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CASE REPORT

KIMURA'S DISEASE OF HARD PALATE: A RARE PRESENTATION OF A RARE DISEASE

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ABSTRACT: Kimura's disease is a rare chronic inflammatory disorder affect mainly. Affecting young Asian males, more so oriental men.Most common sites of involvement are deep Subcutaneous tissue, nodules in head and neck region frequently associated with regional lynphadenopathy or salivary gland involvement. Such clinical Presentation closely mimics a neoplastic lesion. We report a case of kimura's disease arising in hard palate of a 11 year old boy which is a very unusual site of involvement by this disorder and hence merits documentation to create awareness about such as presentation of Kimura's Disease.

KEYWORDS: Kimura's disease, Eosinophilia, Lymphadenopathy follicular Hyperplasia, IgE.

INTRODUCTION: Kimura's disease is a rare chronic inflammatory disease of unknown etiology.[1] Young and middle aged Asian males of Chinese and Japanese origin are primarily affected.[2] The prevalence in patients of other ethnicities is considered low.[3] The typical clinical presentation is characterized by a triad of painless unilateral cervical adenopathy or subcutaneous mass predominantly in the head or neck region, blood and tissue eosinophilia and markedly elevated serum immunoglobulin E(IgE) levels.[2]

Typical areas for the nodules are preauricular, sub mandibular and popliteal regions.[4] Other sites of involvement are oral cavity, groin, limbs, trunk, eyelids, orbit and lacrimal glands.[5] Etiology is thought to be an abnormal immune response to an unknown antigenic stimulus[6] even though it is a benign disease, it can produce devastating renal and thrombotic complications.[6] The diagnosis of Kimura's disease can be very difficult and misleading[4] Kimura's disease has characteristic histologic features that are important to recognize.[7]

CASE REPORT: An eleven years old male child presented to our ENT OPD with history of slowly gnawing mass on hard palate since six months. There was no other significant complaint apart from pruritis. Local examination revealed a lobulated mass on hard palate which was 3.5x3cms in size, firm and non-tender. There was a discrete, mobile, non-tender lymph node on right side of neck- Investigations revealed hemoglobin- 11.5gm%, WBC count -8,200/mm^3 Differential count- Neutrophils 38%, Lymphocytes – 40%, eosinophils – 21% Monocytes – 01% Basophils – 00.

Absolute eosinophils count (AEC) was 1722/mm^3 platelets were adequate.

Biochemical investigations revealed Blood urea – 22.4 mg/dl (N- 15 – 45mg/dl) and S. creatinine 0.7 mg/dl (N- 0.6 – 1.2 mg/dl) FNAC from the palatal mass revealed moderate cellularity with presence of lymphoid cells, benign squamous cells with eosinophilia. Cytological diagnosis of chronic non-specific inflammatory lesion was rendered and the biopsy of the palatal mass was advised to rule out various causes of eosinophilic infiltration of in the palatal mass. Excision biopsy of the mass was done and sent for histo pathological examination.

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Microscopic examination revealed minor salivary gland acini densely infiltrated by eosinophils and at places replacement of gland parenchyma by lymphoid follies which are rich in eosinophils. Histo pathological diagnosis of Kimura's disease was thus made. With this histopathological diagnose few specific investigations were carried out– immunoglobulin levels, urinary protein and renal function tests. Immunoglobulin levels showed increased levels of lgE and normal levels of other immunoglobulin. He had no renal involvement as his renal function tests were within normal limits. Now it has been six months post-surgery. Patient is without local recurrence or renal involvement

DISCUSSION: Kimura's disease was initially described in 1937, in the Chinese literature, by Kim and Szeto as an 'eosinophilic hyperplastic lymphogranuloma' and it became to be known as Kimura's disease after its publication by Kimura et al[2] of similar cases in Japan under the title "atypical granulation associated with hyper plastic abnormalities in the lymphoid tissue." In 1948.[3,6] wells and Whimster in 1969 first reported patients in the west with a similar condition which they called Angiolymphoid hyper plastic with eosinophilia (ALHE) China and Japan are endemic countries although sporadic cases have been described elsewhere.[8]

The clinical course of kimura's disease is generally being and self-limited.[2] It presents as a deep, subcutaneous mass in the head and neck region and is frequently associated with regional lymphadenopathy or salivary gland involvement.[7] Rare sites of involvement include the kidneys, orbits, ears, spermatic cord and nerves.[6] The nodular lesions are deep seated in subcutaneous tissue and clinically may mimic a neoplasm.[7] Lymph node involvement in KD has been reported to range from 67% to 100%.[8]

Although the masse enlarge slowly, patients remain asymptomatic otherwise, purities and dermatitis may occur.[6] Systemic associations includes asthma and nephritic syndrome.[8] Renal involvement may effect upto 60% of patients as membranous glomerulonephritis, minimal change glomerulonephrities, Membrano proliferative glomerulonephritis, and also nephritic syndrome.[9,4] Lesions of Kimura's disease usually precede or coincide with the development of renal disease. Occasionally Kimura disease may present with renal involvement before the appearance of subcutaneous lesions, leading to delayed diagnosis.[2]

Widespread disseminated intravascular thrombosis is also reported in literature, offending mesenteric and renal veins.(thrombotic storm)[(10)]

The cause and pathogenesis of Kimura's disease is unclear although it might be a self-limited allergic or autoimmune response triggered by an unknown stimulus.[(2)] Allergic reactions, infections and auto-immune reactions with an a aberrant immune response have been suggested as possible etiological factors.[5]

It has been speculated that a viral or parasitic trigger may alert t-cell immune regulation or induce an IgE mediated type I hypersensitivity resulting in the release of eosinophilic cytokines.[2] IL-4, IL-5, IL-13. This may finally lead to peripheral eosinophilia and increased level of IgE in serum.[1]

Association has been suggested with Candida albicans,[11] human herpes virus-8[12] and Epstein Barr Virus[13] without any conclusive evidence. The most popular theory is that of candida acting as a source of persistent antigenemia, although neither hyphae nor spores have been isolated.[9]

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Histologicalyl, Kimura's disease presents as preserved lymph node architecture with reactive and prominent germinal center,[4] the pathology of Kimura's disease is characterized by three Components:

(1) Cellular: inflammatory infiltrate inducing eosinophils and follicular hyperplasia.
(2) Fibrocollagenonus and,
(3) Vascular – arborizing vascular proliferation of the post capillary renule.[2,7]

Constant Features Seen Include: Preserved nodal architecture, florid gerimal center hyperplasia, eosinophilic infiltration and post capillary venule proliferation[7] germinal centers are often well vascularized and contain polykaryocytes (Warthin Finkeldy type giant cells) proteinaceous deposits and cosinophils.[5] Frequent features include sclerosis, karyocytosis in both the germinal centers and the paracortex necrosis of germinal canters, eosimophilic[6] abscess and atrophic venules in sderotic areas.[6]

Rarely, progressive destruction of germinal centers may be seen[4] Immune peroxides studies show IgE reticular network in germinal centers, [(2)] and variable amounts of IgG, IgM and fibrinogen [(4)] The aforementioned histologic findings differentiate kimura's disease from more common causes of head and neck masses.[6]

It's differential diagnosis include ALHE, eosinophilic granulona, mikulicz 's disease, acute lymphoblastic leukemia, Hodgkin's disease, angioimmunoblostic lymphadenopalhy[9] Castleman's disease, dermatopathic lymphadenopathy, allergic granulomatosis, drug reaction and parasitic lymphadenitis[9,5] Malignancy should be ruled out first.[6]

The absence of Reed Sternberg cells helps to exclude Hodgkin's disease. Atypical histiocytosis X can present with subcutaneous masses the diagnosis can be made by fending the characteristic abnormal histiocytes and detecting CD1A marker.[6]

The differential diagnosis with ALHE is of particular concern.[5] Differentiating Kimura's disease from ALHE requires strict analysis of clinical and histologic features because the diseases are similar and were once thought to be the same disorder.[6]

Now it is clear that in angiolymphoid hyperplasia with eosinophilia {ALHA} histiocytoid blood vessels with vacuolated endothelial cells are seen with intact germinal centers in contrast to Kimura's disease where germinal centers are destroyed due to heavy infiltration of eosinophils and there is absence of vacuolated endothelial cells[9] the germinal center vascularization and vascular proliferation may be present in hyaline vascular type castleman's disease but the germinal centers are usually atrophic without eosinophilic infiltrate. Florid follicular hyperplasia may show many of the features of Kimura's disease however eosinophilic abscess, eosinophilic infiltrates in germinal centres and folliculolysis are absent[7]

Diagnosis of Kimura disease can be difficult.[2] It can easily be mistaken for a malignant disorder.[9] On radiological examination kimura's disease mimics other chronic and malignant diseases such as tuberculosis or lymphoma[4] FNAC can be misleading[9] Histopathological examination is diagnostic.[5]

The treatment of kimura's disease is problematic.[2] Although spontaneous resolution has been reported most patients have a prolonged course with slow enlargement of the masses.[6] Surgery is the mainstay of therapy although relapses are frequent.[6,7] Additional medical therapies

including regional and systemic steroid therapy, cytotoxic therapy and radiation have also been employed.[5]

We conclude that although rare, kimara's disease is an important differential diagnosis to be considered for all head and neck swelling with eosinophilia as the specific histologic diagnosis reassures the patient about the benign nature of this disease which may have worrisome clinical presentation thus guiding the correct therapeutic approach.

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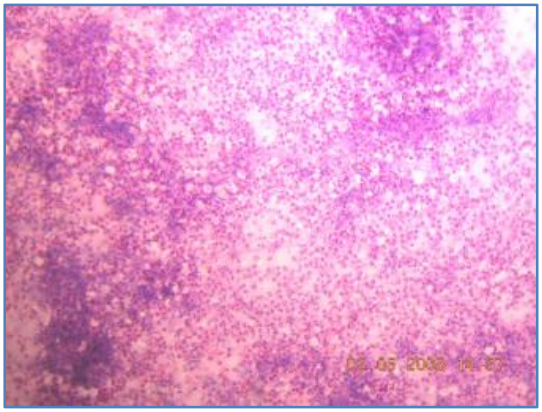

Fig. 1: FNAC (10X10 100x, H and E) image showing eosinophil rich chronic inflammatry infiltrate

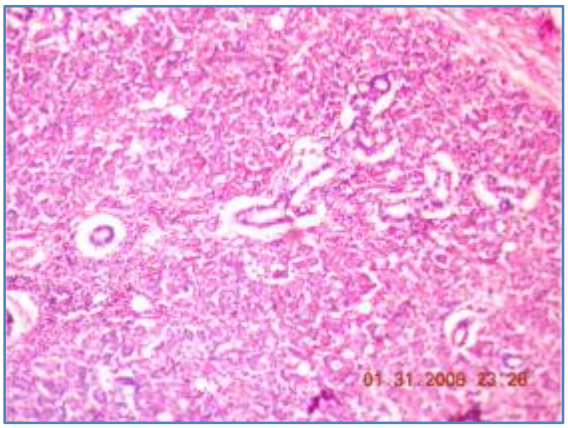

Fig. 2: Photo micro graph (10 x10x, 100X H and E) photo micro graph showing intense eosinophilic infiltration and destruction of gland parenchyma

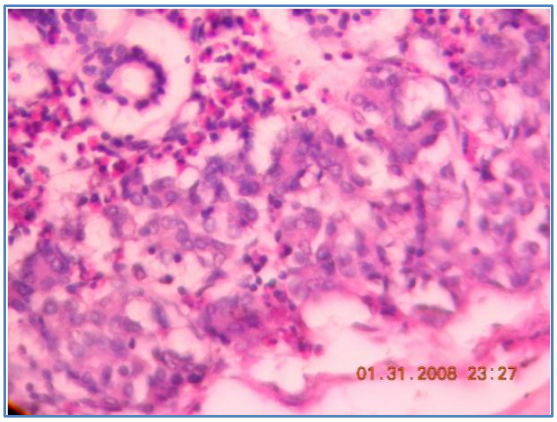

Fig. 3: Photomicro graph (10X 40X, 400X H and E) showing gland parenchyma infiltrated by enosinophils

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