Case report

Kimura Disease - An unusual Presentation

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Abstract-A rare chronic inflammatory disorder with no known cause, Kimura disease (KD) is typically found in young Asian males. The condition is characterized by frequent enlargement of the salivary glands and a painless subcutaneous swelling in the head and neck region. Elevated immunoglobulin E (IgE) levels and eosinophilia in the blood and tissues are frequently associated. Eosinophilic infiltrates, follicular hyperplasia, and postcapillary venule proliferation are typical histological findings of biopsy specimens taken from subcutaneous swellings or lymph nodes. Even so, the course is typically benignly waxing and waning. A patient who receives an early diagnosis may avoid needless intrusive procedures. In this article, we describe a case of KD in a 40-year-old man who had two surgical procedures for subcutaneous edema in the left submandibular region before being reassessed and finally diagnosed as KD in 2022, based upon peripheral blood eosinophilia, raised serum IgE levels and histopathological findings. He subsequently made a good recovery on oral steroids.

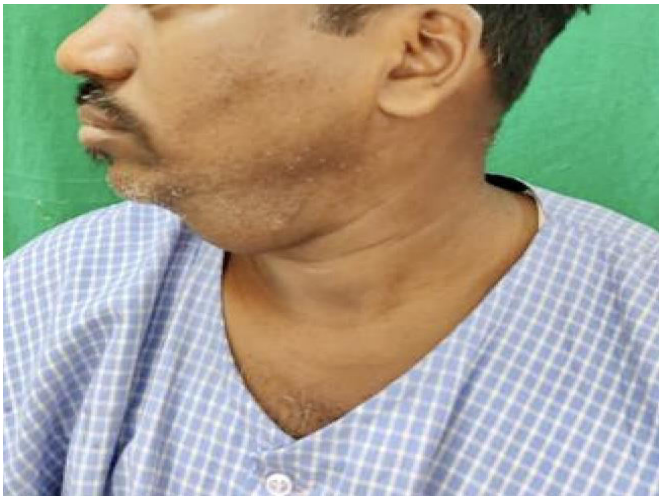
Keywords- Angiolymphoid hyperplasia with eosinophilia, immunoglobulin E, Kimura disease.

Introduction

Kim and Szeto originally described Kimura disease (KD), a rare chronic inflammatory ailment with no known cause, in 1937 in China. However, the disease was first referred to as KD when a Japanese doctor by the name of Kimura et al published a comprehensive analysis of the condition in 1948. [1,2] Only 200 cases of KD have been published in the literature globally since its first histological diagnosis, and the disease has rarely been recorded from India. [3] Although children may also be affected, it is typically found in young adults, with the majority of patients falling within the age range of 20 to 40 years. With a 3:1 ratio, men are impacted more frequently than women. [4,5] Although the condition is sporadic in other racial groups, it is endemic in Asians. Deep head subcutaneous tissue is generally affected by KD.Perivascular blood KD is consistently characterized by eosinophilia and high immunoglobulin E (IgE) levels. Renal disease is a prevalent kind of systemic involvement, with an incidence about 20% or such.[6,7] KD is frequently difficult to diagnose, necessitating a biopsy or removal of the affected mass or lymph node for a histopathological evaluation (HPE). [8] Due to overlapping clinical characteristics and similar histological (HPE) findings, it is sometimes mistaken for an illness known as angiolymphoid hyperplasia with eosinophilia (ALHE)

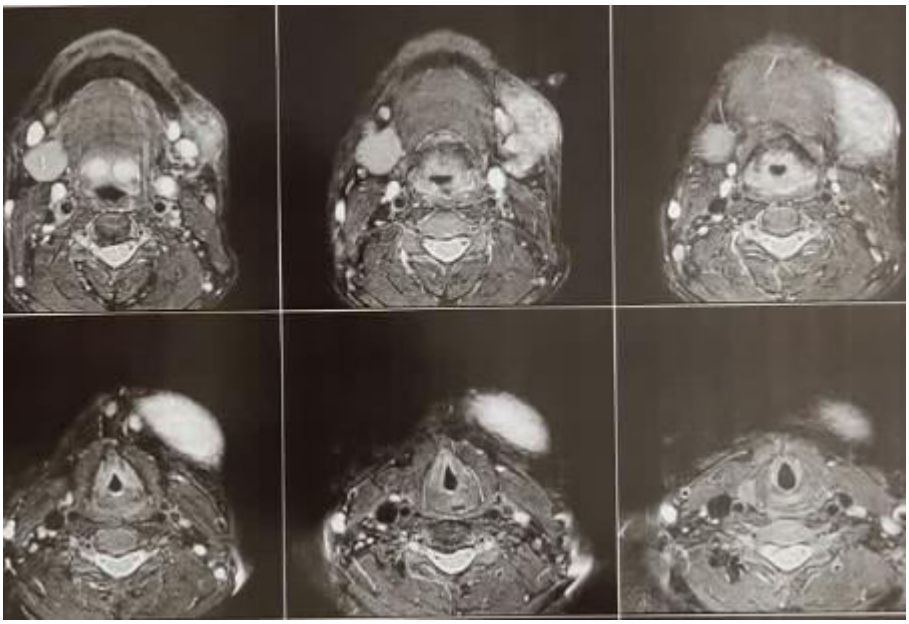
Case Report-

A 40 year-old male, presented to the surgeon in 2017, with complaints of a subcutaneous swelling in the left submandibular region of 1-year duration that was insidious at onset and gradually progressive in nature. A mass lesion measuring 4.3 cm × 2 cm × 2.7 cm with heterogeneous enlargement of left submandibular gland and raised vascularity of gland suggested on color doppler without involvement of skin or mucosa was revealed on an ultrasound (USG) scan. There were discrete enlarged bilateral submandibular and jugular nodes, without necrosis or calcification. (Majority were 0.8 - 1.2 cm in size)

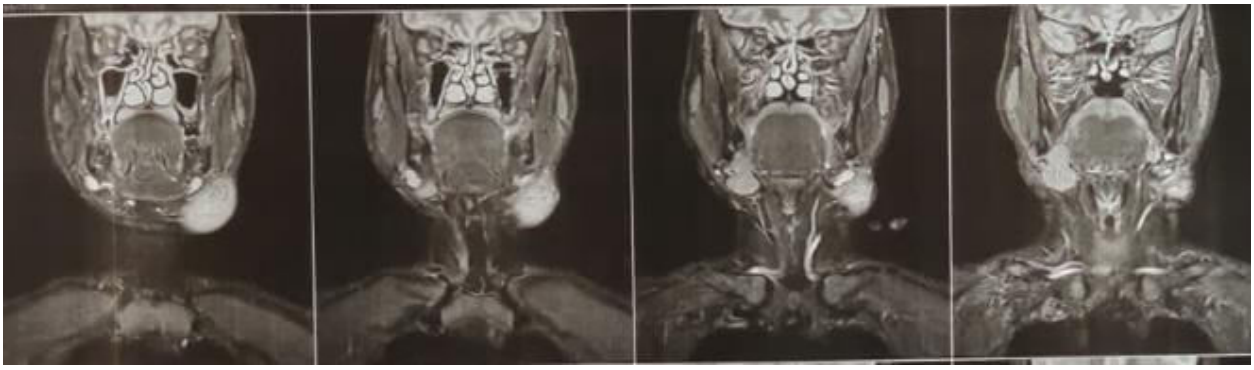


(Figure 1)

The same findings were confirmed on contrast-enhanced computed tomography (CECT) . Fine needle aspiration cytology (FNAC) from the swelling was suggestive of soft tissue lesion, likely fibroblastic in origin .A repeat HPE revealed a lesion with follicular hyperplasia,proliferation of vascular channels lined by plump endothelial cells and accompanying severe inflammatory infiltrate of eosinophils predominantly, consistent with a diagnosis of ALHE. He subsequently remained symptom free. In 2021, he again presented to the otorhinolaryngologist, with a 5 cm x 5 cm globular swelling(Figure 1) . MRI (face and neck) was again suggestive of recurrent/residual lesion involving both cheeks, in a known case of ALHE (postoperative).(Figure 2,3)



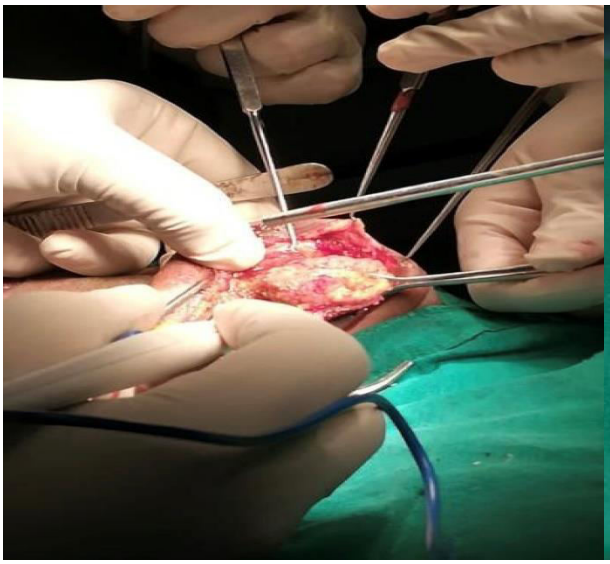
(Figure 2)



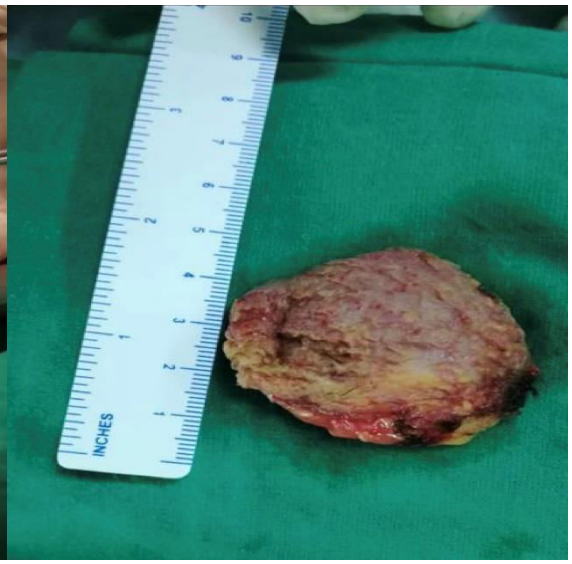
(Figure 3)

FNAC (right level II lymph node) showed polymorphous population of lymphoid cells composed of centroblasts, centrocytes, mature lymphoid cells and immunoblasts, along with clusters of histiocytes, with no atypical lymphoid cells, granulomas or giant cells. Based on previous history the recurrent nature of the swelling in the head and neck region causing cosmetic disfigurement, and substantiated by the HPE finding a diagnosis of KD was entertained (a close mimic of ALHE). He underwent excision of the swelling.(Figure 4,5) The patient was evaluated for serum IgE levels and absolute eosinophil count (AEC). AEC was 2900/cu mm. Peripheral blood smear was suggestive of eosinophilia. The HPE was also reviewed, and the diagnosis was revised to KD . Based on the above, the patient was referred to the Rheumatology OPD, where he was diagnosed as a case of KD. He was managed with oral steroids in the form of Tab prednisolone at a dose of 30 mg with regression in the size of the lump over the next 3 months. Repeat IgE levels at 3 months were 438.7 IU/L. Repeat

MRI showed absence of any significant residual mass.Presently, he is on tapering doses of steroids and received 4 intralesional triamcinolone steroid injections.



(Figure 4)



(Figure 5)

Discussion-

The manner that Kimura's sickness manifests in our patient is distinctive in a number of respects. He initially only had neck lymphadenopathy when presenting. Kimura's disease patients typically exhibit subcutaneous nodules in the head and neck region along with localized lymphadenopathy and salivary gland involvement [4]. Rarely does it manifest other than as a neck lymphadenopathy without subcutaneous skin or salivary gland involvement [9]. Second, our patient had a protracted chronic form of Kimura's disease. At his initial session, the patient was informed of the diagnosis following FNAC.It is well recognised that Kimura's disease is a benign and sluggish disorder. Before making his most recent presentation, our patient had the illness for six years.Third, Kimura's illness with normal serum IgE levels and peripheral eosinophil levels is extremely uncommon. The most reliable laboratory evidence for the diagnosis of Kimura illness has been peripheral eosinophilia and high blood IgE levels [5]. Only a few cases of Kimura's disease with normal peripheral eosinophil and/or serum IgE levels have been documented .[10] While some have reported the swelling as lymphadenitis or malignancies , some authors were able to diagnose Kimura's disease on FNAC.The appearance of substantial numbers of eosinophils on FNAC, along with collagenous tissue fragments, endothelial cells, and sporadic polykaryocytes, are the hallmarks of Kimura's illness .

Kimura's illness is characterized histopathologically by lymphoid tissue with germinal center hyperplasia and hyperplastic vascular structures surrounding it. The germinal centers of lymphoid follicles vary in size and include varying degrees of eosinophilic infiltrates as well as focal eosinophilic microabscesses. Additionally seen are pronounced fibrosis and vascular hyperplasia. There are no unusual cells found. Patients with Kimura's illness have been noted to

have renal damage, with incidence rates ranging from 10 to 60%. In 10-12% of these patients, nephrotic syndrome was observed; other symptoms included membranous glomerulonephritis, minimal change glomerulonephritis, diffuse proliferative glomerulonephritis, and mesangial glomerulonephritis [7].There is still debate over the ideal course of treatment for Kimura's disease [8, 9]. The best treatment option has historically been surgery. A combination of surgery, postoperative systemic corticosteroids, and low-dose radiotherapy have been suggested in cases where the disease affects the subcutaneous tissue without well-defined margins as a recurrence rate of up to 62% has been reported in these cases with surgery alone [10].

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