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Case Report

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Kimura Disease: A Diagnostic Dilemma in A 51 Years Old Male Presenting with Submandibular Swelling

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Abstract

Background: Kimura's disease is a rare benign chronic inflammatory disorder of unknown origin that involves subcutaneous tissue and lymph nodes. Primarily seen in males predominantly in Asian populations like Chinese and Japanese. The lesion is benign, but it may easily be mistaken for a malignant tumor. ***Case Presentation:*** We describe a case of Kimura disease in a 51 years old male presenting with left submandibular swelling. ***Conclusion:*** Kimura disease has been confused with angiolymphoid hyperplasia with eosinophilia (ALHE), for which it should be distinguished separately.

Keywords: ***Kimura Disease, Lymphadenopathy, Eosinophilia.***

Introduction

Kimura's disease is a rare chronic inflammatory disorder of unknown etiology, predominantly seen in Asian population. It is further reported that surgical and medical therapy are extremely beneficial for the treatment of the condition [1]. Male to female is found to be 3:1. Most common in 3rd decade of life. The first case was reported in China by Kimm and Szeto in 1937.The clinical presentation is characterized by Triad of painless unilateral lymphadenopathy or subcutaneous masses predominantly in head and neck region, peripheral eosinophilia and elevated Serum IGE levels. This disease is rare in India with only 200 cases reported in the world after histopathological diagnosis and the primary treatment of choice is surgical approach [2]. Multiple organ involvement has been noted but renal involvement is found to be fatal. Early diagnosis of Kimura's disease may spare the patient from potentially harmful procedures. Unnecessary invasive diagnostic procedures can be avoided with prompt identification of the disease.

Case Report

A 50-year old male patient was presented to the surgical Outpatient Department (OPD) with the complaint of left submandibular swelling (**Figure 6**) for the past 3 years which was insidious in onset and gradually progressive. This swelling grew slowly and was not associated with pain and was occasionally pruritic.

He also noticed similar swellings present in left cheek, left temporal region (**Figure 7**) and right and left hospital regions since 5 years. His medical, surgical and family histories were non-contributory. He was not taking medications and he had no drug allergies. On examination, a 3x4 cm lobulated, non-tender, mobile, non-pulsatile, soft to firm swelling in consistency was noted. The skin over the swelling appeared normal and was freely movable. There was a similar swelling in the temporal area on the left side of size 1x1 cm soft to firm in consistency, non-pulsatile, no erosion of skull bone at the site of swelling. There was also a swelling in the left cheek of size 2x2 cm with soft to firm in consistency, non-pulsatile, not attached to mandible and also a swelling measuring 2x2 cm in the left occipital region, 4cm away from the midline, parallel to the tip of the earlobe which was soft to firm in consistency and non-pulsatile. There was no fever, no enlarged palpable lymph node in the neck. The rest of the examination was unremarkable. Hematological examination revealed 13.90 gm/dl, TLC 6,610 cells/cumm (Neutrophil 60.3%, Lymphocytes 16.8%, Monocytes 2.3%, Eosino.phils 20.4%) and platelets 227 x103/µL. Absolute eosinophil count 700 cells/cu mm, Serum IgE level >1000 IU/ml (normal upto 100 IU/ml) (**Table 1**). Ultrasound (USG) (**Figure 1**) of left submandibular gland and Magnetic resonance Imaging (MRI) (**Figure 2**) was performed. FNAC (**Figure 3,4**) and excision biopsy (**Figure 5**) were also performed.

Table 1: Lab investigations.

CBC	RFT	Lipid Profile
RBC- 4.98 million/cumm	Urea - 18 mg/dl	Total cholesterol - 129mg/dl
Hb -13.90 g%	S. Creat - 0.9mg/dl	Triglycerides - 70 mg/dl
HCT – 41.9%		HDL- 61 mg/dL
PLT- 227×10^3 /µL		LDL- 54 mg/dl
TC- 6.61×10^3 /µL		VLDL- 14 mg/dl
• Neu-16.8%		
• Lym - 16.3%		
• Mon - 2.3%		
• Eos-20.4% (normal 2-3%)		
• Bas - 0.2%		
• LIC - 0.4%		

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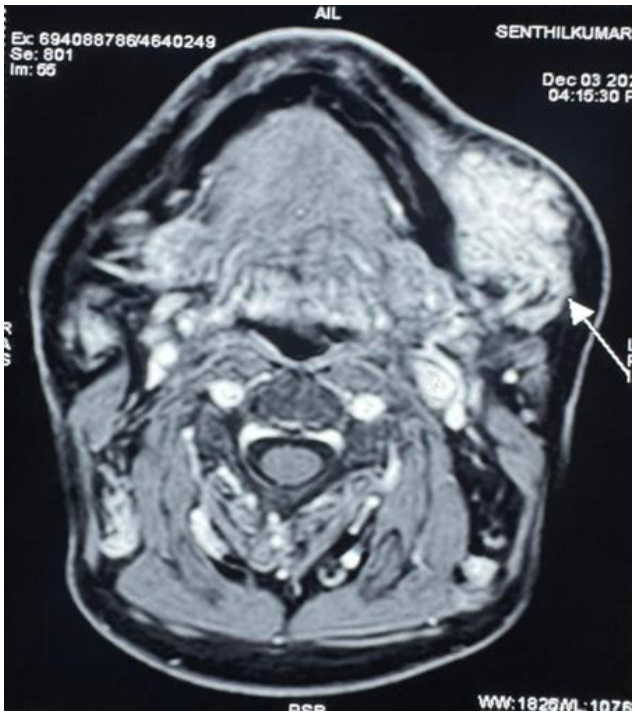

Figure 1: Showing USG of left submandibular swelling(arrow).

USG Left Submandibular Gland
• Ill-defined soft tissue mass 40 x 27 x 23 mm in left submandibular region extending to lateral aspect Pop list mandibular body, Areas of cystic changes with slow flow noted throughout the mass. • The mass is displacing left submandibular salivary gland medially. • No focal lesions in bilateral submandibular salivary glands, no collection. • Few subcentimeter left level two cervical lymph nodes. • Suggestive of soft tissue hemangioma.

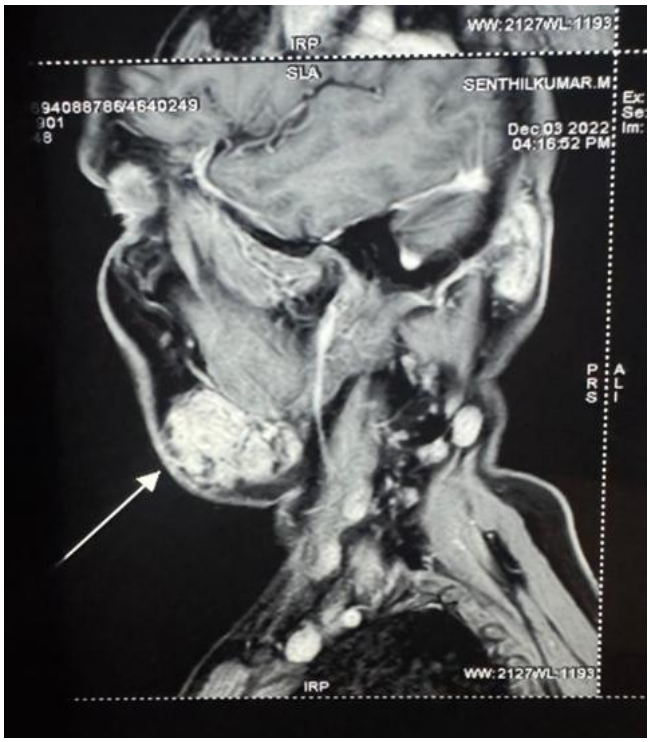

Figure 2: Showing MRI of neck, arrow indicating the left submandibular swelling.

MRI Neck
• Soft tissue lesion noted in left submandibular region measuring 3.7 x 2.6 x 2.3. • No bone erosion noted. • Few subcentimetre submandibular lymph nodes on the left side. • Submandibular gland appears separate and normal. • Moderate enhancement of the lesion noted after contrast enhancement. • Parotid glands appear normal.

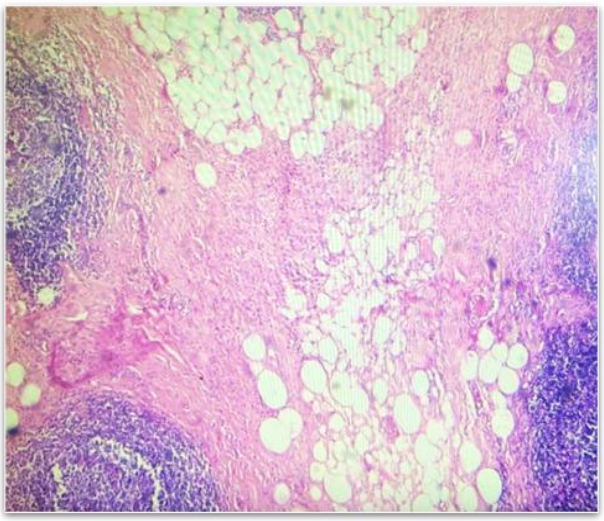

Fig 3. FNAC from swelling showed lymphoid follicles showing and soft tissue (low power).

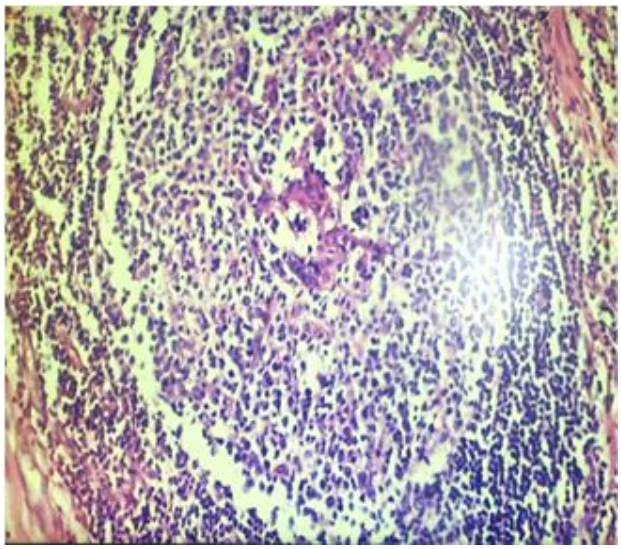

Fig 4. FNAC from swelling showing lymphoid follicles with germinal center (high power).

FNAC Report
• USG-Suggestion of soft tissue hemangioma. • Gross appearance- Swelling Left Submandibular area measuring 3 x 4. • Microscopic appearance-Smears are cellular with lymphocytes in various stages of maturation amidst which are seen clusters of histiocytes, many of them showing abundant eosinophilic cytoplasm. • Impression-Reactive lymphoid cells and eosinophils are seen, no evidence of granuloma or malignancy. • Advised excision biopsy material for evaluation.

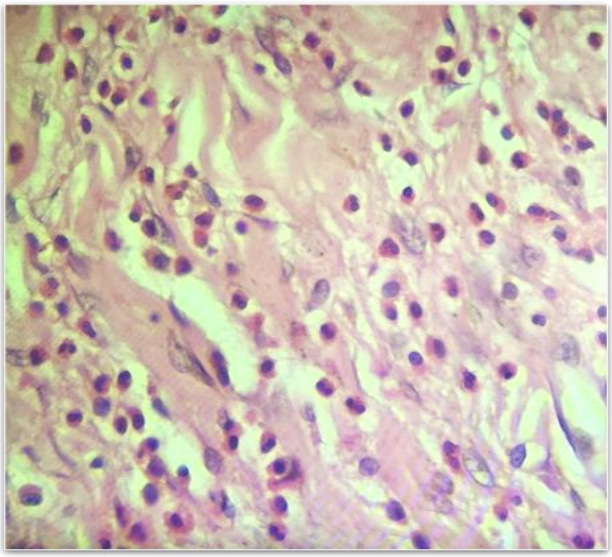

Figure 5: Section shows large number of eosinophils.

Excision Biopsy
• Gross appearance: Received a fibro fatty piece of tissue measuring 4.3 x 2.1 cm. Cut surface yellowish with a grey white area measuring 1.1 x 0.6 cm. Serial sections are similar, areas of haemorrhage identified. • Microscopic appearance: Shows muscle bundle and adipose tissue with blood vessels and nerve fibers with dense. – aggregates of lymphocytes having prominent germinal centres. – abundant eosinophils and plasma cells with proliferating blood vessels with foci showing prominent endothelial cells and few cells with prominent nucleoli. – stromal fibrosis and moderate population of eosinophils with collections of histiocytes. No evidence of granuloma or malignancy seen. • Lymphoid hyperplasia with eosinophilia

Clinical and histological features were suggestive of Kimura's disease. Patient was followed up and was advised to repeat RFT every 6 months, USG abdomen to rule out kidney involvement. Systematically administered steroids was prescribed to get good effect on disease progression.

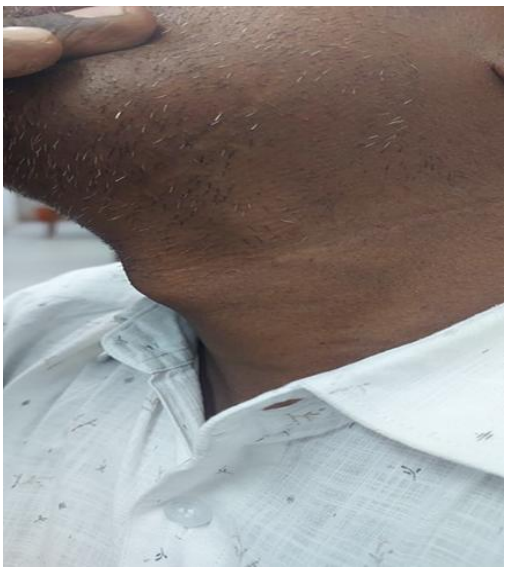

Figure 6: The clinical picture showing left in submandibular gland swelling.

Figure 7: The clinical picture showing swelling left temporal region.

Discussion

Kimura's disease named after Japanese doctor Kimura in 1948, who published rare chronic inflammatory disorder of unknown origin. The etiology of this rare disease is not clear but it is thought to be an immune mediated response triggered by unknown persistent antigenic stimulus. This disease is endemic in Asian males, commonly seen in Japan, China, Philippines occasionally in India. This disease commonly presents with painless soft subcutaneous swelling or lymphadenopathy in head and neck areas. The common lymph nodes involved are post auricular, cervical, inguinal, and epitrochlear. The commonly involved regions are periauricular, epicranium, orbits, eyelids, groin, salivary glands, soft tissues of the trunk, hard palate, larynx [3]. Laboratory findings include peripheral eosinophilia and elevated serum IgE levels. KD is rare in India and only 20 cases have been reported in India. Because of its rarity, accurate histopathological diagnosis is crucial for proper diagnosis and treatment.

KD diagnosis is made histopathologically, preferably from the swelling. In gross HPE multiple reddish white soft tissue are present on outer surface, whereas homogenous whitish color in cut section. Histologically Kimura disease presents as distorted architecture, hyperplasmic follicles, abundant eosinophils in the interfollicular area. Vascular proliferation with polymorphous population consists of many lymphocytes, plasma cells, histocytes and numerous eosinophils [4]. In a nutshell, histological triad of KD is characterized by painless subcutaneous nodules in the head and neck region, blood and tissue eosinophilia, markedly elevated serum IgE levels >1000 IU/ml.

KD should be kept in mind when maximal tumor or swelling diameter is >3cm, symptoms duration is > 5 years, peripheral eosinophil count >20 and serum IgE level >1000 IU/ml. We perused the literature before we made a diagnosis of Kimura disease in our case. About 60% cases show renal involvement (proteinuria and nephrotic syndrome).

Hui et al classified the histological features as constant, frequent and rare features (**Table 2**) [5].

Table 2: Constant, Frequent and rare features of KD

Constant Features
• Preserved nodal architecture • Florid germinal center hyperplasia • Eosinophilic infiltration and • Postcapillary venule proliferation.
Frequent Features
• Sclerosis • Polykaryocytes • Vascularization of the germinal centres • Proteinaceous deposits in the germinal centers • Necrosis of the germinal centres • Eosinophilic abscesses and • Reticular IgE deposition within germinal centers.
Rare Features
• Progressive transformation of the germinal centres. Nodal architecture is largely preserved in most cases, however, capsular fibrosis with subcapsular sinusoid obliteration and perinodal soft tissue involvement is frequently present.

The diagnosis of KD is not easy and differential diagnosis includes angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin's lymphoma, Kaposis Sarcoma, Eosinophilic granuloma, Epitheloid hemangioma, Castleman's disease, Dermatopathic lymphadenopathy, lymphadenopathy of drug reactions, parasitic lymphadenitis and many more (**Table 3**) [6,7]. The closest differential diagnosis is ALHE. Microscopically both show eosinophilic infiltrates and vascular proliferation. But there are few characteristic and distinctive clinicopathologic features that differentiate the two entities (**Table 2**). Differential diagnosis for KD involving parotid gland includes Mikulicz's disease, infective parotitis, salivary gland tumors, Sjogren's syndrome.

Table 3: Kimaru disease versus ALHE

	Kimura Disease	**ALHE**
Prevalence	Asians with male predominance	All races with slight female predominance
Eosinophilia and raised serum IgE	Common	Rare
Gross lesions	Solitary lesions are typically found in the deep subcutaneous tissues, commonly associated with regional lymphadenopathy and salivary gland involvement.	Superficial dermal papulonodules, small and frequently erythematous, accompanied by bleeding and pruritis. Regional lymphadenopathy is rare.
Histological lesions	That issue shows marked reactive follicular hyperplasia with prominent follicles, Accompanied by increased eosinophils, Lymphocytes and mast cells, occasional micro abscess formation and areas of vascular proliferation.	Marked vascular proliferation seen, where endothelial cells aggregate into lobule, which are line by plump cuboidal or hobnail-shaped cells.

Table 4: Cytological and histological features of other differential diagnosis.

Differential Diagnosis	**Cytological Features**	**Histological Features**
Angiolymphoid hyperplasia with eosinophilia (ALHE)	The spindle shaped and polygonal cells show vesicular nuclei and deeply eosinophilic cytoplasm rich in well defined vacuoles and in the immunoblasts.	Marked vascular proliferation seen, where endothelial cells aggregate into lobule, which are line by plump cuboidal or hobnail-shaped cells.
Hodgkin lymphoma	Atypical cells- Reed-Sternberg cells, eosinophils and plasma cells.	Positive diagnosis is determined by presence of Reed-Sternberg cells. Eosinophils, plasma cells and sclerosis are noted but lacks hyperplastic germinal centers and IgE deposits.
Castleman disease	Polytypic population, numerous plasma cells, increased number of lymphocytes and vascular proliferation may show changes	Follicular hyperplasia with abnormal well -formed germinal centers, vascular hyperplasia but lacks eosinophils and increased plasma cells in germinal centers.
Dermatopathic lymphadenopathy	Increased number of reactive lymphocytes, plasma cells large, activated macrophages which contain pigment.	Follicular hyperplasia, evident histiocytes in lymph node sinuses, macrophages contain melanin, hemosiderin.
Kaposi sarcoma	Atypical spindle cells	Slit-like vascular spaces filled with red blood cells, formed by proliferating endothelial cells.

Imaging studies play crucial role in diagnosis and can assist in staging and progression of Kimura's disease as well as in evaluation of lymph nodes [7]. The diagnosis in our case was clinically Hemangioma and histopathological examination of specimen revealed that it is Kimura's disease.

Treatment modalities for KD are: Surgical excision, Steroids therapy, Radiotherapy. Surgical excision is the first line of management [8]. Steroids can be given once diagnosis is made histopathologically. Radiation treatment is usually used for the smaller swellings and also for local control of recurrent lesions not responsive to steroids. In our case surgical excision was done. Recurrence rate in surgery alone is 25.65%-30.5%; Medication alone 35%-45%, Radiotherapy alone 50-60%. Lowest recurrence rate adjuvant therapy with surgery 26.9%. Kimura disease shows characteristic histologic features that helps in distinguishing the condition from hypersensitivity and drug reactions and infections [9]. In fact, Kimura's disease is at times missed diagnosed as malignant tumor that leads to unnecessary radicle surgery and the role of clinician's awareness becomes crucial here [10]. In this condition so far no malignant transformation has been reported but the recurrence rate post excision may be high [11]. It was further stated in another study that this disease exhibits distinct pathologic and clinical features [12].

Conclusion

Kimura's disease should be considered a potential diagnosis in patients presenting with head and neck masses and lymphadenopathy. Investigations should be carried out accordingly. Histopathology examination plays a very important role. Understanding Kimura's disease will enable clinicians to make accurate diagnosis and provide appropriate treatment to patients.

Declaration

Source of Funding

Nil

Conflict of Interest

There was no conflict of interest.

Consent

Informed consent has been taken from the patient.

Ethics Committee Clearance

IHEC clearance has been taken.

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