# Kikuchi-Fujimoto disease: A case report

Shekhar Gurung[1], Ribek Pariyar[1], Saurab Karki[2], Anu Gautam[3], and Hari Sapkota[4]

[1]Shivanagar Primary Health Care Center
[2]Nepalese Army Institute of Health Sciences College of Medicine
[3]Nepalese Army Institute of Health Sciences
[4]College of Medical Sciences Teaching Hospital

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**Abstract**

Kikuchi disease is an uncommon, self-limited disease that mimics malignant lymphoma and tubercular lymphadenitis in presentation but with an excellent prognosis. We present a case of a 20-year-old Asian female with swelling at the angle of the mandible along with fever as a result of Kikuchi disease.

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Shekhar Gurung[1], Ribek Sunam Pariyar[1], Saurab Karki[2], Anu Gautam[3], Hari Sapkota[4]

[1]Shivnagar Primary Health Care Center, Chitwan, Nepal

[2]Military Hospital Itahari, Sunsari, Nepal

[3]Nepalese Army Institute of Health Science, Kathmandu, Nepal

[4]College of Medical Sciences, Chitwan, Nepal

**Correspondence:** Saurab Karki, Military Hospital, Itahari-4 Sunsari, Nepal. Email: *saurabkarki1010@gmail.com,* Phone: +977-9841098336.

## ABSTRACT

Kikuchi disease is an uncommon, self-limited disease that mimics malignant lymphoma and tubercular lymphadenitis in presentation but with an excellent prognosis. We present a case of a 20-year-old Asian female with swelling at the angle of the mandible along with fever as a result of Kikuchi disease.

## KEYWORDS

Kikuchi Fujimoto disease, lymphoma, tubercular lymphadenopathy, case report

## 1 INTRODUCTION

Kikuchi disease also known as Kikuchi -Fujimoto disease is a rare but benign condition that presents commonly with cervical lymphadenopathy and fever. It can be further accompanied by a rash, arthritis, fatigue, hepatosplenomegaly, etc.[1] It was first identified by Japanese pathologists Kikuchi and Fujimoto independently in the year 1972. Since then, it has been diagnosed in all ages and ethnicities around the world. However, it has been found to affect young people of Asian origin more with slight female predominance according to most research.[2,3]

The aetiology of Kikuchi disease is still unknown but two broad theories: infectious and autoimmune have been formulated. As per infectious both viral and bacterial agents have been considered as possible triggers

for the disease. Meanwhile, in the autoimmune hypothesis, human leukocyte antigens (HLAs) class II alleles have been identified in populations more likely to get this disease[4]. Similarly, it has also been found in patients with autoimmune diseases like systemic lupus erythematosus (SLE), Wegener's granulomatosis, Sjogren's syndrome, Still's disease, rheumatoid arthritis, etc.[5] Thus, autoimmune disease may precede, coincide with, or follow a diagnosis of Kikuchi disease[4].

Kikuchi disease is often misdiagnosed due to its rare nature and similarity to other etiologies leading to incorrect management. Here, we present a case of a 20-year female who had presented with cervical lymphadenopathy. She was managed conservatively and on further workup, a diagnosis of Kikuchi disease was made.

## 2 CASE PRESENTATION

20-year female, non-smoker, non-alcoholic with no known co-morbidities presented to the OPD with a complaint of small swelling at the neck behind and beneath the right ear for one and half months. The painless swelling developed insidiously, which was gradually progressive currently to the size as shown in Figure 1. She also had a fever for one month which was intermittent in nature with a maximum recorded temperature of 100 F, not associated with chills and rigors and was partially controlled with antipyretics. There are no alarming signs like loss of appetite, weight loss, cough or contact with a tubercular patient in the past.

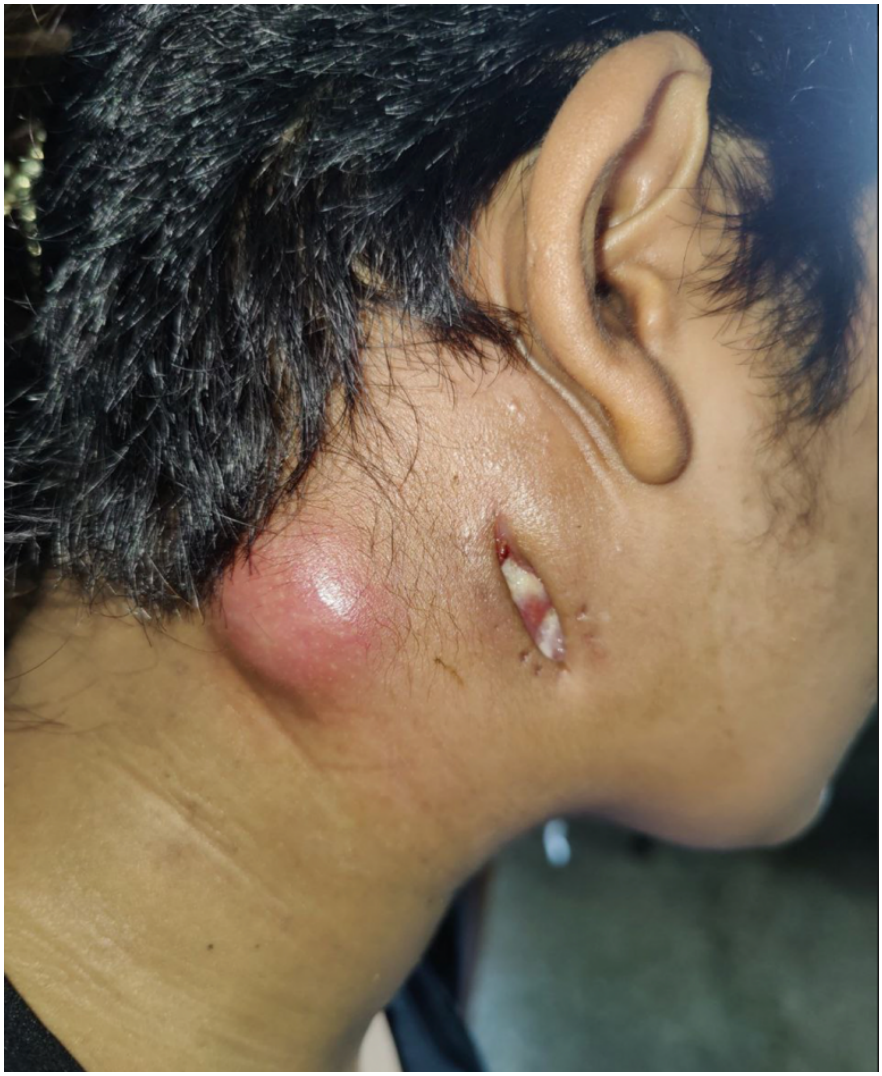

FIGURE 1- 20-year female with swelling behind the angle of mandible

On examination, she was cooperative, alert and well oriented to time, place, and person. She had a BMI of 20.5 kg/$M^2$. Her vital signs were stable. There was no significant finding on her general examination besides cervical lymphadenopathy. Her cervical groups of lymph nodes were enlarged bilaterally with a maximum being 2x2cm. On further examination, the lymph nodes were discrete and non-tender with intact overlying skin and without signs of inflammation, ulceration, and discharge. Oral, otolaryngologic, and systemic examinations were unremarkable.

Her baseline investigations were sent. Her haematological parameters were within normal limits. The thyroid function test was normal. Chest X-ray findings were normal. Serological tests were non-reactive. She was then advised for USG of Neck.

The USG of the neck showed right parotid gland enlargement with multiple round-shaped hypoechoic lesions involving the right parotid and submandibular regions, likely necrotic lymph nodes, one measuring 13*11 mm. For further evaluation, she was advised for FNAC (Fine Needle Aspiration Cytology) along with a biopsy of the affected lymph node. Smear was prepared which showed moderate cellular specimen containing few granulomas composed of epithelioid cells in the background of polymorphous lymphoid cells. Ziehl-Neelsen

stain done for acid-fast bacilli was negative. FNAC showed features suggestive of chronic granulomatous lymphadenitis while a biopsy of the right cervical lymph node carried out revealed widespread necrosis along with disruption of the normal lymph node architecture. The normal follicular arrangement of lymphocytes was lost and focal areas of haemorrhage were also noted. Granulomas or giant cells were absent.

FNAC along with biopsy reports were suggestive of necrotizing lymphadenitis. Based on the histopathological findings diagnosis of Kikuchi Fujimoto disease was made.

The patient was admitted after initial evaluation in OPD. She was then managed conservatively with analgesics, antipyretics, and antibiotics for three days. On the $4^{th}$ day of admission, she was sent home. She along with her family members were reassured that KFD is a self-limiting condition and advised to come for a follow-up in 2 months.

On follow-up 2 months later, her neck swelling and fever were found to have subsided. The patient and patient parties were satisfied with the treatment provided.

## 3 DISCUSSION

Kikuchi disease, since its first description in1972, has puzzled many clinicians because of its rarity and vague clinical presentation and is therefore frequently underdiagnosed. This disease is known to have a higher prevalence in young females of Asian and East European origin, the usual age of presentation being the third to fourth decades of life.[2] The female: male ratio was classically overemphasized to be 4:1 but after the discovery of more and more cases worldwide, the actual ratio is believed to be closer to 1:1. 7 The precise incidence and prevalence of this condition in the general population have not been estimated yet. [8]

Its etiopathogenesis is not entirely understood and remains controversial. A long list of viral triggers, more commonly associated with Epstein Barr Virus but without a direct causal relation, points toward the infectious cause and its association with other autoimmune diseases gives an insight into its possible autoimmune nature. With this limited knowledge, the cause can be explained as an unknown (viral or infectious) trigger leading to an inflammatory process in a susceptible population. [9]

A comprehensive review by Bosch et al. shows unilateral cervical lymphadenopathy involving lymph nodes of the posterior triangle to be the most common clinical manifestation, which was present in 56% to 98% of the patients. Lymphadenomegaly would range from 0.5 to 4 cm, rarely greater than 6 cm, and 30% to 50% of patients had a low-grade fever with upper respiratory symptoms. [2] Our patient also had a low-grade fever with right cervical lymphadenomegaly of 1.3 cm, which is consistent with the review article. The skin might be involved in 40% of the cases, with the presentation varying from nonspecific skin rash to lupus-like findings. Other less frequent manifestations can be nausea, vomiting, weight loss, night sweats, fatigue and arthralgia, which would otherwise mimic B-like presentation.[4]

Diagnostic challenge poses a major problem when approaching patients with this condition, which subjects patients to get inappropriate treatment for alternative etiologies. This happens because of the lack of its pathognomonic signs and symptoms and other common conditions that fit better in the clinical picture like tuberculosis, viral infections, systemic lupus erythematosus and metastatic disease.[10]Excisional lymph node biopsy is required for its definitive diagnosis, which can be supplemented with necessary laboratory and radiological investigations. [2] The histopathological examination reveals well-circumscribed areas of coagulative necrosis with karyorrhectic nuclear debris, a large accumulation of histiocytes in the periphery and relative paucity of neutrophils and eosinophils. [11] The peculiar feature of these histiocytes is the crescent-shaped nuclei and phagocytosed debris, which differentiates this condition from tubercular lymphadenitis and lymphoma. [12]Moreover, Immunohistochemistry (IHC) shows histiocytes positive for myeloperoxidase and CD68, T cells positive for CD8 and infrequent B cells, which rules out lymphoma from the list. Biopsy in KFD fails to show hematoxylin bodies along with areas of vasculitis around areas of necrosis, which are specific to SLE. [11] Fine Needle Aspiration Cytology (FNAC) can also be used for its diagnosis but its use is limited by the fact that specimen collected from this procedure is operator sensitive and it requires careful precautions to preserve the architecture of the lymph nodes. [13] Ultrasound can be used as an imaging

modality in limited-resource settings to support the diagnosis, which can show hypervascularity and lymph node enlargement. [14] In our case, USG findings initially pointed towards tubercular lymphadenitis which was later corrected by FNAC and excisional biopsy.

Through this case, we highlight that Kikuchi disease due to a wide range of clinical presentations can be challenging to diagnose. Dorfman and Berry as part of the Lymphoma Task Force have reported a misdiagnosis rate of 40% for Kikuchi Disease. [15] Most of them are misdiagnosed as malignancy while in our part of the world tuberculous lesion is another frequent misdiagnosis. This misdiagnosis leads to the concern of expensive management procedures for a mild self-limiting illness. This adds a burden on limited health care resources as well as the patients physically, mentally and financially. We want to increase awareness about KDF among clinicians and encourage them to consider it as an important alternate diagnosis in patients with lymphadenopathy of unclear aetiology.

Since the condition has a benign and self-limiting course, observation alone is the commonly used approach in its management. The lack of specific treatment for this condition reiterates the poor understanding of its etiopathogenesis. Antipyretics and analgesics can be used for supportive management and in case of severe disease, prolonged corticosteroid use may be necessary, once the infectious cause has been ruled out. [4] The patient in our case had mild symptoms, which subsided with the symptomatic treatment for pain and fever.

## 4 CONCLUSION

Kikuchi disease is a benign disease and needs to be kept in mind to avoid overtreatment in the line of malignancy or tubercular lymphadenitis whenever a person comes with cervical lymphadenopathy.

## AUTHORS CONTRIBUTIONS

Shekhar Gurung (SG)= Led data collection, literature review, contributed to writing the case information. Ribek Sunam Pariyar (RSP)= Literature review, led data collection and concept of study. Saurab Karki (SK)= Literature review, revising, and editing the rough draft into the final manuscript. Anu Gautam (AG)= Literature review, writing an introduction, case report, discussion and editing manuscript. Hari Sapkota (HS)= Literature review, led data collection. All authors were involved in manuscript drafting and revising, and approved the final version.

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**CONFLICTS OF INTEREST:** None.

**ETHICAL APPROVAL:** N/A.

## CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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