INTRODUCTION

Histiocytic Necrotising Lymphadenitis was first described in Japan independently by Kikuchi and Fujimoto in 1972 as a benign self limiting condition usually presenting with cervical lymphadenopathy in young females of Asian decent. It was next reported from Germany in 1982. Since then it is reported from various parts of world, including Brazil, Spain, Korea, UK, Saudi Arabia and India.

CASE REPORT

A 56 years Hindu male, non diabetic, non hypertensive, non smoker, presented with history of fever for three weeks for which he was treated with different antibiotics successively including Ciprofloxacin, Amoxycillin, Cloxacillin and Cephalexin. The fever was high grade, intermittent with chills and rigor, with a rise towards the evening. It was not associated with dysuria, night sweats, bone pain or joint pain. There was no history of bleeding diathesis.

There was history of cough with scanty mucoid expectorations without any breathlessness but with one episode of haemoptysis. Patient had history of loss of appetite.

At presentation patient had history of abnormal behavior and confusion which developed one day before admission but there was no history of unconsciousness, convulsions, headache, weakness of any limb or features suggestive of any cranial nerve involvement. Since the last five years there was a history of tremors and bradykinesia and for this he was not on any medicine. There was no past history like tuberculosis or bronchiectasis.

The general physical examination revealed mild pallor. There was no edema or venous engorgement. Cervical lymph nodes were palpable in left supraclavicular region, 2-3 in number with each lymph node about 2cm in diameter, discrete, non tender, firm, mobile, not fixed to the underlying tissue and local temperature normal. Bilateral posterior cervical lymph nodes were palpable, 2 in number, 2 centimeter in diameter with similar features. No other lymph node was palpable in axilla, epitrochlear, inguinal region. Thyroid gland was normal. There was no sternal tenderness. His temperature was 102°F, pulse-126/minute, regular, BP-130/82mmHg, respiration-32/minute.

Examination of respiratory system revealed increased vocal fremitus in the left infrascapular region with bronchial breath sounds and coarse crackles. Vocal resonance was also increased in this region. In gastrointestinal system, liver was palpable 2 centimeter, firm, non-tender with sharp margins and smooth surface. Spleen was not palpable. On examination of central nervous system, patient was drowsy and disoriented. He had cog-wheel rigidity in all limbs with tremors. There was no sign of any cranial nerve palsy or weakness in any limb. Plantars were bilateral flexors. Cardiovascular system was normal.


Abstract

A 56 years adult male presented with fever for 3 weeks with neutropenia and cervical lymphadenopathy with left sided pneumonitis. Histopathology of lymphnode was consistent with Kikuchi’s Necrotizing Lymphadenitis. Kikuchi’s disease is usually a self-limiting illness characterized by pyrexia, neutropenia and cervical lymphadenopathy in young women of Asian decent. This often leads to the misdiagnosis of lymphoma or tuberculosis. The notable feature here is an older male presented with severe neutropenia and pneumonia with hypoplastic marrow.

*Associate Professor, **Assistant Professor, ***Post Graduate Trainee-3rd year, Professor, Department of Medicine; #Assistant Professor, Department of Pathology, Calcutta National Medical College, Calcutta.

Received : 29.8.2005; Revised : 22.11.2005; Accepted : 7.7.2006
Investigations done on admission revealed: TLC-250 (P-32, L-67, M-1), Hb-11g/dl, ESR-128mm in 1st hour, Reticulocytes-0.3%, Platelets-128000/dl, Chest X-ray showed consolidation in right mid zone and left lower zone (Fig. 4).

After three days of supportive therapy (I.V. fluid, Inj Cefipime 1 gm b.d. and Amikacin 500 mg, b.d., vitamins, H2 receptor blocker) bronchial breath sounds and crackles had disappeared and patient became alert and conscious. However fever and lymphadenopathy persisted for 3 weeks.

Subsequent investigations revealed gradual rise of TLC from 250/dl to 5200/dl in 2 weeks and ESR was decreased from 128mm (in 1st hr.) to 30mm (in 1st hr.). Antibiotic therapy was withdrawn after one week.

During this two week period, repeated blood culture was negative. Blood biochemistry revealed mild hyponatremia with normal sugar, urea and creatinine level. LFT was normal. Sputum examination with Gram stain showed a few epithelial cells and pus cells. Culture showed no growth. Routine urine examination showed pus cells-2-3/hpf and albumin was trace. Urine culture showed no growth. Sputum for AFB was negative. Bactec culture for AFB and IgM ELISA for Koch’s was negative. Blood ANF, anti ds-DNA and ANCA were also negative.

USG of whole abdomen revealed mild hepatomegaly with prostatomegaly. Blood for PSA was normal.

Bone marrow examination in initial phase (one week after admission) showed (Fig. 1) hypocellular marrow with relative increase in fat cells. Two weeks later it was perfectly normal (Fig. 3).

Lymph node biopsy (one week after admission) revealed areas of necrosis containing karyorhectic debris and infiltration by histiocytes. There was no definite granuloma. Features were suggestive of Kikuchi’s necrotising lymphadenitis (Fig. 2).

When reviewed after four weeks in OPD, he was normal and all lymph nodes disappeared but Cefipime 1 gm b.d. and Amikacin 500 mg, b.d., vitamins, H2 receptor blocker) bronchial breath sounds and crackles had disappeared and patient became alert and conscious. However fever and lymphadenopathy persisted for 3 weeks.

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Parkinsonian tremor and rigidity persisted.

**DISCUSSION**

Combination of left sided segmental pneumonia, pyrexia for 3 weeks, neutropenia and cervical lymphadenopathy initially suggested Septicemia from Pneumonitis. A lot of other diseases were brought forward in differential diagnosis, like Tuberculosis, Lymphoma, Toxoplasmosis, viral infection, SLE (Systemic lupus erythematosus), malignancy with secondary infection, and metastasis, even AML (acute myeloid leukemia) was thought for (preleukemic stage). Lymph node biopsy report showed multiple areas of cortical necrosis with histiocytic infiltration without any granuloma formation, which confirmed the diagnosis. Raised ESR, neutropenia, leucopenia and lymphocytosis were additive factors for suggesting Kikuchi’s disease.

Kikuchi Fujiimoto disease was first described by Kikuchi and Fujimoto independently in 1972 as a cervical lymphadenopathy. It has been recently shown to have wide variety of clinical presentations like aseptic meningitis or encephalitis, polyarthritis, hepatosplenomegaly and skin rash. The disease is self limiting and commonly occurs in young Asian women before fourth decade. It usually presents as lymphadenitis with pyrexia. Sometimes there is association with SLE, Hashimoto’s disease, mixed connective tissue disorder (MCTD), Epstein Barr virus and Parvo virus. But, so far we have checked, marrow hypoplasia or lung infection is not reported in literature.

Etiology of Kikuchi’s disease is not clearly known. Various virus (EBV, HSV, HZV, Adenovirus, Parvovirus B19, CMV), bacteria (Mycobacterium szulgai), Yersinia, protozoa- all have been reported to be associated with the disease. A recent study by Stephen JL et al suggested EBV plays an important role. Neoplastic conditions like haemophagocytic syndrome, autoimmune disorders like SLE have been proposed to play a role via cell mediated cytotoxic response.

The diagnosis is based on histopathologic finding, differentiating it from a number of infections, autoimmune diseases or lymphoproliferative diseases. Early recognition helps in avoiding unnecessary investigations and treatment. It is seen commonly in young women of Asian descent with a mean age of 30. Isolated painless lymphadenopathy (mainly posterior cervical) is seen in 83% of cases.

The unique abnormality in this case is that the disease affected an aged male of 56 years with severe neutropenia, hypoplastic marrow and lung infection alongwith cervical lymphadenitis.

**REFERENCES**


**Announcement**


For further details contact: Dr. A Ghatak, Organising Secretary, Deputy Director, Division of Clinical and Experimental Medicine, Central Drug Research Institute, Chattar Manzil Palace, Lucknow-226001.

E-mail: bpcon2006@gmail.com; ashimghatak@gmail.com
Website: www.hypertensionindia.com; Fax: 0522-2623405 / 2623938 Attention Dr. A. Ghatak
Phone: 0522-2612411-18 (FABX Extn. 4371); Mobile: 9450906511