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A Rare Case of Kimura Disease with Pulmonary Involvement

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Abstract
Kimura disease (KD) is a rare chronic inflammatory disorder of unknown etiology, typically characterized by painless subcutaneous nodules or masses in the head and neck region, regional lymphadenopathy, and eosinophilia. The disease predominantly affects young males from East Asia. This case report describes an unusual presentation of KD in a 36-year-old male with initial symptoms of dry cough, weight loss, and peripheral neuropathy, followed by the development of subcutaneous nodules in the right hand and right axillary lymphadenopathy. This case highlights the diverse clinical manifestations of KD and emphasizes the importance of considering this diagnosis in patients with atypical presentations.

Keywords: *Kimura Disease, Pulmonary Involvement*

Introduction

Kimura disease is a rare inflammatory disorder primarily affecting young Asian males [1]. The classic presentation involves cervical or preauricular lymphadenopathy, often accompanied by subcutaneous nodules in the head and neck region [1]. Epidemiologically, KD is most frequently reported in Japan, China, and other parts of Asia, with an estimated incidence of 0.013 per 100,000 individuals in endemic regions. Cases are rare in Western populations, leading to under recognition outside Asia [3]. From a community health perspective, KD underscores the importance of including rare but regionally relevant diseases in differential diagnoses, especially in areas with high prevalence of eosinophilic disorders [1].

While eosinophilia and elevated serum IgE levels are common laboratory findings, atypical presentations can pose diagnostic challenges [3]. This report presents a unique case of KD with initial symptoms mimicking respiratory and neurological conditions, later accompanied by subcutaneous nodules.

Case presentation

A 36-year-old previously healthy Sri Lankan male from Ampara, Sri Lanka presented with a one-month history of dry cough, unintentional weight loss of approximately 5 kg over five months with preserved appetite, and a one-year history of intermittent numbness and burning sensations in his hands and feet, which had recently worsened. At the time of presentation, he had a burning sensation of almost the whole body.

The cough was predominantly dry, nocturnal, and associated with mild wheezing. He denied sputum production, hemoptysis, and shortness of breath, fever, night sweats, or loss of appetite. He had no history of

recent foreign travel and high-risk sexual behaviors. He denies significant environmental exposures to chemicals, biomass fuel, animals, birds, paddy, paint, silica, asbestose and gases or contact with tuberculosis.

Other than the burning sensation and numbness there were no other complaints related to the central or peripheral nervous system. Notably he had no weakness, loss of consciousness, headache, nausea, vomiting or visual disturbances.

History was unremarkable for connective tissue disorder or vasculitis associated symptoms such as small and large joint arthritis, thickening of the skin, photo sensitive rashes or other skin rashes, red eyes and hair loss. His urine output and bowel habits were normal.

Nearly 2 weeks after the initial presentation, the patient developed subcutaneous nodules in the right forearm with some right axillary lymph nodes enlargement. This progressively increasing in size, itchy and not painful. Patient recalled an event of enlargement of the right-side submandibular area swelling which was transient and disappeared in three days couldn't observe such swelling during the follow up clinic visit.

His past surgical history was significant only for renal stone removal in 2018 with a unremarkable recovery. He had a 0.5 pack-year history of cigarette smoking with a strong willingness to stop smoking and occasional alcohol consumption. He does not have any allergies. He did not have any past medical illnesses or childhood diseases. His family history was unremarkable for asthma, autoimmune diseases, lung cancer, or tuberculosis. He is a father of two children with very good family support and it is a middle-income family. He worked as a recovery officer at a people's leasing company and was the primary provider for his family.

Physical examination

On physical examination, he was afebrile and not pale. A single enlarged, non-tender right axillary lymph node was noted. Two weeks after initial presentation, during a follow-up visit, multiple small, non-tender, mobile subcutaneous nodules, with one larger dominant nodule, were discovered on his right hand. No similar nodules were found elsewhere on his body.

Respiratory examination revealed occasional coarse and fine crepitation in bilateral lower zones of the lung, with equal air entry. Cardiovascular and abdominal examinations were normal. Neurological examination revealed altered sensations in his hands and feet, consistent with peripheral neuropathy, but no obvious motor weakness.

Investigations
Biochemical examination

His full blood count revealed high eosinophilic count.

Table 1: Full blood count of the patient

Test	Result		Flag	Reference Value
Total White Cell Count	10.35	10^9/L		4.00 - 11.0
Neutrophils	33.3	% 3.5	L	40-80% (2.0-7.0)
Lymphocytes	24.5	% 2.5		20-40% (1.0-3.0)
Monocytes	2.8	% 0.3		02-10% (0.2-1.0)
Eosinophils	38.9	% 4.0	H	01-06% (0.02-0.5)
Basophils	0.5	% 0.1		<0.1-02% (0.02-0.1)

Haemoglobin	14.7	g/dL		13.0-17.0
Red Blood Cells	4.79	10^12/L		4.5-5.5
Mean Cell Volume	90.6	Fl		76.0-96.0
Haematocrit	43.4	L/L		40.0-47.0
Mean Cell Haemoglobin	30.6	Pg		27.0-33.0
M.C.H. Concentration	33.8	g/dL		32.0-36.0
Red Cells Distribution Width	12.9			12.0-15.0
Platelet Count	254	10^9/L		150-400

Renal function tests including electrolytes, blood urea and serum creatinine were found to be normal.

Table 2: Renal function tests of the patient

Test	Result		Flag	Reference Value
Blood Urea	13.2	mg/dL		12.8-42.8
Serum Creatinine (Enzymatic)	0.91	mg/dL		0.7-1.2
Serum Sodium	136	mmol/L		136.0-146.0
Serum Potassium	4.10	mmol/L		3.50-5.1
Serum-Chloride	101	mmol/L		101.0-109.0
Serum Total - Calcium	2.20	mmol/L		2.20-2.65
Serum Inorganic Phosphorus	4.2	mg/dL		2.7-4.5
Serum Uric Acid	6.2	mg/dL		3.5-7.2
Blood Urea Nitrogen	6.1	mg/dL		6.0-20.0

Table 3: Lipid profile

Test	Result (mg/dL)	Flag	Reference Value (mg/dL)
Total Cholesterol	271.8	H	140.0 - 239.0
Triglycerides	108.4		10.0 - 200.0
HDL Cholesterol	63.0		35.0 - 85.0
Non-HDL Cholesterol	208.8	H	55.0 - 189.0
LDL Cholesterol	187.1	H	40.0 - 159.0
VLDL Cholesterol	21.6		10.0 - 41.0
Chol/HDL Ratio	4.3		2.0 - 5.0
LDL/HDL Ratio	2.97		0.01 - 3.31

Table 4: Other investigations

ESR	13mm/hr
CRP	1.56g/dl
TSH	1.676micrIU/ml
Filarial antigen Filarial antibody	Negative Positive
HBA1C	5.9%
Total IgE Aspergillous specific IgE	683IU/ml Moderately positive
PANCA/CANCA	Negative
LDH	300U/L

Blood picture: His total white cell differential counts show *moderate eosinophilia* with some reactive cells. Red cells and platelets are normal in number and morphology.

Radiological Examination

Chest X-ray: Shows Bilateral lower zone reticular shadow (Figure 1)

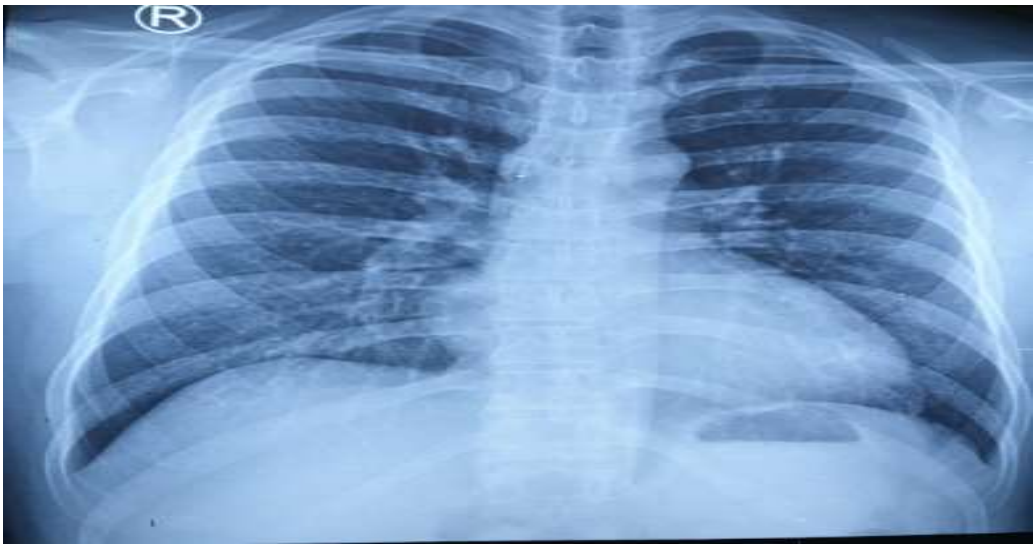

Figure 1

CT scan: To evaluate the cause of the cough and assess for any lung parenchymal involvement or mediastinal lymphadenopathy a CT scan was carried out. Found to have patchy small ground glass density nodules with mild bronchiectasis in the bilateral lower lobe. The reporting by radiologist suggested that changes could be due to eosinophilic lung disease. ABPA was considered as a differential diagnosis. Enlarged right axillary lymph node with right pectoraris minor group lymph nodes was noticed, and USS scan guided biopsy was planned.

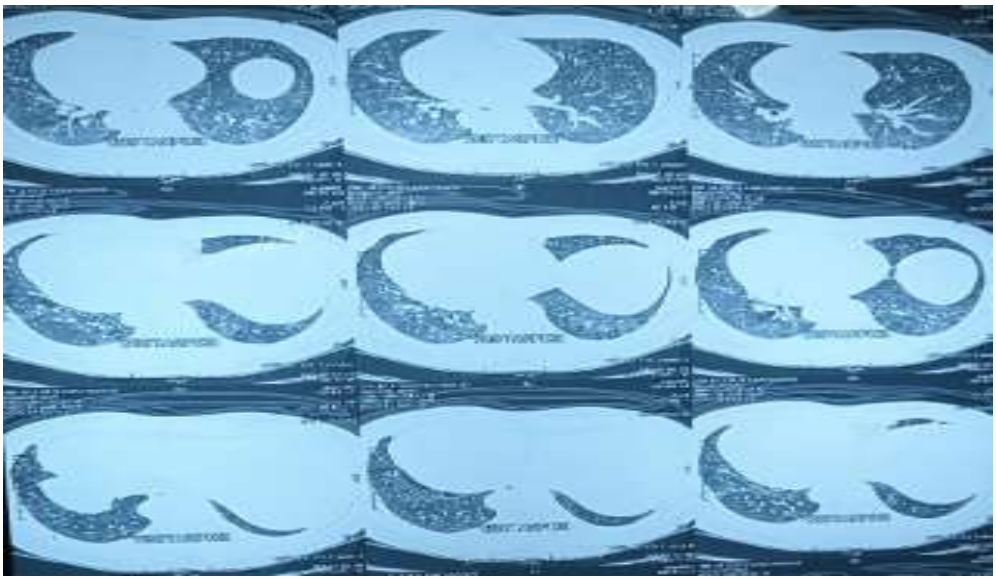

Figure 2

USS abdomen: Grade 1 fatty liver. The rest of the study is normal.

Imaging of the affected areas (e.g., ultrasound of the hand): To further characterize the subcutaneous nodules a USS of the affected area was carried out which showed multiple lymph nodes in the right axillary region. Largest node measures approximately 1.2 x 2.4 cm. Right mid arm 2.4x0.7cm size linear hypo echogenic area with high vascularity in the subcutaneous tissue was found. The lymph nodes are described as "reactive," suggesting an inflammatory process. A biopsy of the lymph node was planned.

Special Examinations.

Nerve conduction studies: The study to objectively assess the peripheral neuropathy. He had moderate bilateral carpel tunnel syndrome. No evidence of large fiber neuropathy. Small fiber neuropathy cannot be excluded with this study

2D echo was normal

Bone marrow trephine biopsy: The study concludes a normal cellular marrow. No evidence of bone marrow infiltration by haematological and non haematological malignancies.

Bronchoscopy: bilateral bronchial tree was inflamed, edematous and had thin secretions. Gobble stone appearance was noted. Broncheoalveolar lavage TB culture, TB gene xpert was negative. BAL cytology: Smear shows scattered alveolar macrophages, benign bronchial epithelial cells and metaplastic squamous cells. The background shows lymphocytes. Malignant cells are not seen. Gram stain and culture showed no growth. Fungal studies: No fungal elements seen and no growth.

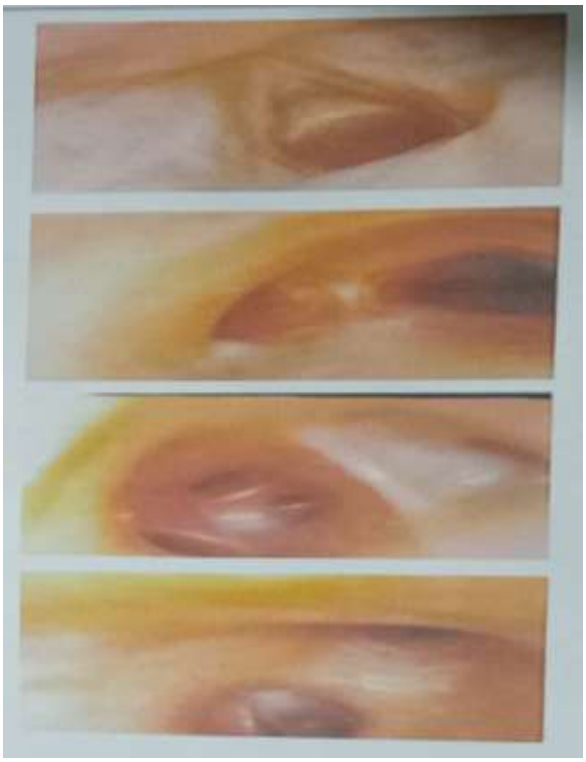

Figure 3

Right axillary lymph node biopsy

The microscopy reveals a lymph node biopsy with preserved architecture. There is an abundance of reactive lymphoid follicles, some of which are enlarged, showing polarization of the germinal centers. The areas between the follicles are widened and contain numerous eosinophils, forming small clusters known as eosinophilic microabscesses. Scattered plasma cells are also present. Importantly, no Reed-Sternberg cells or

Hodgkin cells are observed, and there is no evidence of suppuration or granulomatous inflammation. No abnormal lymphoid cell proliferation is seen.

Biopsy report consistent with florid reactive lymphoid follicular hyperplasia with eosinophilia and suggested immune chemical assessment.

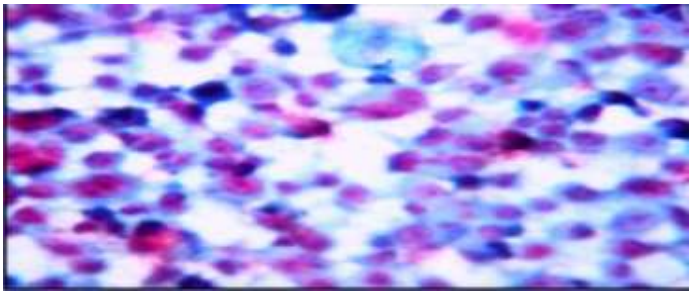

Figure 4

Right forearm lump excision biopsy

The section shows lymph nodal tissue with preserved architecture with scattered lymphoid follicles of various sizes. Germinal centers and mantle zones are well defined. There are singly scattered eosinophils and eosinophilic abscess formation.

The differential diagnoses include Kimura disease, hyper eosinophilic syndrome, and parasitic infestation was considered.

The report suggests a biopsy from a different lymph node group to exclude other possibilities like lymphoma.

Differential Diagnosis:

The initial presentation raised suspicion for various conditions, including:

- Infections (e.g., tuberculosis, fungal infections): Given the cough and weight loss. Broncho alveolar lavage did not find any infection.

- Connective tissue diseases (e.g., vasculitis): Due to neuropathy and potential for lymphadenopathy. He did not have symptoms suggestive of connective tissue disease such as raynouds phenomenon, rahses, joint pain or hair loss.

- Malignancies:

  Hodgkin's Lymphoma: While less likely with his age, Hodgkin's lymphoma can present with eosinophilia and lymphadenopathy.

  T-cell Lymphomas: Some T-cell lymphomas can have eosinophilic infiltration. Considering lymphadenopathy. But bone marrow biopsy was normal.

- Tropical pulmonary eosinophilia: Filarial antigen was negative still the filarial antibody became positive, and this condition was also taken as a possibility and treated him with antifilarial medication.

- Other eosinophilic lung diseases (eg: churg strauss): we could not find any granulomatous inflammation in the subcutaneous nodule biopsy.

- Hyper eosinophilic syndromes: This is a group of disorders characterized by persistent eosinophilia without an identifiable cause.

- Drug Reactions: Certain medications can cause drug-induced eosinophilia and systemic symptoms.
- Allergic Diseases: Although less likely in this case due to the lack of specific allergic triggers, severe allergic reactions can present with eosinophilia and systemic symptoms.

Further Investigations to Narrow Down Diagnosis:

The following investigations were considered to narrow down the diagnosis:

- Histopathological examination of the lymph node or subcutaneous nodule: This is crucial for confirming Kimura disease or other conditions.
- Serum IgE levels: Elevated IgE levels are often seen in Kimura disease.
- Chest CT scan: To evaluate for mediastinal lymphadenopathy or other lung abnormalities.
- Ultrasound of the abdomen: To assess organ involvement.
- Stool examination for ova and cysts: To rule out parasitic infections.
- Filarial antigen tests: If parasitic infections are suspected.
- Autoimmune serology: To rule out autoimmune conditions.

6. Management

Patients started on oral prednisolone 40mg daily for three months and tapered over 5 weeks. His blood sugar levels were monitored using a home glucometer. Subcutaneous nodules disappeared 2 weeks after starting the treatment. His cough resolved and weight became static.

Itraconazole:

The patient was started on itraconazole for three months due to a positive skin prick test for Aspergillus and moderately elevated Aspergillus-specific IgE. Itraconazole is an antifungal medication, and its use in KD is not standard and requires further justification.

Diethylcarbamazine (DEC):

Diethylcarbamazine (DEC) was administered for three months as the filarial antibody test was positive. DEC is an anti-parasitic medication primarily used for filariasis.

Considering Other KD Treatments:

After the diagnosis of KD is confirmed, other treatment options were considered, especially if the patient doesn't respond well to corticosteroids or has recurrent disease. These include:

- Immunomodulators (e.g., cyclosporine, tacrolimus): For more severe or refractory cases.
- Surgery: The preferred initial treatment localized kimura disease is surgical excision.
- Other treatment: Surgical exicion with post-operative law dose radiotherapy, radiation therapy alone.

Monitoring Response:

Closely monitor the patient's response to treatment, including symptom improvement, changes in lymph node size, and repeat blood tests (especially eosinophil count).

Discussion

Kimura disease (KD) is a rare, chronic inflammatory disorder most commonly seen in young Asian males and is characterized by painless subcutaneous nodules, regional lymphadenopathy, peripheral eosinophilia, and elevated serum IgE levels. Despite its benign course, KD can pose significant diagnostic challenges, particularly when it presents with atypical features [1]. In this case, a 36-year-old male initially presented with symptoms not classically associated with KD—dry cough, weight loss, and peripheral neuropathy—followed later by the development of subcutaneous nodules and axillary lymphadenopathy, which eventually guided the diagnostic trajectory toward KD. While cervical and preauricular lymphadenopathy are the most common sites of involvement, the presence of right axillary lymphadenopathy, subcutaneous nodules in the right hand, and peripheral neurological symptoms broaden the clinical spectrum, emphasizing the diagnostic complexity in this case.

The markedly elevated eosinophil count is a key laboratory finding that strongly supports the diagnosis of KD, as eosinophilia is a hallmark feature of this disease. In addition, the detection of enlarged, non-tender axillary lymph nodes—described on imaging as "reactive"—and the recurrence of nodal swelling at a prior biopsy site reinforce the inflammatory nature of the disease [1,3]. Subcutaneous nodules, particularly those that are mobile and non-tender, are also consistent with KD and aid in its differentiation from neoplastic or infectious etiologies [4]. However, the patient's initial systemic symptoms and neurological findings prompted consideration of a broad range of differential diagnoses, including parasitic infections, connective tissue disorders, lymphoma, and vasculitis.

Although peripheral neuropathy is rarely reported in KD, it has been described in select cases and could be related to eosinophil-mediated nerve injury or coexisting immune dysregulation [1,3,5]. This finding further complicates the clinical picture and highlights the need for thorough neurological evaluation in KD patients presenting with atypical symptoms. The cornerstone of diagnosis remains histopathological confirmation via biopsy of an involved lymph node or subcutaneous nodule. Characteristic histological features include preserved lymphoid architecture with follicular hyperplasia, dense eosinophilic infiltration, eosinophilic microabscesses, and proliferation of postcapillary venules [4].

In terms of management, corticosteroids remain the first-line therapy for KD. The patient was appropriately started on prednisolone, which is effective in reducing lesion size and controlling eosinophilic inflammation [4,5,6,7]. However, the empirical use of antifungal (itraconazole) and antiparasitic (diethylcarbamazine) agents requires further justification, as there was no documented clinical or microbiological evidence of fungal or parasitic infection [7]. This underscores the importance of evidence-based prescribing and careful exclusion of mimicking conditions through targeted investigations. If corticosteroid therapy fails or if the disease relapses, alternative treatment options such as immunosuppressants (e.g., cyclosporine, azathioprine), radiation therapy, or surgical excision may be considered [7,8].

Conclusion

In conclusion, this case illustrates the diverse and occasionally deceptive clinical presentation of Kimura disease. It emphasizes the value of maintaining a high index of suspicion for KD in patients with eosinophilia and nodal/subcutaneous swellings, even in the presence of atypical symptoms such as respiratory and neurological involvement. A definitive diagnosis should be secured via biopsy, and treatment must be guided by clinical response and histopathological confirmation. A multidisciplinary approach—including dermatology, pathology, hematology, neurology, and infectious disease expertise—is essential for accurate diagnosis and comprehensive care. Long-term follow-up is recommended to monitor for recurrence and manage chronic inflammatory sequelae.

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