



ORIGINAL RESEARCH PAPER

Arts

KIKUCHI - FUJIMOTO DISEASE : A RARE CASE OF NECROTIZING LYMPHADENITIS.

KEY WORDS:

Ramakrishnan. C Post Graduate Student Department of General Surgery Mahatma Gandhi Medical College and Research Institute Sri Balaji Vidyapeeth – Deemed to be University .

Siddhartha Gowthaman* Assistant Professor Department of General Surgery Mahatma Gandhi Medical College and Research Institute Sri Balaji Vidyapeeth – Deemed to be University . *Corresponding Author

Ramanathan.M Professor Department of General Surgery Mahatma Gandhi Medical College and Research Institute Sri Balaji Vidyapeeth – Deemed to be University .

ABSTRACT

Kikuchi – Fujimoto disease (KFD) or necrotizing histiocytic lymphadenitis is characterized by fever, lymphadenopathy (usually cervical) and malaise. We are presenting a 26 year old women from Puducherry, with one month history of fever and cervical lymphadenopathy. On examination, she had level 2 and level 3 enlarged lymphnodes in the right side of her neck and her systemic examination was normal. Her ESR was mildly elevated and her total leucocyte counts were normal. Ultrasound guided FNAC revealed only a hemorrhagic aspirate and was inconclusive. So lymphnode biopsy was done which showed abundant necrosis, nuclear karyorrhexis. Usually the disease is self limiting and needs only symptomatic treatment as opposed to other causes of lymphadenitis like tuberculosis which is more common in our country.

INTRODUCTION:

Kikuchi – Fujimoto disease (KFD) is histologically characterized by histiocytic necrotizing lymphadenitis. The exact etiology of the disease is still not known. It was first described in Japan by Kikuchi and Fujimoto et al in 1972 [1] [2]. The disease is more common in Asians. The histopathological and blood picture in KFD suggests a possible viral etiology [3] [4] [5]. KFD is associated with Human Leucocyte Antigen (HLA) class II alleles mainly HLA DPA1 and HLA DPB1 which are more prevalent in Asians which makes the disease more common in the Asian ethnicity. KFD is also seen in association with other autoimmune conditions like Systemic Lupus Erythematosus (SLE) [7]. There are no definitive diagnostic test available for KFD but a histopathological picture of lymphnode with abundant necrosis, nuclear karyorrhexis and occasional presence of histiocytes at the edge of the necrosis. Treatment is mainly conservative as the disease is self limiting. Early identification of this condition is essential since the disease is self limiting and thus prevents the patient to undergo a radical and potentially toxic treatment for the other conditions with lymphadenitis.

CASE REPORT:

A 23 year old old woman from Puducherry presented to our hospital with one month history of swelling in the right side of her neck for one month duration. The swelling was associated with low grade fever which was intermittent and generalized myalgia.

She had significant loss of weight and appetite. There were no known co morbidities in the patient. On examination she had two lymphnodes on the right side level 2 and level 3 measuring 2 X 2 cm, firm, non tender and mobile. ENT examination, Oral cavity and Systemic examination revealed no other abnormality.

Her blood investigations showed normal leucocyte count, mild anemia and elevated ESR. X-Ray chest was normal. Ultrasound of Neck showed two enlarged right cervical lymphnodes with features of necrosis. Ultrasound guided FNAC was done which was inconclusive due to hemorrhagic aspirate. So patient was posted for excision biopsy of the lymph node under general anaesthesia. Biopsy revealed features of necrotizing lymphadenitis ie Kikuchi Fujimoto

disease. Patient was managed conservatively with systemic steroids. Patient improved symptomatically and was discharged.

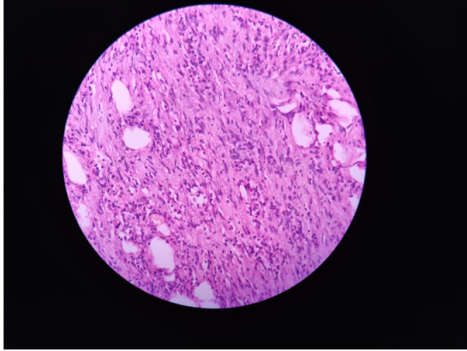
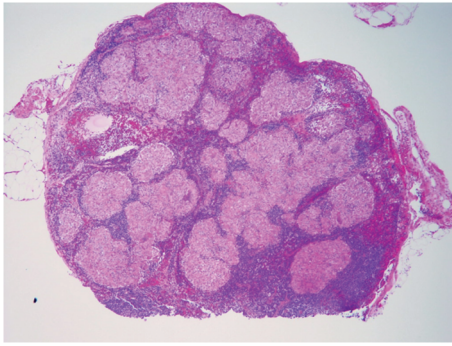
DISCUSSION:

Kikuchi Fujimoto Disease is a benign condition characterized by necrotizing lymphadenitis which is often self limiting or can be treated with steroids. Mostly it occurs in young women of Asian ethnicity [8,9]. Kikuchi disease has an onset which is mostly insidious and can be symptomatic for a period of 2 – 3 weeks. Fever is the primary symptom and usually associated with unilateral lymphadenopathy [10]. There are no specific laboratory investigations to diagnose the disease. The disease is usually non fatal but very rarely fatality has been reported [10]. The best approach to treat these patients is by NSAIDs and steroids are usually recommended only in refractory cases. There are also reports which have successfully treated the disease with Hydroxychloroquin, cyclosporine, azathioprine and intravenous immunoglobulins [11].

CONCLUSION:

Diagnosing kikuchi disease is of importance because it rules out other causes for lymphadenitis like tuberculosis, lymphoma and so on. This could prevent the patient from undergoing potentially toxic therapies.





REFERENCES:

1. Kikuchi M. Lymphadenitis showing focal reticulum cell hyperplasia with nuclear debris and phagocytosis. *Nippon Ketsueki Gakkai Zasshi.* 1972;35:378–380.
2. Fujimoto Y, Kozima Y, Hamaguchi K. Cervical necrotizing lymphadenitis: a new clinicopathological agent. *Naika.* 1972;20:920–927.
3. Bosch X, Guilabert A, Miquel R, Campo E. Enigmatic Kikuchi-Fujimoto disease: a comprehensive review. *Am J Clin Pathol.* 2004;122(1):141–152.
4. Hutchinson CB, Wang E. Kikuchi-Fujimoto disease. *Arch Pathol Lab Med.* 2010;134(2):289–293.
5. Pepe F, Disma S, Teodoro C, Pepe P, Magro G. Kikuchi-Fujimoto disease: a clinicopathologic update. *Pathologica.* 2016;108(3):120–129.
6. Tanaka T, Ohmori M, Yasunaga S, et al. DNA typing of HLA class II genes (HLA-DR, -DQ and -DP) in Japanese patients with histiocytic necrotizing lymphadenitis (Kikuchi's disease). *Tissue Antigens.* 1999;54(3):246–253.
7. Imamura M, Ueno H, Matsuura A, et al. An ultrastructural study of subacute necrotizing lymphadenitis. *Am J Pathol.* 1982;107(3):292–299.
8. Kido H, Kano O, Hamai A, Masuda H, Fuchinoue Y, Nemoto M, et al. Kikuchi-Fujimoto disease (histiocytic necrotizing lymphadenitis) with atypical encephalitis and painful testitis: a case report. *BMC Neurology.* 2017;17:22.
9. Mrowka-Kata K, Kata D, Kyrzcz-Krzemien S, Sowa P. Kikuchi-Fujimoto disease as a rare cause of lymphadenopathy—two cases report and review of current literature. *Otolaryngologia polska=The Polish otolaryngology.* 2013;67(1):1.
10. Dalugama C, Gawarammana IB. Fever with lymphadenopathy—Kikuchi Fujimoto disease, a great masquerader: a case report. *Journal of medical case reports.* 2017;11.
11. Srikantharajah M, Mahendra P, Vydianath B, Lowe GC. Case Report: Kikuchi-Fujimoto disease: a rare but important differential diagnosis for lymphadenopathy. *BMJ case reports.* 2014;2014.