

# Autoimmune Disorders in Association with Hairy-Cell Leukaemia

Mary John, Kapil Chattree

## Abstract

Autoimmune diseases in the form of cutaneous leucocytoclastic Vasculitis (CLCV), polyarteritis nodosa (PAN), cerebral arteritis and cryoglobulinemia are uncommon associations of Hairy Cell Leukaemia (HCL). We report a case of a 55 year old male with HCL, carcinomatous meningitis, cryoglobulinemia and cutaneous leukocytoclastic vasculitis, which preceded the diagnosis of HCL, leucocytoclastic vasculitis was confirmed on skin biopsy, the cutaneous lesions regressed on steroid therapy.

## Key Words

Cutaneous leukocytoclastic vasculitis, Hairy cell leukaemia

## Introduction

Hairy cell leukemia (HCL) is known to be associated with various syndromes, especially PAN and cutaneous leukocytoclastic vasculitis, cerebral vasculitis and chronic active hepatitis (1). There appears to be no relationship between the presence of vasculitis and the severity or progression of the underlying malignant disease. Cutaneous vasculitis usually precedes the malignant process and responds well to steroids, immunosuppression with cyclophosphamide, splenectomy and interferon alpha-2b therapy.

## Case Report

A 55 year old male presented with three weeks history of intermittent fever, generalized weakness and dry cough. Two days before admission the patient became drowsy, restless and had urinary incontinence. There was no history of vomiting, headache, seizures or any focal neurological deficit.

Examination revealed a normosthenic elderly male who was febrile with a temperature of 100° F, pulse rate of 100/min and blood pressure of 100/60 mmHg. He had marked pallor and oral thrush. He was drowsy and disoriented to time, place and person. There was no cranial

nerve palsy. There was no motor or sensory deficit. He had clinical signs of meningitis and moderate splenomegaly. His complete blood count revealed a hemoglobin of 4.7gm%, TLC of 5500/cumm, DLC N-4%, L-96%, platelet count of 2.07 lacs, ESR-110 mm/1<sup>st</sup> hour; blood urea 40mg/dl and serum creatinine 0.8mg/dl; liver function tests normal; serum calcium 8.5 mg% and LDH 432 IU/litre; Chest X-ray-normal; ECG-normal; HIV, HBs Ag-negative; blood culture-negative; CSF examination showed 660 WBCs, with 60% Lymphocytes, 40% Neutrophils, protein 138mg/dl, sugar-38mg/dl, with the corresponding blood sugar of 120mg/dl. CSF gram stain, AFB smear, India ink preparation and CSF culture were negative. A bone marrow aspiration and biopsy was consistent with a diagnosis of Hairy cell leukaemia. The patient was started on injection ceftriaxone for meningitis and fluconazole for candidiasis. On the second hospital day he developed multiple papules and plaques over the extremities which progressed to involve the trunk and face. The skin biopsy showed leukocytoclastic vasculitis. Serum cryoglobulin was positive. Patient was pulsed with intravenous steroids and subsequently started on oral steroids. The skin lesions cleared and he regained

From The Department of Medicine, Christian Medical College & Hospital, Ludhiana-141008, Punjab.

Correspondece to: Dr. Mary John, Professor, Department of Medicine, Christian Medical College & Hospital, Ludhiana, Punjab.

normal sensorium. A repeat CSF after ten days of antibiotics showed a WBC count of 60/ml. The examination of the CSF at this time showed numerous lymphocytes showing fine cytoplasmic projections, consistent with hairy cells.

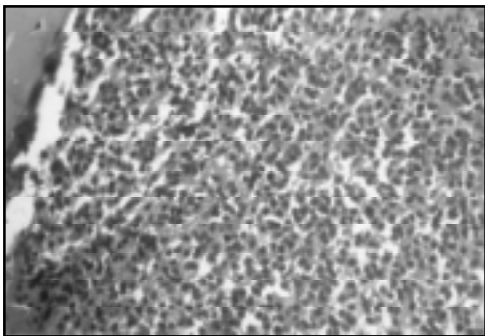


Fig. 1 Bone marrow trephine biopsy showing sheets of lymphoid cells with moderate amount of cytoplasm and clearing around individual cells (H & E X 400)



Fig. 2 Skin biopsy showing dermal vessels with fibrinoid necrosis and dense acute inflammatory infiltrate

### Discussion

Autoimmune manifestations in form of joint pains, fever, malaise, skin rash and variable visceral involvement are rare but recognized complications of HCL (1). Systemic vasculitis in the form of PAN and CLCV are also recognized unusual complications of HCL (2).

Gabriel et al (3) performed a retrospective of 129 patients with HCL at Mayo Clinic between 1976 and 1983, and identified two cases of HCL with evidence of systemic vasculitis. Immunological studies performed were unable to demonstrate the presence of shared antigen on hairy cells and endothelial cells (3).

Hasler et al when reviewing 42 cases of vasculitis coexisting with HCL, found 17 cases of PAN

and 21 cases of CLCV. In our patient also, CLCV was diagnosed along with HCL. They also observed that PAN generally occurred after the diagnosis of HCL, splenectomy or infection (4).

There was no relationship between the presence of vasculitis and the severity of malignant disease (1). Leucocytoclastic vasculitis is often preceded by infection and is frequently detected before HCL. PAN generally occurs after the diagnosis of HCL, splenectomy or infection, with positive HBs antigen (1), cerebral vasculitis (5) and chronic active hepatitis (6).

Serum immunoglobulins are generally elevated. Complement levels, antinuclear antibodies, rheumatoid factor and cryoglobulins have not shown a clear association with vasculitis in HCL (3). However, occurrence of cryoglobulinemia with HCL is very rare (7). This association presents a therapeutic challenge and emphasizes need for individualized treatment.

Our patient showed dramatic response to intravenous and oral steroids. Westbrook et al after reviewing 37 patients with HCL and vasculitis concluded that these patients respond promptly to splenectomy, corticosteroids and cytotoxic therapy in form of cyclophosphamide and interferon alfa 2b (1). Failure to recognize these complications may lead to increased morbidity and mortality.

### References

1. Westbrook CA, Golde DW. Autoimmune disease in hairy-cell leukaemia: Clinical syndromes and treatment. *Hr J Haematol* 1985; 61(2): 349-56.
2. Farcet JP, Weschsler J, Wirquin V, Divine M, Reyes F. Vasculitis in hairy-cell leukemia. *Arch Intern Med* 1987; 147(4): 660-64.
3. Gabriel SE, Conn DL, Phylly RL, Scott RE. Vasculitis in hairy cell leukaemia: Review of literature and consideration of possible pathogenic mechanisms. *J Rheumatol* 1986; 13 (6): 1167-72.
4. Hasler P, Kistler H, Gerber H. Vasculitides in hairy cell leukemia. *Semin Arthritis Rheum* 1995; 25 (2):134-42.
5. Lowe J, Russel NH. Cerebral Vasculitis associated with hairy cell leukemia. *Cancer* 1987; 60 (12): 3025-28.
6. Begley CG, Mackay IR, Bhathal PS. Another immune-mediated disease associated with hairy cell leukemia: chronic active hepatitis. *Acta Haematol* 1985; 73 (2): 104-05.
7. Raju SF, Chapman SW, Dreiling B, Tavassoli M. Hairy-cell leukemia with the appearance of mixed cryoglobulinemia and Vasculitis. *Ach Intern Med* 1984; 144 (6): 1300-02.