Case Report

Kimura disease: A rare cause of painless lymphadenopathy in South Asia

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Abstract

Kimura disease is an uncommon disease entity that typically involves the lymph nodes predominantly in the head and neck region together with frequent involvement of salivary glands. Very few cases of it have been reported in literature globally, and in the context of India, it is even rare. Early suspicion of Kimura disease may prevent the patient from unnecessary invasive diagnostic tests. We present a case scenario of a 35 years old female, from a hilly area who presented with painless neck swelling for 3 months that was followed by fever, new onset pain at the site of neck swelling, and skin rashes. Diagnosis of Kimura disease was made based on histopathological findings aided by peripheral eosinophilia and elevated serum Immunoglobulin E (IgE) levels. Following the diagnosis, the patient was treated with a short course of oral steroids which produced an excellent response with a consequent decrease in the size of lymph nodes and resolution of the skin rashes.

Keywords: Kimura, lymphadenopathy, painless

Introduction

Kimura disease is a rare chronic inflammatory condition of unknown etiology characterized by painless lymphadenopathy with predominant involvement of the head and neck region, with frequent involvement of the salivary glands.[1,2] This disease entity is rare in India with only 200 cases reported in the world after histopathological diagnosis.[3] Kimura disease was first described by Kim and Szeto in the Chinese literature as "Eosinophilic hyperplastic lymphogranuloma" and has been renamed as Kimura`s disease later on after being published in literature by in 1948. Common age of presentation is between 20 and 40 years with the male preponderance of 3:1[4] Peripheral eosinophilia and serum Immunoglobulin (IgE) levels aid in the diagnosis however confirmatory diagnosis can be made only on histopathology. It is assumed to be an autoimmune reaction to antigenic stimulus for which the treatment is based on anti-inflammatory drugs mainly steroids. Multiple organ involvement has been seen with renal involvement being the deadly one. This case report will make the doctors aware of the consideration of this illness in the differential diagnosis of lymphadenopathy in young adults. High clinical suspicion and early diagnosis prevent patients from getting unnecessary investigations and also aid in the early recovery of the patient.

Case Summary

A 35 years female from a middle-class family in a remote village of Uttarakhand with no addictions or comorbidities, P4L4, presented with insidious onset, gradually progressive, painless neck swellings (Right > left) for 3 months. She developed high-grade fever with chills and rigor for 10 days associated with the burning sensation in the swelling. The fever subsided with over-the-counter medications. She did not have a history of sore

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throat, cough, chest pain, pain abdomen, dysuria, per vaginal discharge, or altered bowel habits. She developed acute onset pruritic erythematous and maculopapular rashes distributed all over the body for 5 days, not associated with any drug ingestion, exposure to new objects or environment, or jaundice. The pruritus was so intense that there were excoriation marks in different parts of the body that were readily appreciable.

She was admitted from the medicine outpatient department (OPD) for the evaluation of lymphadenopathy with rashes. Her vitals at the presentation were normal except for her temperature which was 101°F. On examination, she had multiple cervical lymph nodes in the cervical area levels Ia, Ib, II, III, IV, and post-auricular nodes, the largest measuring 3*3 cm at level II, non-tender, firm, non-matted, and not fixed to the underlying tissue. [Figure 1] She also had palpable bilateral inguinal lymph nodes of similar consistency and relatively smaller size; however, no genital ulcers were evident. There were bilateral anterior axillary lymph nodes palpable with the size of 2*2 cm. She had erythematous and maculopapular rashes dispersed all over the body, predominantly over the bilateral upper and lower extremities, including the facial area, while the anterior abdomen was relatively less involved [Figure 1].

Baseline blood investigations revealed leucocytosis with eosinophilia. Serum IgE levels were elevated. However, kidney function was normal [Table 1]. A stool routine examination did not reveal any parasite and Imaging of the chest was also normal. She was initially started on oral antihistaminic after a dermatology consultation. Empirically oral Azithromycin was started considering the likelihood of reactive lymphadenitis. Over the course of treatment, pain in the swelling subsided but there was only minimal decrement in the size of lymph nodes. The rashes settled over the course of 7 days of antihistaminic and the fever also subsided. A lymph node biopsy was done as a part of the etiological workup, and the patient was discharged on oral antihistaminic and followed with a biopsy report in 10 days. The patient reported back in OPD with a biopsy report that was suggestive of Kimura disease [Figure 2].

After an opinion from hematology, prednisolone was started in an oral dose of 30 mg per day for 10 days and tapered over the next 2 weeks. Deworming with Ivermectin was done before starting the prednisolone. On follow up the patient was examined for lymph nodes, where the lymph nodes were barely palpable [Figure 3] and the rashes were almost absent.

Discussion

Kimura disease is a rare disease entity, whose etiology is still not clear. However, it has been postulated that the disease occurs due to the autoimmune response mounted against unknown antigenic stimulus which preferentially stimulates the TH2 cells with resultant production of cytokines like macrophage-colony stimulating factor, tumor necrosis factor (TNF) alpha, interleukin 5 (IL-5), interleukin 6 (IL-6), eotaxin, and regulated upon activation normal T cell expressed and secreted (RANKES). Clonal T-cell proliferation leads to disease occurrence as well as recurrence.[5]

The clinical course is usually benign and self-limited. It can involve organs other than lymph nodes like orbit, eyelid, lacrimal gland, and even kidney. Up to 60% of the patients exhibit renal involvement in the form of extramembranous glomerulonephritis and nephrotic syndrome. Renal involvement is a warning sign.[6] Our case did not show any evidence of renal involvement.

The histological features of Kimura`s disease have been divided into constant, frequent, and rare by Hui *et al.* The constant features include preserved nodal architecture, florid germinal center hyperplasia, eosinophilic infiltration, and post-capillary venule proliferation. The nodular picture is largely preserved with capsular fibrosis, subcapsular sinusoidal obliteration, and perinodal soft tissues being frequently involved.[7]

The nearest differential of Kimura`s disease is angiolymphoid hyperplasia with eosinophilia (ALHE) with a very small margin of differentiation between the two. Kimura`s disease occurs predominantly in males and Asians with a male predilection. Peripheral eosinophilia and elevated IgE levels are more in favor of Kimura`s disease while ALHE occurs in all racial groups.[8] In

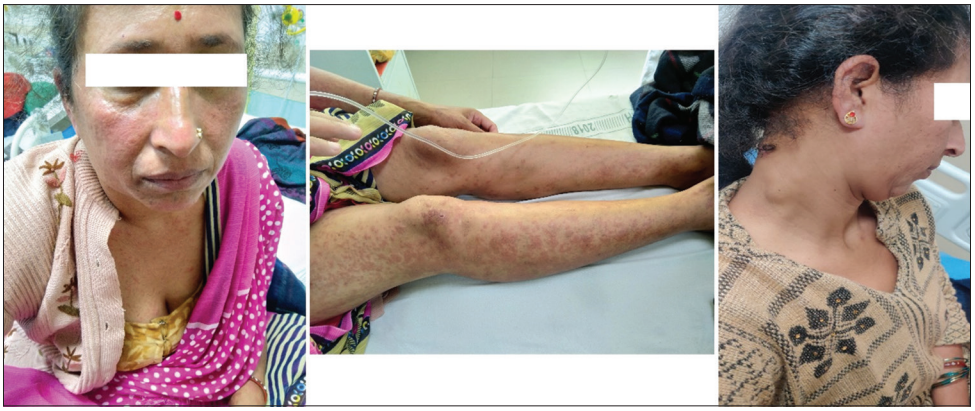

Figure 1: Rashes and cervical lymphadenopathy at the time of presentation (Pre-treatment)

Table 1: Investigations

Investigations	15/11	16/11	20/11	21/11	22/11
Hemoglobin	12.6	13.6	12.3	10.9	11.6
Total leucocyte count	9*1000	15.46*1000	18.54*1000	11310	12.1*1000
Platelets	251000	261000	190000	165000	157000
N/L/M/E	35/11/8/46	35/17/13/35	60/14/4/22	55/21/4/20	71/10/18
PT-INR		12.5/1.15			
Bilirubin (T)	0.79	0.99			
Bilirubin (D)	0.22	0.28			
SGPT	66	42			
SGOT	65	36			
ALP	332	144			
S. Protein	5.9	6.5			
S. Albumin	3.4	3.3			
Blood Urea	17		17		
S. Creatinine	0.79		0.6		
S. Sodium	135		136		
S. Potassium	4		3.9		
S. Calcium	8.5		8.3		
S. Uric acid	4.2				
S. Phosphorus	2.4		2.9		
HIV Ab/HBsAg/Anti-HCV Ab	Non-Reactive				

Special Investigations:
USG Abdomen: Hepatosplenomegaly (Liver=16.5 cm, Spleen=14.5 cm). No free fluid present.
Serum IgE=602.3 (normal=0-300 IU/ml)
20/11: Leptospira IgM/IgG: Negative
Scrub Typhus IgM: Negative
22/11: Blood culture: No growth
16/11: Procalcitionin: 0.16
URINE Routine Examination: Within Normal Limits
CRP: 41ng/ml
URINE CULTURE: No Growth
USG ABDOMEN: Bulky Uterus
18/11: Absolute Eosinophil Count: 1612
APTT: 25.2s

SGPT=Serum Glutamate Pyuruvate Transaminase, SGOT=Serum Glutamatic Oxaloacetate Transaminase, ALP=Alkaline Phosphatase, PT/INR=Prothrombin Time/International Normalized Ratio

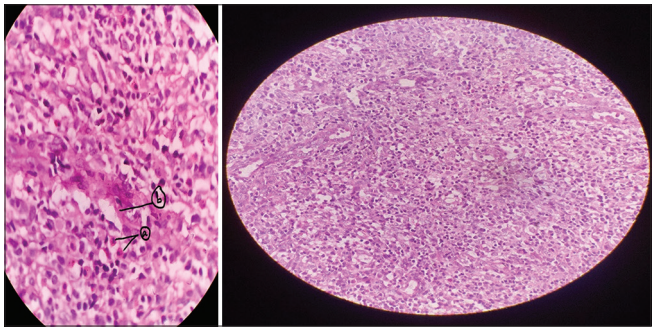

Figure 2: Histopathological Examination. (i) 40× magnification. (a) Increased eosinophils admixed with inflammatory cells (lymphocytes and occasional histiocytes). (b) There is also the prominence of plump endothelial cells. (ii) 20× magnification. Image showing preserved nodal architecture

Figure 3: Post-treatment resolution of rashes

contrast to Kimura`s disease, ALHE shows prominent vascular proliferation, forming aggregates or plumps with epithelioid and histiocytoid changes.[9]

Regarding the treatment, oral steroids remain the first-line treatment, which at times may need to be upgraded to a higher immunosuppressant depending upon the response. These drugs suppress the disease activity by inhibiting inflammatory responses. In our case, a short course of steroid therapy has provided a sounding response over a short period of time.

Conclusion

Kimura disease though rare is still an enigma of medical science. The relevance of this case is due to the rarity of disease which mimics neoplastic conditions. Hence, it should be put in differential diagnoses of the patient presenting with painless cervical lymphadenopathy and peripheral eosinophilia. Knowledge of this disease will put physicians in a better condition to diagnose and treat it.

Consent

Informed consent has been taken from the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Ioachim HL, Medeiros LJ, editors. Ioachim's Lymph Node Pathology. Lippincott Williams & Wilkins; 2009.
2. Abhange RS, Jadhav RP, Jain NK. Kimura disease: A rare case report. Anl Pathol Lab Med 2018;5:C53-5.
3. Rajesh A, Prasanth T, Sirisha VN, Azmi MD. Kimura's disease: A case presentation of postauricular swelling. Niger J Clin Pract 2016;19:827-30.
4. Tseng CF, Lin HC, Huang SC, Su CY. Kimura's disease presenting as bilateral parotid masses. Eur Arch Otorhino Laryngol Head Neck 2005;262:8-10.
5. Meningaud JP, Pitak-Arnnop P, Fouret P, Bertrand JC. Kimura's disease of the parotid region: Report of 2 cases and review of the literature. J Oral Maxillofac Surg 2007;65:134-40.
6. Abhay H, Swapna S, Darshan T, Vishal J, Gautam P. Kimura's disease: A rare cause of local lymphadenopathy. Int J Sci Stud 2014;2:122-5.
7. Chen H, Thompson LD, Aguilera NS, Abbondanzo SL. Kimura disease: A clinicopathologic study of 21 cases. Am J Surg Pathol 2004;28:505-13.
8. Vimal S, Panicker NK, Soraisham P, Chandanwale SS. Kimura disease. Med J Dr. DY Patil Univ 2013;6:208.
9. Swarnkar M, Agrawal A. Kimura's disease. Formos J Surg 2018;51:26.