International Journal Dental and Medical Sciences Research
Volume 5, Issue 2, Mar - Apr 2023 pp 865-867 www.ijdmsrjournal.com ISSN: 2582-6018

Kimura's Disease - An Unusual Presentation Involving Unilateral Lid

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Date of Submission: 15-04-2023 Date of Acceptance: 25-04-2023

ABSTRACT: This paper analyses a case of Kimura's Disease (KD) in a 31-year-old female, who reported with a complaint of swelling over Right Eye (RE) upper eyelid (UL) since 4 years. On examination, there was a firm, non-tender, circumscribed mass of size 4 x 2 cm in RE UL beneath eyebrow. Laboratory investigations revealed raised TLC, ESR and Eosinophils. Excision Biopsy was done under LA and was sent for Histopathological Examination (HPE). Follow-up was done at 1 week, 6 weeks and 3 months. HPE revealed lymphoid proliferation with lymphoid follicles having prominent germinal centres with few showing vascular proliferation; and Eosinophilic infiltration. KD is common in young men of East Asian population and is rare in Indian population. The present case is an exception described in an Indian female.

KEYWORDS: Kimura's disease, eyelid, TLC, ESR, Eosinophils

I. INTRODUCTION

KD is a rare benign chronic inflammatory disorder of unknown etiology which frequently affects young men of Asian race,[1] characterized by deep subcutaneous masses mostly in the head and neck region with coexisting lymphadenopathy often accompanied by eosinophilia and elevated serum IgE levels.[2] Orbital, eyelid and lacrimal gland involvement in KD is extremely rare.[3,4,5] Treatment options include surgery, oral or intralesional steroids, radiotherapy, photodynamic therapy and laser fulguration. Recently, it has been reported that cyclosporine, azathioprine, pentoxifylline, pranlukast and imatinib are valid treatment options.[6] The case presented here is an exceptional case described in an Indian female. KD is predominantly seen in Eastern Asian population and is rare in Indian population. This case report analyses a case of Kimura's disease in a 31 year old female.

II. CASE REPORT

A 31 year old female patient was presented to our centre with swelling over RE UL since 4 years, which was insidious onset, non-progressive and painless. Ocular examination revealed firm, non-tender, fairly circumscribed mass of size 4 x 2 cm in RE UL beneath eyebrow without any signs of inflammation [Fig.1].

No other abnormalities were detected on ocular & general physical examination. Laboratory investigations revealed raised Total Leukocyte Count (TLC), ESR and Eosinophils. Excision Biopsy of mass was done under LA and was sent for HPE [Fig.2].

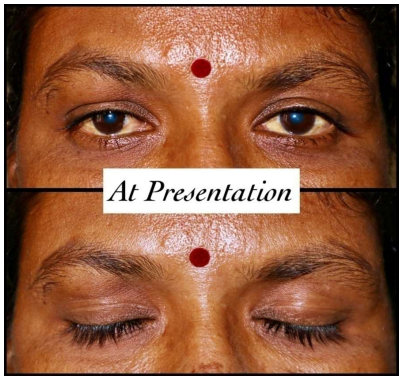

Fig.1: Status on Presentation

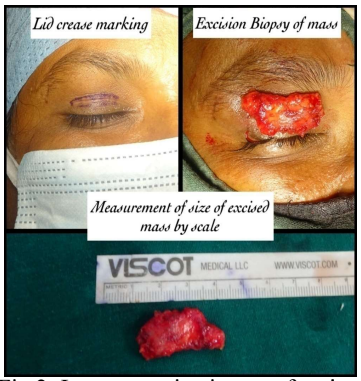

Fig.2: Intra-operative image of patient

International Journal Dental and Medical Sciences Research
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Post-operatively, oral corticosteroid therapy (Tab. Prednisolone 50 mg OD for 5 days) was given. HPE showed dense lymphoid proliferation with lymphoid follicles having prominent germinal centres with few showing vascular proliferation, and eosinophilic infiltration [Fig.3].

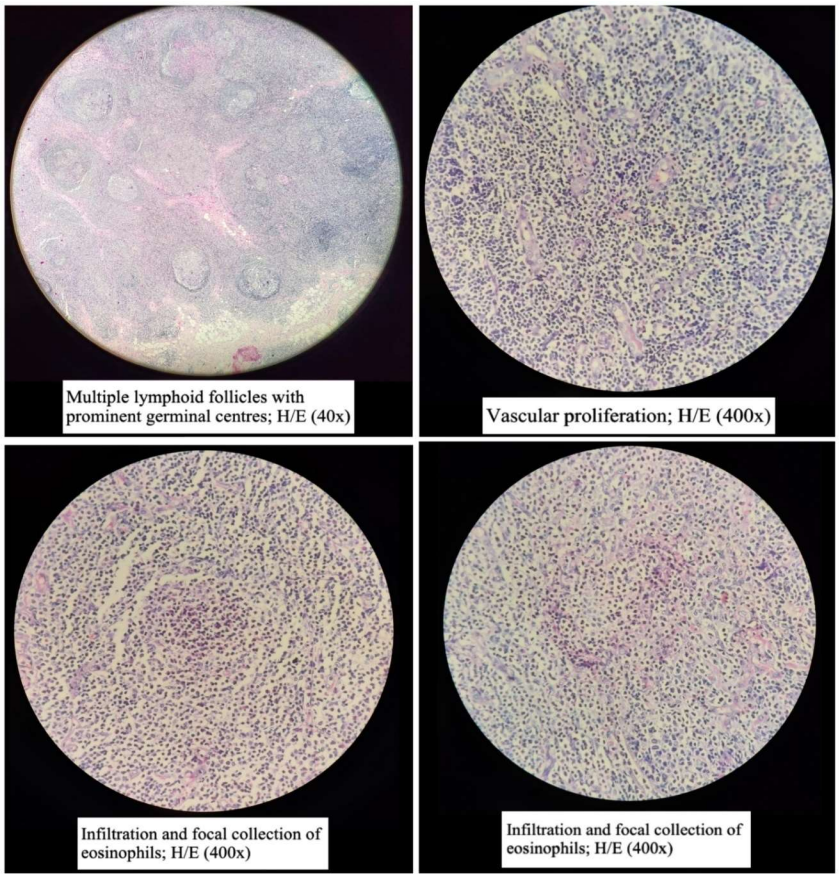

Fig.3: HPE of excised mass

Diagnosis of Kimura's disease was established on HPE. Follow-up was done at 1 week, 6 weeks and 3 months [Fig.4]. Patient was under regular follow-up thereafter.

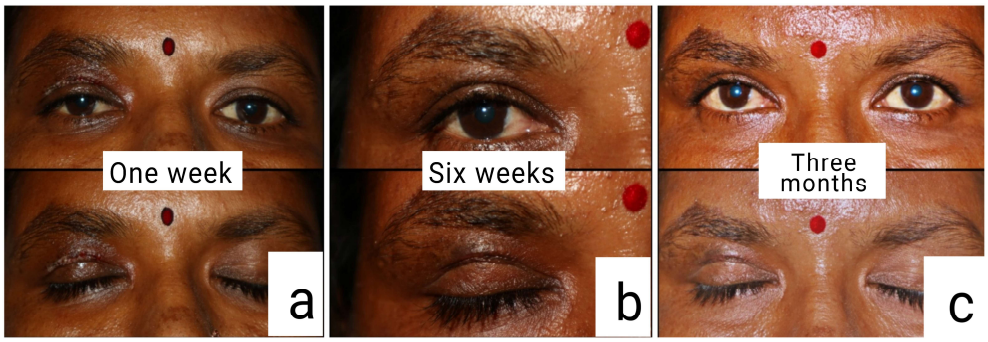

Fig.4: Follow up of patient

III. DISCUSSION

Kimura's disease is a chronic inflammatory disorder of unknown origin. The disease can become apparent at any age, but most commonly occurs in the second and third decades of life, with 80-87% of the affected patients being males.[7] In the present study, the patient was a female who presented in her 4th decade.

The usual clinical presentation is one or more slowly increasing subcutaneous nodules in the head and neck regions, accompanied by regional lymphadenopathies and/or salivary gland enlargement.[3] Laboratory findings in Kimura disease include raised peripheral blood eosinophil counts and markedly elevated serum immunoglobulin E (IgE) level.[8] In our study, the patient showed increased TLC, ESR and eosinophil count. However, serum IgE was not assessed.

Histologically Kimura's disease presents as preserved lymph node architecture with reactive and prominent germinal centers. Dense eosinophilic infiltration of the interfollicular zones, lysis of the follicles, and occasionally microabscesses are seen. Granuloma formations contain infiltration of eosinophils, lymphocytes, plasma cells, and histiocytes. Tissue fibrosis, sclerosis, and vascular proliferation are also present. Vessels remain thin-walled with cubical endothelial cells.[3]

Generally, Kimura's disease is a localized condition with a benign clinical course but recurrence is not uncommon. Therapeutic options include surgery, radiotherapy, laser fulguration, photodynamic therapy and steroids.[9] Steroid is effective in reducing size of the mass, but the lesions may recur while reducing the dose of steroid.[10]

IV. CONCLUSION

Kimura's disease is common in East Asian population and is rare in Indian population. Recurrence is not uncommon. Overall outcome is good as there's no association with the malignancy. The case presented in this paper is an exceptional case described in an Indian female.

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