

## KIKUCHI-FUJIMOTO DISEASE IN YOUNG MALE : A CASE REPORT

Bharti Taksande, U. Jajoo

Department of Medicine, Mahatma Gandhi Institute of Medical Science, Sevagram, Wardha, Maharashtra - 442102, India

**Abstract:** A 23-year-old male presented with fever, cough and lymphadenopathy of one months' duration. Lymph node biopsy revealed a diagnosis of Kikuchi's disease. Although the disease has been recognized worldwide, to our knowledge no cases have been reported previously from Vidharbha region.

**Keywords:** Kikuchi-Fujimoto disease, Histiocytic necrotizing lymphadenitis.

### INTRODUCTION

Kikuchi-Fujimoto disease- KD (histiocytic necrotizing lymphadenitis) was first described in Japan in 1972 and is now recognized worldwide<sup>1</sup> with a higher prevalence in the east Asian population there have been sporadic case reports from Europe and America and by late 1994 there had been some 120 cases described in the world literature<sup>2-3</sup>. It is a benign disorder, predominantly affecting young women with a predilection for cervical lymphadenopathy. It is a distinct clinicopathological entity of unknown etiology. About 16.6% to 40% of patients with KD are reported to have cutaneous involvement<sup>4</sup>. No diagnostic laboratory tests are available for KD. The definite diagnosis of KD can be made reliably only via histopathologic study from an open biopsy of the affected lymph nodes. The intermingling of distinctive crescentic histiocytes, karyorrhectic debris, and plasmacytoid monocytes in the form of nodules and the paucity of neutrophils are consistent findings that should permit a confident histopathologic diagnosis of KD<sup>5</sup>.

### CASE REPORT

A 23-year-old male presented to us with fever, cough and lymphadenopathy of one month duration. Firm, discrete, non-tender lymphadenopathy involving bilateral deep cervical and left axillary groups was present. Vitals was stable. Systemic examination did not reveal unusual findings. His hematological investigations showed anemia (Hb, 8 g%), leucopenia (total leucocyte count, 3,200/cmm) and mild thrombocytopenia (platelet count, 1,10,000/cmm); the ESR was 95 mm/h and direct coombs test was negative. Urine analysis showed traces of proteins and the 24-hour urine protein was 164mg. The blood glucose, liver function tests, blood urea, serum creatinine, and ECG were normal. A chest X-ray and ultrasonography of abdomen were normal. Blood VDRL, HIV(ELISA), Mantoux test and rheumatoid factor were negative. FNAC of the axillary lymph node was suggestive of Kikuchi's disease. Hence excision biopsy was performed which revealed a dense inflammatory infiltrate and fibrosis. The infiltrate was characterized by lymphoid tissues with germinal centers and numerous eosinophils with eosinophilic microabscess formation. A proliferation of small venule-sized vessels was noted. There was no evidence of malignancy and no organisms were noted. The patient was started on antibiotics. After a follow up period of one month patient was symptomatically better. On clinical examination all the cervical and axillary glands disappeared; the antibiotics were then stopped.

### DISCUSSION

Kikuchi-Fujimoto disease (KD) is now a well-known, benign, self-limiting disease. The youngest patient described to date was 8 years, 8 month old<sup>6</sup>. KD predominantly affects females (male:female ratio 1:4), with a mean age of 30 years. The disease commonly presents with cervical lymphadenopathy, which may be painless and isolated, or accompanied by diffuse lymphadenopathy, fever, chills, myalgia and non-specific skin

lesions. Systemic symptoms are severe when extra-nodal involvement is present. Although recurrences are possible, it clears spontaneously in 1-4 month<sup>7</sup>. This disease usually involves cervical lymph nodes; however, involvement of axillary, mesenteric, mediastinal, inguinal, intraparotid, iliac, celiac, and peripancreatic lymph nodes has been reported as well<sup>3,8,9</sup>. The cause of this rare, self-limiting disease remains unknown. It may be due to a hyperimmune reaction to an infectious agent. Several infectious agents, including Epstein-Barr virus, HHV-6, HIV, Parvovirus b19, Yersinia, and Toxoplasma have been suggested<sup>7</sup>.

Clinically, tuberculous lymphadenitis remains a major differential diagnosis, especially in developing countries like India. Patients may be empirically started on antituberculous therapy (ATT), with no significant response. Biopsy of the lymph node reveals the actual diagnosis<sup>10</sup>. The clinical manifestations include lymphadenopathy, fever, cutaneous erythema, diarrhea, vomiting chest pain, arthralgia, and hepatosplenomegaly<sup>3-5</sup>. Patients with Kikuchi-Fujimoto disease may develop anemia, leukopenia, atypical lymphocytosis and raised erythrocyte sedimentation rate<sup>11</sup>. Histologically, KD has to be differentiated from India and<sup>12</sup>. KD is self-limiting, as spontaneous improvement and disappearance of symptoms frequently occur within 1 to 6 months of initial onset. However, recurrence of lymphadenopathy; and fatal cases have been reported<sup>13</sup>. Corticosteroids are suggested as an appropriate treatment<sup>12</sup>. Recent therapeutic options for KD include ciprofloxacin<sup>14</sup>, chloroquine and hydroxychloroquine<sup>15</sup>. Response to antimicrobials suggests a possible microbial etiology<sup>14,15</sup>; KD should be considered in the differential diagnosis of cervical lymphadenopathy.

### REFERENCES

1. Kikuchi M. Lymphadenitis showing focal reticulum cell hyperplasia with nuclear debris and phagocytosis. *Nippon Ketsueki Gakkai Zasshi* 1972; 35:379-380.
2. Cho KJ, Le SS, Khang SK. Histiocytic necrotizing lymphadenitis: a clinicopathologic study of 45 cases with in situ hybridization for Epstein-Barr virus and hepatitis B virus. *J Korean Med Sci* 1996; 11:409-414.
3. Irish JC, Kain K, Keystone JS, Gullane PJ, Dardick I, Kimura? Disease: An unusual cause of head and neck masses. *J Otolaryngol* 1994;23:88-91.
4. Yasukawa K, Matsumura T, Sato-Matsumura KC, Takahashi T, Fujioka Y, Kobayashi H, et al. Kikuchi's disease and the skin: case report and review of the literature. *Br J Dermatol* 2001;144:885-9.
5. Tsang WYW, Chan JKC, Ng CS. Kikuchi's lymphadenitis. A morphologic analysis of 75 cases with special reference to unusual features. *Am J Surg Pathol* 1994;18:219-231.
6. Lien Ch, Yang W, Tsai YC, Huang PH. Kikuchi's disease (histiocytic necrotizing lymphadenitis): report of one case. *Acta paediatr Taiwan* 199;40:344-347.
7. Yen A, Feameyrough P, Raimer SS, Hudnall SD EBV-associated Kikuchi's histiocytic necrotizing lymphadenitis with cutaneous manifestations. *J Am Acad Dermatol* 1997;36:342-6.
8. Thongsukasi P, Kayasut K. Histiocytic necrotizing lymphadenitis (Kikuchi's disease): clinicopathologic characteristics of 23 cases and literature review. *J Med Assoc Thai* 1999;82:812-818.
9. Madle-Samardzija N, Turkulov V, Vukadinov J, Stajnic S, Canak G. Histiocytic necrotizing lymphadenitis (Kikuchi-Fujimoto disease). *Med Pregl* 2000;53:513-516.
10. Basu D, Mutha SM. Histiocytic necrotizing lymphadenitis (Kikuchi-Fujimoto disease)- A report of four cases. *Indian J Pathol Microbiol* 2002;45:89-92.
11. Kuo TT. Kikuchi's disease (histiocytic necrotizing lymphadenitis): clinicopathologic study of 79 cases with an analysis of histologic subtypes, immunohistology, and DNA ploidy. *Am J Surg Pathol* 1995;19:798-809.
12. Martinez-Vazquez C, Hughes G, Brodon J, Alonso-Alonso J, Anibarro-garcia A, Redondo-Martinez E, et al. Histiocytic necrotizing lymphadenitis, Kikuchi-Fujimoto's disease, associated with systemic lupus erythematosus. *Q J Med* 1997;90:531-3.
13. Shih-Hua L, Wang-Sheng K, Heng-Sheng L, Wei-Shou H. Kikuchi's disease associated with lupus-like syndrome: a fatal case. *J Rheumatol* 1992;19:1995-1996.
14. Mahajan VK, Sharma NL. Kikuchi-Fujimoto disease: immediate remission with ciprofloxacin. *Int J Dermatol* 2004;43:370-2.
15. Rezaei K, Kuchipudi S, Chudni V, Ariga R, Loew J, sha BE. Kikuchi-Fujimoto disease: hydroxychloroquine as a treatment. *Clin Infect*

Correspondence: Dr. Bharati Taksande

e-mail : ankurbaru26@yahoo.com