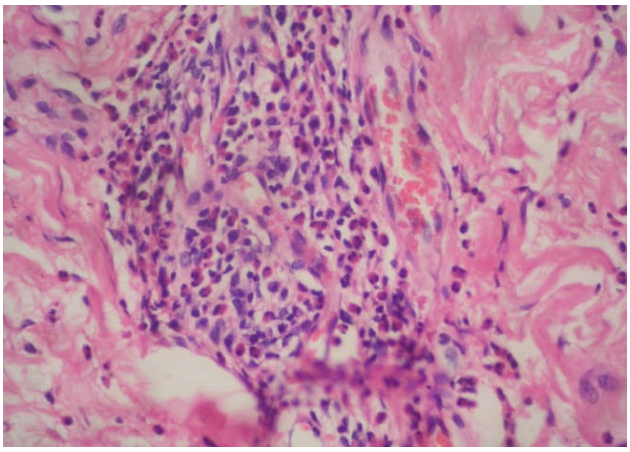

Figure 2. Hematoxylin and Eosin : (40X10x): Numerous eosinophils are seen in lymphoid aggregate

Histopathology

Gross examination showed a single reddish brown firm tissue measuring 20x10x8mm in size.

Microscopic examination: On haematoxylin and eosin staining showed multiple reactive lymphoid follicles with moderate to intense eosinophilic infiltration in the section. Vascular and endothelial proliferation was seen along with few congested blood vessels. Typically, lymphocytes and lymphoid follicles with distinct germinal centers were present (Figure 2). There was no evidence of neurofibroma, parasite, tuberculosis or malignancy in the section. Excision biopsy showed features typically suggestive of KD (eosinophilic lymphogranuloma) on histopathology.

DISCUSSION

KD is an inflammatory process with a predilection for the head and neck region in young men histologically characterised by an angiolymphoid proliferation with eosinophilia and elevated serum immunoglobulin IgE. This disorder got its name as KD following the report in the Japanese literature by Kimura (Kimura *et al*., 1948). First published case with orbital involvement was reported by Nakai in 1966 (Amemiya 1981). Coexisting renal disease is common, with an incidence ranging from 10% to 60% while 10% to 12% of patients may suffer from nephrotic syndrome characterized by clinically relevant proteinuria in 12% to 16% of cases due to immunocomplex-mediated damage or to Th2-dominant immune response disorders (Dixit *et al*., 2004). In a literature search for this disease, cases involving the orbit, lacrimal glands, eyelids, conjunctivae and probably the chorioretina were found with few bilateral cases (Buggage *et al*., 1999). Though rare but KD involves the extraocular muscles (Lee *et al*., 2009). KD should be suspected and included in the differential diagnosis of a caruncular mass lesion (Kim *et al*., 2013). KD occurs predominantly in younger age group of Chinese and Japanese, with longer duration of the disease than ALHE; the lesions are large masses of deep soft tissue without distinct borders. ALHE presents for a short duration in older patients and the lesions are small erythematous papules or nodules which itch and bleed easily. The aetiology is not fully understood. ALHE was considered to be a proliferative reaction of vascular endothelial cells with a secondary inflammatory response whereas an immunological disturbance or an allergic reaction was suggested in KD. First case of ALHE involving the orbit was reported in 1982. Recently it is reported that histologic features of KD differ from those of ALHE (Bostad and Pettersen, 1982). It is believed that KD represents an allergic or autoimmune response, whereas ALHE is considered a benign neoplasm of endothelial cells. There is no evidence for the progression of lesions from the characteristic picture of ALHE to that of KD with the passage of time (Buggage *et al*., 1999). Long-term behaviour of the disease is not documented well. Although some authors use the terms KD and ALHE interchangeably but recent studies suggest that they may be different entities. KD presents as a large solitary lump or multiple lumps, with an insidious onset, long duration, and a frequent occurrence of peripheral blood hypereosinophilia. KD occurs most commonly in young males, usually teenagers.

However, orbital KD tend to occur in older age (30-50 years). Clinically, KD patients present with exophthalmos, palpable orbital with or without eyelid lesions and eyelid edema (Amemiya, 1981). The peripheral blood of KD patients usually show eosinophilia of up to 54% and elevated serum IgE titers (Kuo *et al*., 1988). KD lesions either enlarge gradually and increase in number over time or spontaneously regress so presentation of this disease is late. Systemic associations include asthma and the nephritic syndrome (Kung *et al*., 1984; Hidayat *et al*., 1983). Regional lymph node involvement occurs in up to 75% of cases and recurrence occur in 15%-40% of cases but this entity is nonfatal (Kuo *et al*., 1988). Previously reported cases of periorbital KD were successfully treated with excisional biopsy. Excision is the most widely used therapeutic modality but oral or intralesional steroids, radiation, and chemotherapy can also be used. Incomplete excision can cause recurrence (Kung *et al*., 1984). Being inflammatory process this disease has an excellent prognosis. Cyclosporine has also been reported to be effective in the treatment of KD (Wang *et al*., 2005). In orbital disease radiotherapy and intralesional steroids have not been tried instead conservative management may be the best option, but should include biopsy and histopathological examination to rule out lymphoma. In the present case 25 year old Indian male patient presented with a nodule beneath the right upper lid with history of operated postauricular nodules on both sides. Mass was excised from the orbicularis and similarly located mass lesion has been reported in 50 years old Indian male (Lee *et al*., 2009). In the present case the age is 25 years which is different from that case report.

Conclusion

KD has been predominantly described in oriental males in China and Japan but in Indian population this entity is very rare. Despite its rarity, KD should be included in the differential diagnosis of eye lid nodular lesions but their diversity can make clinical diagnosis difficult. Signs and symptoms of the disease are clueless about this rare diagnosis. Precise and correct diagnosis can be achieved with histopathologic examination of excised tissue. In conclusion, we have added second Indian case of KD to the literature but with a difference of young age of the presentation.

Conflicts of interest: Nil

Financial Support: Nil

Acknowledgements

We acknowledge Mr. Surendra Singh Bhandari, Office Assistant, Department of Ophthalmology for his technical support and assistance in photographic documentation.

REFERENCES

Amemiya T. 1981. Eosinophilic granuloma of the soft tissue in the orbit. *Ophthalmologica*., 182:42-48.

Bostad L, Pettersen W. 1982. Angiolymphoid hyperplasia with eosinophilia involving the orbit: A case report. *Acta Ophthalmol*., 60:419-426.

Buggage RR, Spraul CW, Wojno TH, Grossniklaus HE. 1999. Kimura disease of the orbit and ocular adnexa. *Surv Ophthalmol*., 44:79-91.

Dixit M.P, K. M. Scott K.M, Bracamonte *et al*. 2004. "Kimura disease with advanced renal damage with anti-tubular basement membrane antibody," *Pediatric Nephrology*., vol. 19, no. 12, pp. 1404–1407.

Hidayat AA, Cameron JD, Font RL, Zimmerman LE.1983. Angiolymphoid hyperplasia with eosinophilia (Kimura's disease) of the orbit and ocular adnexa. *Am J Ophthalmol*., 96: 176-189.

Kim WJ, Chung MJ, You IC. 2013. Kimura disease involving a caruncle. *Korean J Ophthalmol*., 27(2):137-140.

Kimura T, Yoshimura S, Ishikawa E. 1948. Unusual granulation combined with hyperplastic changes of lymphatic tissue. *Trans Soc Pathol Jpn.,* 37:179-180.

Kung IT, Gibson JB, Bannatyne PM. 1984. Kimura's disease: a clinico-pathological study of 21 cases and its distinction from angiolymphoid hyperplasia with eosinophilia. *Pathology.,* 16:39-44.

Kuo TT, Shih LY, Chan HL. 1988. Kimura's disease: Involvement of regional lymph nodes and distinction from angiolymphoid hyperplasia with eosinophilia. *Am J Surg Pathol*., 12:843-854.

Lee JKS, Almousa R, Thamboo TP, Arrith S.2009. Kimura disease of the eyelid in an Indian man. *Clin & Exper Ophthalmol*., 37:412-414.

Lee SJ, Song JH, Kim SD.2009. Kimura's disease involving the ipsilateral face and extraocular muscles. *Korean J Ophthalmol.,*23:219-223.

Moroz I, Rosen N, Rosner M.1998. Bilateral Kimura's disease with devastating visual outcome. *Eye*., 12:102-103.

Wang YS, Tay YK,Tan E, Poh WT. 2005. Treatment of Kimura's disease with cyclosporine. J Dermatolog Treat., 16:242-244.

****