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A rare case report: Kimura's disease presenting as bilateral post auricular swellings in a young male.

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Abstract: Kimura disease (KD) is a chronic inflammatory disorder with angiolymphatic proliferation, usually affecting young men of Asian race but is rare in other races. The disease is characterized by a triad of painless subcutaneous masses in head and neck region, blood and tissue eosinophilia and moderately elevated serum immunoglobulin E levels.
Keywords: Kimura's Disease; Eosinophilia, Lymphadenopathy.

INTRODUCTION

Kimura's disease (KD) is a rare chronic angiolymphoid proliferative soft tissue disorder of unknown origin [1]. It occurs most often in young and middle aged Asian male. It is most common in head and neck region, with a predilection for preauricular area. Typical clinical presentations are painless subcutaneous masses, regional lymph node enlargement, blood and tissue eosinophilia, and markedly elevated serum IgE levels. According to the previous medical literatures, Kimura's disease has a high recurrence rate so early & definitive diagnosis of the disease is vital for effective treatment plan [1]. Here we report a case of Kimura's disease with clinico-pathological differential diagnosis.

CASE REPORT

A 24 year old man reported to outpatient department with chief complaint of painless swelling both sides post auricular region since 4 months slowly increasing in size. With a provisional diagnosis of lymphnodal masses, an incisional biopsy was performed. The section stained with Haematoxylin & Eosin revealed unencapsulated ill-defined lesion with lymphoid follicle formation, fibrosis and chronic inflammatory cell infiltration with predominant eosinophils (Fig-1 to Fig-3). Inter follicular areas show increased number of venules with endothelial cell proliferation. In some areas lymphoid follicle were infiltrated by eosinophils with resultant eosinophilic micro abscesses. With a diagnosis of Kimura's disease, the patient underwent excisional biopsy both the swellings.

The excisional biopsy specimen received was firm nodular soft tissue measuring approximately 4.5 X2.2X1.5 cm3 and two in number. The histopathological feature of excisional biopsy was matched with that of incisional one. The patient was supplemented with oral Dexamethasone for total duration of 6 weeks (0.5 mg twice daily for 1 week and then tapered over 5 weeks). The patient was apparently normal without any local recurrence & systemic involvement within 6 month of follow-up period.

DISCUSSION

Kimura's disease first described in1937 in Chinese literature by H.T.Kimm and C.Szeto and they termed it as "eosinophilic hyperplastic lympho granuloma" [2]. The disease became widely known as Kimura's disease after Kimura and colleagues reported two cases of unusual granulation combined with hyperplastic changes of lymphoid tissue [1]. The etiopathogenesis of KD remains unknown and it is considered nowadays as an allergic disease and it seems to be a systemic immunological disorder.) The etiology of KD is still unknown but may be due to impairment or interference with immune regulation, atopic reaction to a persistent antigenic stimulus by arthropod bites, virus, and neoplasm. The most interesting hypothesis suggests Candida acting as a source of persistent antigenaemia, although neither hyphae nor spores have been isolated. The disease is manifested by hyperplasia of lymphoid follicles and vascular endothelium. Peripheral eosinophilia and the presence of eosinophils in the inflammatory infiltrate suggest that KD might be a kind of hypersensitivity reaction. With lymphocyte, T-helper 2 (Th2) might play a role [6].

It occurs predominantly as a unilateral manifestation in the head and neck and it is frequently associated with regional lymphadenopathy with or without involvement of salivary glands [4]. In the present case, presence of bilateral postauricular lymphnodal swellings led to the clinical diagnostic dilemma of other pathologic entities affecting salivary glands & lymph nodes. The differential diagnosis of

Kimuras disease frequently in previous literatures are Lymphomas, salivary gland neoplasms, benign lymphoepithelial lesions (BLL / Mikulicz's disease), Angiolymphoid hyperplasia with eosinophilia (ALHE/ Epitheloid hemangioma) and angioimmunoblastic lymphadenopathy (AIL). Constant classical features of KD include numerous lymphoid follicles, mixed inflammatory infiltrate composed mainly of eosinophils and increased amount of post capillary venules [5]

The diagnostic challenge of KD is generally solved by histological study: although there is no specific diagnostic feature of Kimura disease, fine-needle aspiration cytology is helpful in some cases, and definitive diagnosis can be obtained by histological examination of the excised lesion [6].

Treatment for Kimura disease includes surgical resection and regional or systemic steroid therapy. Cytotoxic therapy and radiation have also been utilized. The disease has an excellent prognosis, although it may recur locally [7]. Sun *et al.;* [8], reported that Imatinib—previously to be useful for treatment of hyper eosinophilic syndrome and may work by selectively blocking protein-tyrosine kinases—might be an effective drug for the treatment of the disease.

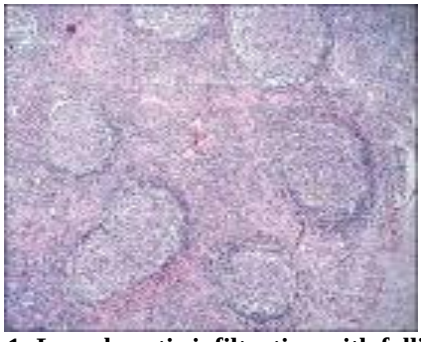

Fig-1: Lymphocytic infiltration with follicle formation and fibrosis. Well defined lymphoid follicle with prominent germinal centre, (10x)

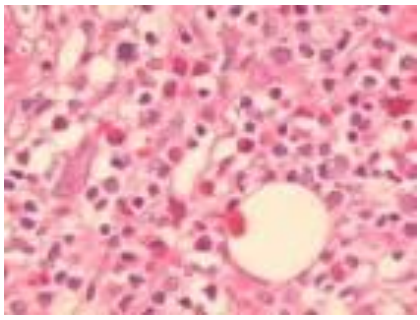

Fig-2: Chronic Inflammatory cell infiltration with predominant eosinophils with micro abscesses. (40x)

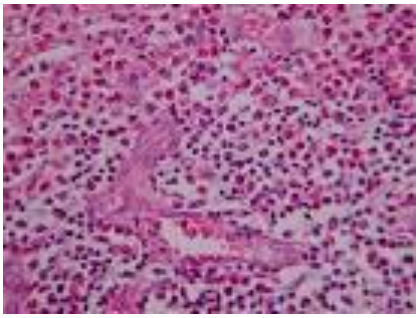

Fig-2: Interfollicular areas showing mixed inflammation with prominent eosinophils and proliferation of venules. (40x)

CONCLUSION

In conclusion kimuras disease is a rare chronic inflammatory disease which mimics neoplastic conditions. This disease should be considered in differential diagnosis of patients presented with head & neck mass and lymphadenopathy and investigated accordingly as this disease has good prognosis. Knowledge of important features of Kimura's disease put the clinicians in a better position to evaluate its clinical outcome and optimal treatment regimen. .

Competing interests:

We have no competing interests

Consent:

Taken from the patient

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REFERENCES

1. Chung YG, Jee WH, Kang YK, Jung CK, Park GS, Lee A *et al.;* Kimura's disease involving a long bone. Skeletal Radiology 2010; 39(5):495-500.
2. Pitak-Arnnop P, Bellefqih S, Chaine A, Dhanuthai K, Bertrand JC, Bertolus C; Head and neck lesions of Kimura's disease: Exclusion of human herpesvirus-8 and Epstein-Barr virus by in situ hybridisation and polymerase chain reaction. An immunohistochemical study. Journal of Cranio-Maxillofacial Surgery 2010; 38 (4):266-70.
3. Briggs PL; Kimura disease is not angiolymphoid hyperplasia with eosinophilia: clinical and pathological correlation with literature review and definition of diagnostic criteria. Anais Brasileiros de Dermatologia 2006; 81(2):167-73.
4. Kung ITM, Gibson J, Bannatyne P; Kimura's Disease: A Clinico-Pathological Study of 21 Cases and Its Distinction from Angiolymphoid Hyperplasia with Eosinophils. Pathology1984; 16(1):39-44.
5. Jani A, Coulson M; Kimura's disease. A typical case of a rare disorder. Western Journal of Medicine 1997; 166 (2):142-4.

6. Tseng CF, Lin HC, Huang SC, Su CY; "Kimura's disease presenting as bilateral parotid masses," European Archives of Oto-Rhino-Laryngology, 2005; 62(1): 8–10.
7. Abuel-Haija M, Hurford MT; "Kimura disease," Archives of Pathology and Laboratory Medicine, 2007; 131(4): 650–651.
8. Sun QF, Xu DZ, Pan SH, Ding J.G, Xue Z.Q, Miao C.S *et al.;* "Kimura disease: review of the literature," Internal Medicine Journal, 2008; 38(8): 668–672.