Kikuchi necrotizing lymphadenopathy: a tip of the iceberg disease?

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ABSTRACT

Kikuchi-Fujimoto disease (KFD) is a rare benign condition also called histiocytic necrotizing lymphadenitis, which typically presented as fever with cervical lymphadenopathy in previously healthy individual. We presented a case of 11 year old boy with fever and cervical lymphadenopathy since 2 months. Lymph node biopsy was performed which suggested of KFD and was treated symptomatically. KFD incidence is rare but clinicians should be alert if young patient comes with fever and cervical lymphadenopathy to lower the chance of unwanted laboratory test and harmful treatment.

Keywords: Fever with lymphadenopathy, Kikuchi-Fujimoto disease, Histiocytic necrotizing lymphadenitis

INTRODUCTION

KFD or histiocytic necrotizing lymphadenitis is rare, benign self-limiting systemic lymphadenitis of unknown etiology. This disease was first reported in 1972 in Japan by Kikuchi and Fujimoto.¹,² Misdiagnosed rate is up to 40% because of shared clinical features.³ Females are more commonly affected than males, less than 30 years age group is more affected than older age groups.⁴ With unknown etiology of KFD, its associated with viral or post-viral etiology has been proposed.⁵

CASE REPORT

A 11 year old boy presented in OPD with history of fever for 7 days. Fever was continuous, high grade, accompanied by chills. His parents had noticed swellings on the left side of his neck, which were painful and increasing in size over the last 2 months. There was no history of vomiting, rash, arthralgia, breathlessness, mucosal bleed or contact with tuberculosis. On examination he was conscious, febrile, non-toxic, had 5 to 7 tender, enlarged matted left cervical lymph nodes of 3xl cm size. There were no rashes or generalized lymphadenopathy. The examination of other systems was normal, treated symptomatically. Fever persisted with increasing in size of cervical lymph node hence relevant investigations sent which revealed hemoglobin 12.1 g/dl, TLC 5,300 cells/mm³, neutrophils 46.7%, lymphocyte 42.7%, monocyte 9.7%, eosinophils 0.7%, basophils 0.2%, platelet count 2.9 lacs/mm³ and ESR 40 mm/hr, CRP 4. Peripheral smear was normal, no atypical cells seen. Widal, dengue IgG, NS1 antigen, MP by card were negative. Mantoux, chest X-ray was normal. IgM for EBV (Epstein Barr virus) was negative. Leptospirosis IgM, HbsAg and HIV by ELISA were negative. Blood and urine cultures were sterile. Ultrasound neck showed multiple enlarged matted heterogeneous lymph nodes and few sub centimeter sized non-specific lymph nodes were seen in right cervical region. FNAC showed reactive specific lymph nodes hence put on macrolide (azithromycin) and NSAIDS. On follow up fever was subsided but left cervical lymph node remains enlarged and tender hence excision biopsy of the enlarged left cervical node was done. Histopathology revealed partially effaced architecture, paracortical collection of histiocytes, focal...
necrosis, karyohexis, fibrin deposit and many foamy macrophages were also seen findings suggestive of necrotising histiocytic lymphadenitis or KFD.

Following this child was treated with NSAIDs and responded well to treatment and clinically improved.

**DISCUSSION**

KFD or histiocytic necrotising lymphadenitis was described in 1972 as a rare benign disease more prevalent in Asia especially among Japanese that was characterized by lymphadenopathy, lymph node tenderness, fever. Less common symptoms included malaise, night sweats, chills, cough, arthralgia, nausea, vomiting and weight loss also presented with hepato-splenomegaly.5 KFD is a rare but self-limiting process which typically resolve over few weeks to months in majority of cases.5 Posterior cervical lymph nodes was most commonly involved with size of 0.5 cm to 4 cm.7 Our case presented with fever, lymphadenopathy with negative TB screening. With unknown aetiology of KFD, its associated with viral or post-viral aetiology had been proposed which included EBV, HIV, dengue virus, parovirus B19, HSV.5 Other possible associations with autoimmune had been proposed as electron microscopic studies showed tubular reticular structures in cytoplasm of stimulated lymphocytes and histiocytes in KFD with SLE patients.8 Various systemic complications had been seen in KFD such as cardiac tamponade, acute renal failure, mesenteric lymphadenopathy, hepatitis, hemophagocytic syndrome, interstitial lung disease and pleural effusion, isolated mediastinal lymphadenopathy, panuveitis reported beside this uncommon central nervous system complications such as aseptic encephalitis and meningitis and peripheral neuropathy were also reported.9 Differential of KFD were tubercular adenitis, lymphoma, SLE, Kawasaki disease. With no specific diagnostic laboratory tests lymph node biopsy and histopathological examination confirmed the diagnosis of KFD showing focal or complete loss of follicular architecture associated with necrosed cortical and paracortical areas consisted of immunoblast cells, small lymphocytes, macrophages and so-called plasmacytoid T-cells.10

**CONCLUSION**

Physicians should keep differential of KFD if young patient present with fever and cervical lymphadenopathy because KFD resembles tuberculosis adenitis, lymphoma, SLE, Kawasaki disease. Because very few case have been reported in medical literature. It appears to be tip of iceberg disease in India, probably TB being more common and expertise to pick up KFD in biopsy appears to be limited too. This will save the child and family the trauma, stigma and unnecessary cost of treating other similar diseases. Patience for workup, expertise in biopsy and continuous communication with family appears to be key.

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**REFERENCES**

1. Kikuchi M. Lymphadenitis showing focal reticulum cell hyperplasia. Nippon Ketsueki Gakkai Zasshi. 1972;35:378-80.
2. Fujimoto Y, Koizima Y, Yamaguchi K. Cervical subacute necrotizing lymphadenitis. A new clinicopathological agent. Naika. 1972;20:920-7.
3. Ramirez AL, Johnson J, Murr AH. Kikuchi Fujimoto’s disease: an easily misdiagnosed clinical entity. Otolaryngol Head Neck Surg. 2001;125(6):651-3.
4. Kuo T. 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.
5. Sudhiakar MK, Sathyamurthy P, Indhumati E, Rajendran A, Vivek B. Kikuchi’s disease: a case report from South India. Int J Case Rep Image. 2011;2(1):15-8.
6. Bosch X, Guibert A, Miquel R, Campo E. Enigmatic Kikuchi-Fujimoto disease: a comprehensive review. Am J Clin Pathol. 2004;122(1):141-52.
7. Kuo T. 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.
8. Imamura M, Ueno H, Matsuur A, Kamiya H, Suzuki T, K Kikuchi K, et al. An ultrastructural study of subacute necrotizing lymphadenitis. Am J Pathol. 1982;107(3):292-9.

9. Kapoor S. Rare complications of Kikuchi’s disease: beyond pain control. Korean J Pain. 2012;25(4):281-2.

10. Rivano MT, Falini B, Stein H, Canino S, Ciani C, Gerdes J, et al. Histiocytic necrotizing lymphadenitis without granulocytic infiltration (Kikuchi’s lymphadenitis). Morphological and immunohistochemical study of eight cases. Histopathology. 1987;11:1013-27.

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