

Kikuchi lymphadenitis

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Abstract

Introduction: Kikuchi-Fujimoto disease is also known as Necrotising histiocytic lymphadenitis. It is a very rare disorder, benign condition usually self limited to six months period. It mainly affects the young asiatic females and very few cases has been reported from paediatric age group. It simulates like Tuberculous lymphadenitis, Systemic Lupus Erythmatosus, other benign and malignant conditions. The aetiology is unknown, labarotary investigations are usually not remarkable. Diagnosis is done by histopathology of cervical lymphnodes.

Keywords: Histiocytic necrotizing lymphadenitis; Kikuchi disease; Lymphadenopathy.

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INTRODUCTION

Kikuchi's disease or histiocytic necrotising lymphadenitis is an uncommon, benign and self-limited condition of unknown aetiology that was initially described in Japan.¹ Although it has been reported worldwide, it is most commonly seen in Asia specially India and Japan; some of the occasionally-reported cases in Europe and the United States have involved patients of Asian descent.^{2,3} While initially described in young women, Kikuchi's syndrome clearly also occurs in men. The ratio of affected males to females in three series was 1:4, 1:1.6, and 1:1.26, respectively.¹ It mainly affects young adults and is clinically characterised by tender regional lymphadenopathy, fever, and occasional systemic involvement. The lymphadenitis in Kikuchi's disease reveals characteristic histopathologic features of coagulative necrosis and karyorrhectic debris.¹ Differential diagnosis includes lymphoma and lymphadenitis associated with systemic lupus erythematosus (SLE), and certain infectious aetiologies, which share similar clinicomorphologic features.¹

OBJECTIVES

The aims of this study is how kikuchi lymphadenopathy present in paediatric population. To increase the awareness of clinicians to treat such patient presenting fever with cervical lymphadenopathy.

CASE REPORT

A 8 year old female girl presented with history of fever since 2 to 3 weeks, noticed glands in cervical region on right side. Three small cervical lymphnodes were enlarged. There was no rash on trunk and limbs. All other lymphnodes are normal. Systemic examination within normal reports, no hepatosplenomegaly. She had received Amoxycillin/clavulanate in view of bacterial lymphadenitis without reduction of the lymphnode volume. There is no history of exposure to animals, insect bites or contact with infection. Serology titres of Epstein-Barr virus, cytomegalo virus, adenovirus and HIV were negative. Mantoux test was negative after 48 and 72 hours. Haematological investigation were normal except raised erythrocyte sedimentation rate (E.S.R.) of 32mm/hr. Chest x-ray was normal. Her ANA levels are normal. Her cervical lymphnode biopsy was performed, Histopathological examination of cervical lymphnodes revealed necrosis with fibrinoid deposits and apoptotic cells and histiocytes, reactive germinal centers are present. All these changes are suggestive of Histiocytic necrotizing lymphadenitis (Kikuchi Lymphadenopathy). No specific therapy was given and just regular follow-up was done. After 8 weeks she recovered spontaneously and completely. Her fever and cervical lymphnodes were subsided. During follow-up period the general condition of patient was good and no relapse was observed.

DISCUSSION

Though the presentation mentioned above is very common in Indian OPD settings, but the diagnosis is rare²². Most of the times, it was seen that the patient was given antibiotics on the basis of empirical diagnosis and sent to home with reassurance. The patient gets panicked when lymph nodes don't subside in one week. The onset of Kikuchi's disease is usually acute or subacute with fever and regional lymphadenopathy which is mostly cervical; in a previously healthy young adult.⁶ Enlarged lymph nodes range from 0.5 to 4 cm in size and are tender and painful. Other anatomic sites of lymphadenopathy are involved in 2% to 40% of patients. Involvement of mediastinal, peritoneal and retroperitoneal regions is uncommon. Less frequent symptoms such as fatigue, arthralgia, joint pains, nausea, vomiting, anorexia, and sore throat have been reported in 2-7% of patients. Weight loss and night sweats, though rare, have also been observed. Numerous causative agents have been proposed, including Epstein Barr virus (EBV)^{2,3}, human herpesvirus 6, human herpesvirus 8², human immunodeficiency virus (HIV), parvovirus B19², paramyxoviruses, parainfluenza virus, *Yersinia enterocolitica*, and toxoplasma. The diagnosis of Kikuchi's disease is only by histo-pathological studies. It is a form of histiocytic necrotizing lymphadenitis with characteristic histological features (FIG 1). Classic pathological findings in this disease include patchy areas of necrosis in the cortical and paracortical areas of enlarged lymph node, together with nuclear debris or extensive karyorrhexis. Cellular infiltration consists of CD68-positive plasmacytoid histiocytes and transformed lymphocytes (immunoblasts), predominantly of T-cell origin. Neutrophils, eosinophils or plasma cells are sparse or totally absent^{2,3}.

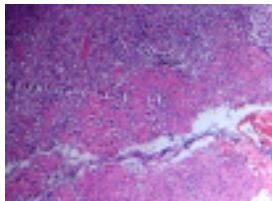


Figure 1: H and E stained section (10 X) of cervical lymph node showing foci of necrosis containing abundant karyorrhectic debris

The most commonly involved site is the cervical lymph nodes region (56–98%). Most often the lymph nodes are tender, involve the posterior cervical triangle (88.5%) and are generally unilateral, like in our first case. Lymph node size ranges from 0.5 to 4 cm, but occasionally lymph nodes may be larger than 8 cm in diameter. In only a few cases lymphadenopathy was generalized or it involved the mediastinal, peritoneal and retroperitoneal region.

Sometimes pain, indurations or nodal adherence to surrounding tissue are referred²

CONCLUSION

Kikuchi's disease is a self limiting disorder of unknown aetiology. It mainly affects the adult population but very few cases have been reported from paediatric age group. First time reported in 1998 by Mathew's. Uptil now only 20 cases have been reported from INDIA.⁶ It is diagnosed histopathologically and can be considered as differential diagnosis of any child with fever of unknown origin and cervical lymphadenopathy. Early recognition will minimize unnecessary investigation and prolonged empirical treatment.

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