

HAIRY CELL LEUKAEMIA

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INTRODUCTION

Hairy cell leukaemia is a rare form of lymphoproliferative disease that affects 2% of leukaemia patients and was first described in 1958 as leukaemic reticuloendotheliosis¹. The typical presentation is pancytopenia, splenomegaly and abnormal lymphoid cells in blood with hairy cytoplasmic projections^{2,3}. The pathological hairy cell resembles in many ways B-lymphocyte⁴ as in chronic lymphocytic leukaemia (CLL). The disease occurs predominantly in older males, median age 50 years, and male/female ratio is 4:1. It is important to differentiate it from CLL because of its different behaviour and management^{2,5}.

CASE REPORT

Mr. S.Z., a 47 year old male presented in June 1985 with a history of mass in the left upper abdomen and general weakness. He had 8 cms palpable spleen, no hepatomegaly and minimal lymphadenopathy, all other systems were normal. Blood count showed a haemoglobin of 12.6 g/dl, platelets of 185000/cmm, total leucocyte count of 190,000/cmm with 3% neutrophils and 97% of lymphocytes which were mature cells of small and medium type. Bone marrow report showed "a diluted marrow with no marrow particles and marked infiltration by mature lymphocytes". A diagnosis of chronic lymphocytic leukaemia (CLL) was made and the patient was started on chlorambucil 6 mg and deltacortril 20 mg orally daily. After one month of therapy the TLC was 136,000/cmm, chlorambucil was increased to 10 mg daily. On 16 July TLC was 128,000/cmm, his general weakness had increased and there was intermittent low grade fever. The patient was lost to follow up for 4 months. He came to the hospital on 11/11/85 with a haemoglobin of 13.2 Wdl and a TLC of 128,000/cmm. As he had discontinued our treatment, he was restarted on chlorambucil 6 mg and Prednisolone 20 mg daily. In view of poor response to therapy and patient's increasing spleen size and symptoms, his clinical findings and blood cytology were critically reviewed. It was noted that many of the larger lymphocytes had hairy cytoplasmic projections; diagnosis was, therefore, revised to hairy cell leukaemia. Bone marrow cytochemistry showed these cells to be Sudan Black-Neg. PAS-Neg, a naphthal acetate Neg, but Tartrate resistant Acid Phosphatase (TRAP), positive⁴. Chlorambucil and Prednisolone were therefore continued in low doses but his drug compliance was unsatisfactory. By January 1986 his TLC count had come down to <45,000/cm, spleen had slightly regressed and his general condition had much improved. He was advised splenectomy to which he did not agree so he was managed on intermittent Chlorambucil and Prednisolone. His spleen, however, showed progressive enlargement and was about 12 cm, below the costal margin by November 1986 and his TLC was above 10^5 /cmm. As 2 interferon was not available locally so the previous therapy was continued. The patient was lost to follow up from December 1986 until 29/4/87 when he was seen again had a high grade temperature, body pains, heaviness in the abdomen and his general condition was poor. His blood counts showed a haemoglobin of 12.8 gm/dl, platelets of 78,000/cmm and TLC of 386,000/cmm. Besides other supportive treatment he was started on CVP regimen of Cyclophosphamide 500mg i/v weekly x4, Vincristine 2mg i/v weekly x 4, and Prednisolone 60 mg orally daily for six weeks. During the next 6 weeks his condition improved and on 2.6.1987 his counts were haemoglobin of 10 g/dl, Platelets of 96,000/cmm, and TLC = 72,000/ cmm. From July 1987 to September, 1988 he has had 4 courses of $\alpha 2$ interferon each of 4.5

million units subcutaneously on alternate days x 12 injections. The deviation from the recommended treatment schedule of 3 million units daily was due to non-availability of the drug here and its very high cost. During each course the patient had severe "flu like" symptoms, high temperature, body aches and low performance index. His blood counts and subjective feeling of well being had been much better after each course compared to previous Chiorambudil and Prednisolone/CVP therapy. Due to non availability of interferon, the disease process in this patient could not be controlled effectively, although his response to this drug had been quite from good. His last blood counts done in September, 1988 were haemoglobin of 6.8 g/dl, TLC of 380,000/ cmm and spleen was 24 cmm. We therefore, intend to combine leukapheresis with Chlorambucil/ Prednisolone therapy for future management of this case.

DISCUSSION

Lymphoproliferative disorders are now extensively classified on the basis of surface markers on the lymphoid cells. This has enabled better management of these disorders. Hairy cell leukaemia is a slow progressive chronic disorder and is considered a variant of CLL which it closely resembles. In fact, this patient was initially diagnosed as CLL, but due to his poor response to conventional therapy his diagnosis was re-evaluated. He had all the features of hairy cell leukaemia, i.e., hairy lymphoid cells in the blood, pancytopenia, splenomegaly and marrow that was difficult to aspirate. These cases should be differentiated from CLL because they respond poorly to chemotherapeutic agents like Chiorambucil! besides which these drugs may further suppress the normal cells of the marrow. These patients used to be treated with splenectomy and intermittent Prednisolone but the over-all treatment has been unsatisfactory. Recently dramatic results have been reported with $\alpha 2$ interferon. This newer compound is now freely available in a highly purified form, made possible by monoclonal recombinant DNA technique. In hairy cell leukaemia which had hitherto no satisfactory drug for treatment, interferon is the first agent to bring about significant control of disease process. There are multicentric reports of 90% complete/partial remissions in patients including those who did not do well on other chemotherapeutic agents and splenectomy. These patients suffer from low granulocyte values, increased infections, decreased platelets and haemoglobin levels. Treatment with interferon has resulted in improvement of some or all the above haematological variables at sometime during therapy. In our patient the results of interferon treatment were encouraging but its irregular supply and high cost precluded its continual use and potential benefits.

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