Asian Journal of Medicine and Health

Volume 21, Issue 11, Page 97-101, 2023; Article no.AJMAH.106687
ISSN: 2456-8414

A Rare Case of Kikuchi-Fujimoto Disease in an Indian Female

Shubham Darda [a++], Aarjuv Majmundar [a++*], Puneet Upadhyay [a++], Harshita H. Nouni [a#], Sucheta Lakhani [b#] and Jitendra D. Lakhani [a]

[a] Department of Medicine, SBKS MI&RC, Vadodara, India.
[b] Department of Microbiology, SBKS MI&RC, Vadodara, India.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/AJMAH/2023/v21i11923

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/106687

Case Report

Received: 21/07/2023
Accepted: 25/09/2023
Published: 29/09/2023

ABSTRACT

Kikuchi-Fujimoto disease is a self-limiting illness characterised by non-specific symptoms, most commonly reported symptoms being fever and lymphadenopathy. Unknown in its cause, Kikuchi-Fujimoto disease (KFD) is a benign, uncommon disorder usually presenting under the age of 30 years in women. This disease is mainly seen in Asian population mainly in Japan but it is rare in Indian population. The etiological cause of this disease remains unknown. This condition needs to differentiated from other closely resembling conditions such as tuberculosis, lymphoma, Systemic Lupus Erythematosus (SLE) and adenocarcinoma. The physician needs to be aware about this condition to avoid misdiagnosis and improper treatment. This case report is about a female of age 26 years who was diagnosed as having Kikuchi Fujimoto Disease on the basis of clinical and biopsy findings.

[++]Resident;
[#]Professor;
**Corresponding author: E-mail: aarjuv1996@gmail.com;*

Keywords: Kikuchi-Fujimoto syndrome; benign; lymphoma; tuberculosis; lymphadenopathy.

1. INTRODUCTION

Kikuchi-Fujimoto disease also known as histiocytic necrotizing lymphadenitis, Kikuchi necrotizing lymphadenitis, phagocytic necrotizing lymphadenitis, subacute necrotizing lymphadenitis, and necrotizing lymphadenitis, was first described in 1972 in Japan by Dr Masahiro Kikuchi and by Y. Fujimoto independently [1-5]. Kikuchi-Fujimoto disease (KFD) is a rare, self-limiting disorder that typically affects the cervical lymph nodes [1]. The presentation of Kikuchi-Fujimoto disease is variable, and there is no specific set of symptoms or laboratory features that reliably establishes the diagnosis [6]. Although autoimmune reactions and unknown pathogenic organisms have been recognized as the main culprits, the aetiology of KFD is still unknown [7]. Kikuchi disease occurs sporadically in people with no family history of the condition [5]. Very few cases of Kikuchi-Fujimoto disease have been described in Indian patients. It is especially important to differentiate this condition from tuberculosis because of the high prevalence of tuberculosis in Indian population, close resemblance and different treatments. Other close differential diagnosis includes lymphoma and adenocarcinoma [1]. Despite over 50 years having passed, it is still difficult to identify KFD from other lymph node illnesses, which poses a serious danger of misdiagnosis and inappropriate therapy [8]. A physician needs to be aware of this condition to prevent misdiagnosis and inappropriate treatment. We hereby describe a case of Kikuchi-Fujimoto disease in a young adult female of Indian descent.

2. CASE PRESENTATION

A 26-year-old Indian female who had no major prior medical history arrived at our outpatient department with complaints of fever and neck swelling that developed over a course of 30 days. She was treated at a local hospital with empirical antibiotics with no improvement of symptoms. No drug or alcohol use was reported. Family history was unremarkable. She was a home-maker from Madhya Pradesh and was married for 4 years. Initially, she was given treatment for seven days at a local hospital with no improvement in symptoms. She was referred to our hospital for additional treatment due to her ongoing fever and bilateral cervical region swelling. Indoor admission consent was obtained from the patient and attendants. On admission to the female medical ward, the patient was noted to be febrile. She also had bilateral cervical lymphadenopathy which ranged from 0.8-1.2 cm in size with no complains of cough or breathlessness. On admission, her heart rate was 128/min, her respiratory rate was 20/min, her blood pressure was 120/70 mmHg, and her temperature was 40.2°C. Investigations performed on admission are mentioned in the following table.

Table 1. Investigations on admission

Laboratory Data	Results	Reference value
Haemoglobin	11.7 gm%	13-17 gm%
WBC	11,600 cell/cumm	4000-11000 cell/cumm
Platelet	2.4 lacs/cumm	1.5-4.5 lacs/cumm
ESR	20 mm	<18 mm
Urea	32 mg%	8-52 mg%
Serum Creatinine	0.8 mg%	0.6-1.3 mg%
RBS	74 mg/dL	70-126 mg/dL
HIV/HCV/HBSAG (Rapid tests)	Negative	
Sodium/Potassium	134/4.4 mmol/L	135-145/3.5-5.5 mmol/L
T. Bilirubin	0.6 mg%	0.2-1.0 mg%
CRP	28.4 mg/L	0-6.0 mg/L
SGOT	24 mmol/L	Up to 40 mmol/L
SGPT	24 mmol/L	Up to 40 mmol/L
Albumin	2.6 gm%	3.4-5.4 gm%
EBV mono spot test	Negative	
HIV (ELISA)	Negative	
P.S. For Immature or Atypical cells	Negative	
ENT examination	Normal	
H1N1 RTPCR	Negative	

Laboratory Data	Results	Reference value
COVID-19 RTPCR	Negative	
ANA Profile	Negative	
RA factor	Negative	
Anti-CCP Antibodies	Negative	
Urine Routine	Micro Albuminuria absent, Sugar absent, no cast	
Lymph Node Biopsy	Focal Areas of Non-suppurative Necrosis with Histiocytic and Plasmacytoid cell infiltrates	

A pulmonary and ENT (ear, nose and throat) examination was normal. Lab work revealed mild leucocytosis, elevated acute phase reactants and hypoalbuminemia with normal aspartate aminotransferase, alanine aminotransferase and platelet counts. Peripheral smear came out negative for atypical or immature cells. The serology for HIV, HCV and HbSAg was negative. The EBV mono-spot test and indirect fluorescent antibody test both were negative. Both the COVID-19 and H_1N_1 RTPCR results were negative. The samples of blood and sputum were sent for sensitivity and culture testing. The patient was given standard symptomatic treatment including antibiotics. Investigational blood was sent, and blood for blood cultures was obtained. A lymph node biopsy was planned on the 4th day of admission and after meeting all pre-procedural requirement a lymph node biopsy was performed on the 5th day of admission.

The biopsy showed focal areas of non-suppurative necrosis with histiocytic and plasmacytoid cell infiltrates, The lymphnode biopsy was negative for AFB (Acid fast bacilli) stain, atypical cells, immature cells and Gram's stain. Based on biopsy results, negative EBV serology and peripheral smear results a diagnosis of Kikuchi-Fujimoto syndrome was suspected.

She was managed symptomatically with anti-pyretics and NSAIDs for the pain. Fever resolved on the 7th day of admission and she was discharged on similar oral treatment. On follow up after 15 days, her fever had resolved with reduction in the size of lymphnodes. On further follow up after 2 months the lymphadenopathy had completely resolved with no other complains.

3. DISCUSSION AND CONCLUSION

Kikuchi-Fujimoto disease (KFD), was first observed in Japan in 1972. Since then, it has been reported in a number of nations. It is widely acknowledged that KFD is a form of lymphoid reactive hyperplasia that causes non-neoplastic lymph node hypertrophy [7]. KFD was formerly believed to be more common among women, however current research suggest the condition may affect both sexes [9].

Fever and cervical lymphadenopathy have a wide range of potential differential diagnoses, which frequently results in a thorough workup. Our patient was tested for lymphoma, tuberculosis and adenocarcinoma. The diagnosis was aided by the results of the lymph node biopsy. Necrosis and a histiocytic cellular infiltration can be found in one or more locations within the lymph node, which is the hallmark histology of KFD. There is a chance that the node's capsule will be penetrated, and perinodal inflammation is usual [10].

Even though autoimmune and infectious aetiologies have been put out as potential causes, KFD illness still has no established aetiology. There have been some suggested causal organisms. These include Brucella, Yersinia enterocolitica, Epstein-Barr virus, human T-cell leukaemia virus type 1, human herpesvirus type 6, B19 parvovirus, cytomegalovirus, and parainfluenza virus [11]. Case reports include KFD developing concurrently with or after circumstances that may have acted as triggers, such as vaccination and COVID-19 infection [12-17].

Unilateral cervical lymphadenopathy, with or without systemic signs and symptoms like fever and skin rash, is the most typical clinical symptom of Kikuchi illness [4,18-20]. In this case, our patient had bilateral cervical lymphadenopathy and fever.

With a few exceptions, the patterns of lymphadenopathy in TB, KFD, and lymphoma are comparable. In KFD, the lymph nodes are typically unilateral cervical or jugular with a perinodal infiltration and karyorrhetic debris, and their size ranges from 0.5 to 2.5 cm. These conclusions were made in this instance, favouring KFD more. Both patients may have nodal necrosis, although TB is more likely to have it [21,22].

Almost typically, Kikuchi disease has a benign course and clears up in a few weeks to months. For KFD, there is no proven effective treatment. It's a benign, self-limiting condition that goes away in one to four months. However, patients should be followed up later on because they might develop Systemic Lupus Erythematosus (SLE) or in rare cases, relapse with KFD. The latter does not frequently occur again [23-25].

KFD is often treated with supportive care. Symptomatic relief is the main goal of treatment, which includes using analgesics and antipyretics to reduce fever and sensitive lymph nodes. For severe cases or recurring illnesses, corticosteroids are only used. It is advised that patients have a comprehensive survey and follow-up to assess any subsequent development of SLE because of the relationship with the disease.

For steroid-resistant or recurring cases – hydroxychloroquine is indictated.

Intravenous immunoglobulin is indicated for situations that are resistant to steroids.

Our patient was managed symptomatically with anti-pyretics and NSAIDs for the pain. Fever resolved on the 7th day of admission and she was discharged on similar oral treatment. On follow up after 15 days, her fever had resolved with reduction in the size of lymphnodes. On further follow up after 2 months the lymphadenopathy had completely resolved with no other complains.

SIGNIFICANT OF THE STUDY

It is particularly important to distinguish this disorder from tuberculosis due to the high prevalence of tuberculosis in the Indian population, similarity, and distinct therapies. Other near differential diagnosis include lymphoma and adenocarcinoma [1]. To avoid an incorrect diagnosis and the wrong kind of treatment, a doctor needs to be aware of this illness. In our case we present a young adult female of Indian heritage who had Kikuchi-Fujimoto Disease.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Rammohan A, Cherukuri SD, Manimaran AB, Manohar RR, Naidu RM. Kikuchi-Fujimoto Disease: A Sheep in Wolf's Clothing. J Otolaryngol Head Neck Surg. 2012;41(3):222–26. PMID 22762705.
2. Kaushik V, Malik TH, Bishop PW, Jones PH. Histiocytic necrotising lymphadenitis (Kikuchi's disease): A rare cause of cervical lymphadenopathy. Surgeon. 2004; 2(3):179–82. DOI:10.1016/s1479-666x(04)80084-2. PMID 15570824.
3. Bosch X, Guilabert A. Kikuchi-fujimoto disease. Orphanet J Rare Dis. 2006;1:18. DOI:10.1186/1750-1172-1-18. PMC 1481509. PMID 16722618.
4. Bosch X, Guilabert A, Miquel R, Campo E. Enigmatic Kikuchi-Fujimoto disease: A comprehensive review. Am. J. Clin. Pathol. 2004;122(1):141–52. DOI:10.1309/YF08-1L4T-KYWV-YVPQ. PMID 15272543.
5. Kikuchi M. Lymphadenitis showing focal reticulum cell hyperplasia with nuclear debris and phagocytes. Acta Hematol Jpn. 1972;35:379–80.
6. Abdu Ahmed BA, MedSci*; Pajkrt Dasja MD, PhD, MBA†; Bijker, Else M. MD, PhD‡. Kikuchi Disease in children: Case report and review of the literature. The Pediatric Infectious Disease Journal. 2022; 41(3):199-204.
7. Perry AM, Choi SM. Kikuchi-fujimoto disease: A review. Arch Pathol Lab Med. 2018;142:1341–6.
8. Lahma J, Arkoubi Z, Hejjouji R, Nitassi S, El Ayoubi A, Bencheikh R, et al. About a rare disease misdiagnosed as malignant lymphoma or tuberculosis: Kikuchi-Fujimoto's disease. Pan Afr Med J. 2018; 31:1–5.
9. Lin HC, Su CY, Huang CC, Hwang CF, Chien CY. Kikuchi's disease: A review and

analysis of 61 cases. Otolaryngol Head Neck Surg. 2003;128:650–3.
10. Loachim HL, Ratech H. Loachim's lymph node pathology. 3rd ed. New York: Lippincott Williams & Wilkins; 2002.
11. Menasce LP, Banerjee SS, Edmondson D, Harris M. Histiocytic necrotizing lymphadenitis (Kikuchi-Fujimoto disease): Continuing diagnostic difficulties. Histopathology. 1998;33(3):248–254.
12. Jaseb K, Nameh Goshay Fard N, Rezaei N, Sadeghian S, Sadeghian S. COVID-19 in a case with Kikuchi-Fujimoto disease. Clin Case Rep. 2021;(3):1279-1282.
13. Al Ghadeer HA, AlKadhem SM, AlMajed MS, AlAmer HM, AlHabeeb JA, Alomran SH, et al. Kikuchi-Fujimoto Disease Following COVID-19. Cureus. 2022; (1):e21049.
14. Tan HM, Hue SS, Wee A, See KC. Kikuchi-fujimoto disease post COVID-19 vaccination: Case report and review of literature. Vaccines (Basel). 2021;9(11).
15. Racette SD, Alexiev BA, Angarone MP, Bhasin A, Lima K, Jennings LJ, et al. Kikuchi-Fujimoto disease presenting in a patient with SARS-CoV-2: a case report. BMC Infect Dis. 2021;21(1):740.
16. Soub HA, Ibrahim W, Maslamani MA, A Ali G, Ummer W, Abu-Dayeh A. Kikuchi-fujimoto disease following SARS CoV2 vaccination: Case report. IDCases. 2021; 25:e01253.
17. Guan Y, Xia X, Lu H. Kikuchi-fujimoto disease following vaccination against COVID-19. J Hematop. 2022;(1):21-23.
18. Alam H, Saeed MO, Saeed AB, Zaidi A. Kikuchi disease. J Pak Med Assoc. 2015;65(12):1349-50.
19. Lamzaf L, Harmouche H, Maamar M, Adnaoui M, Aouni M, Tazi Mezalek Z. Kikuchi-fujimoto disease: Report of 4 cases and review of the literature. Eur Ann Otorhinolaryngol Head Neck Dis. 2014; 131(6):329-32.
20. Kuo TT. Kikuchi's disease (histiocytic necrotizing lymphadenitis). A clinicopathologic study of 79 cases with an analysis of histologic subtypes, immunohistology, and DNA ploidy. Am J Surg Pathol. 1995;19(7):798-809.
21. Iguchi H, Sunami K, Yamane H, et al. Apoptotic cell death in Kikuchi's disease: A TEM study. Acta Otol Laryngol. 1998;118:250–3. [PubMed] [Google Scholar]
22. Kucukardali Y, Solmazgul E, Kunter E, et al. Kikuchi-Fujimoto disease: Analysis of 244 cases. Clin Rheumatol. 2007;26:50–54
23. Litwin MD, Kirkham B, Henderson DR, Milazzo SC. Histiocytic necrotising lymphadenitis in systemic lupus erythematosus. Ann Rheum Dis. 1992; 51(6):805–807.
24. Hsiang-Cheng C, Jenn-Huang L, Guo-Shu H, et al. Systemic lupus erythematosus with simultaneous onset of Kikuchi-Fujimoto's disease complicated with antiphospholipid syndrome: A case report and review of the literature. Rheumatol Int. 2005;254:303–306.
25. Leyral C, Camou F, Perlemoine C, Caubet O, Pellegrin JL, Viallard JF. Pathogenic links between Kikuchi's disease and lupus: A report of three new cases [in French].Rev Med Interne 2005;268: 651–655.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/106687