Table 1. Clinical features.

Pts	Sex/Age	Involved Nodes	Symptoms	Abnormal laboratory test	Therapy	Time from diagnosis to recovery
1	F/36	Left, cervical	Fever	ESR	Antibiotic+steroids	Six weeks
2	M/53	Cervical, axillary	Fever, fatigue	ESR	Antibiotic	Four weeks
3	F/31	Cervical	Fever, fatigue, sweats, cough	–	–	Six weeks

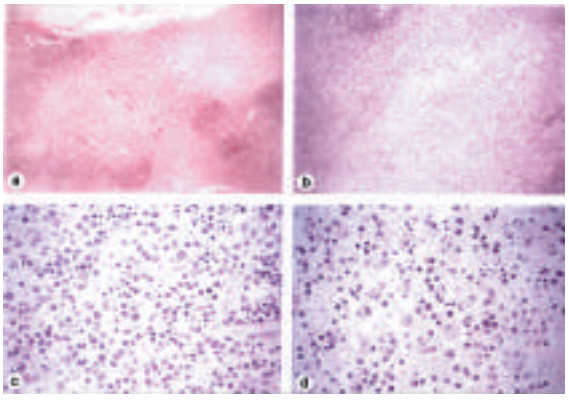

Figure 1 (left). Typical lymph node histology of Kikuchi-Fujimoto disease (case #1).
a) lymphoid tissue with multiple nodular necrotic foci; b) a necrotic area adjacent to a normal follicle; c) a necrotizing lesion with karryorrhectic nuclear debris; phagocytizing histiocytes and a few lymphocytes; neutrophils are absent; d) a necrotic area containing lymphocytes, histiocytes and nuclear debris.

SLE.[3,8-9] Therefore, prolonged follow-up of patients with KFD to reveal possible connections with SLE is suggested. The prevalence of the disease may be underestimated, since the spontaneous regression of the symptoms may hinder the diagnosis in some cases. All our patients were resident in rural areas surrounding the city, 20-30 km away from each other. We have found only two other cases of KFD from Italy reported in the literature; one was a patient with AIDS.[4,10] Thus, no estimate of the incidence of KFD in Italian populations can be attempted. We do not know whether the identification of three cases over a few years in a relatively restricted area is a casual event, possibly related to a greater attention to this disease, or whether it indicates the existence of local factors responsible for an endemic spread. Our patients recovered in about six weeks; neither relapse nor onset of other diseases has been observed with follow-ups of sixty-three, fifteen and thirteen months, respectively.

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Key words

Adenopathy, necrotizing lymphadenitis, Kikuchi-Fujimoto disease

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α-interferon as induction and maintenance therapy in hairy-cell leukemia: a long-term follow-up analysis

Sir,

The treatment of hairy cell leukemia (HCL) has improved greatly over the last two decades thanks to the introduction of first IFN[1,2] and later new purine analogs.[3,4] In particular the latter have revolutionized prognosis of HCL by increasing the number of long term complete responses.[5] The real impact on overall survival, however, is not yet clear. In this study the long term outcome of a cohort of patients treated with IFN is reported. Sixty-four patients observed between 1980 and 1996 with a histologic diagnosis of HCL, including 11 patients who had undergone splenectomy, received IFN 3MU ×3/week for 12-18 months as first line treatment. The overall response rate was 91%. Sixty-five percent achieved PR and 26% went beyond PR (13% CR and 13% GPR) (Table 1). Forty-one patients (71%) were administered IFN 3 MU/week as maintenance therapy after first line therapy (Table 1). After IFN induction therapy 20 patients relapsed and were retreated (8 under maintenance). Twenty-six patients received a second course of treatment (6 non responders and 20 relapsed after first line therapy). IFN, 2CdA, and DCF were similarly effective, with 2CdA and DCF producing higher CR and GPR rates than IFN. IFN as second line therapy (employed in patients who had previously responded to IFN) produced an overall response rate (90%) identical to that which had been observed in first line therapy. In October 1998 the median follow-up for surviving patients was 97 months. Figure 1 reports the ten year projected survival of patients achieving objective response to first line therapy (CR and GPR 100%; PR 95%) and that of non responders (NR) and clearly shows that the type of response does not affect survival.

Figure 2 shows that patients receiving IFN maintenance after IFN first line therapy had a statistically higher PFS than those not receiving maintenance therapy ($p < 0.01$).

During the last few years 2-CdA has emerged as the treatment of choice for HCL patients. However the possible effects of the profound and protracted immunosuppression caused by the drug are not completely known, thus caution is required.[6]

The results of our experience confirm that IFN is still an excellent treatment for HCL patients, producing 91% of objective responses, while having a low toxicity. In particular, IFN is not associated with an increased incidence of autoimmune diseases . As already pointed out,[7] IFN induced responses were mostly partial. The type of response does not, however, affect survival since patients achieving CR/GPR and PR had a ten year projected survival probability of 100% and 95%, respectively. The series by Rai[8] and

Table 1. Patients' features and outcome.

No.	64*	
M/F	66/16	
Mean age (range)	52	(23-73)
Spleen size - cm under costal rib (range)	8	(0-24)
% pts with splenomegaly	55	
Hb g/dL	11	(4-14)
Ht	33	(10-46)
% pts with Hb < 10 g/dL	40	
WBC $\times 10^9$/L (range)	5	(1-17)
% pts. with WBC < 3×10^9/L	51	
Plt $\times 10^9$/L (range)	93	(20- 390)
% pts with Plt < 100×10^9/L	68	
PMN % (range)	27	(6-55)
L % (range)	58	(9-83)
M %(range)	6	(2-15)
% peripheral blood HCs (range)	32	(0-87)
% pts with HCs in peripheral blood	50	
% Bone marrow cellularity (range)	56	(5-92)
% Bone marrow HC infiltration (range)	50	(5-95)
HCI (range)	0.31	(0.01-1)
NR	6(9%)	
Overall responses	58 (91%)	
PR	42 (65%)	
GPR	8 (13%)	
CR	8 (13%)	
PR-GPR-CR progression free period°	41-59-71	
Mean survival of PR-GPR-CR (months)	76-91-100	
Mean survival of SD-PD	57	

**preceded by splenectomy in 11; °41/58 (71%) received IFN maintenance therapy: response duration in maintained pts = 65 months (7-144); response duration in non maintained pts: = 42 months (9-72). HCI = hairy cell index; GPR = good partial response.*

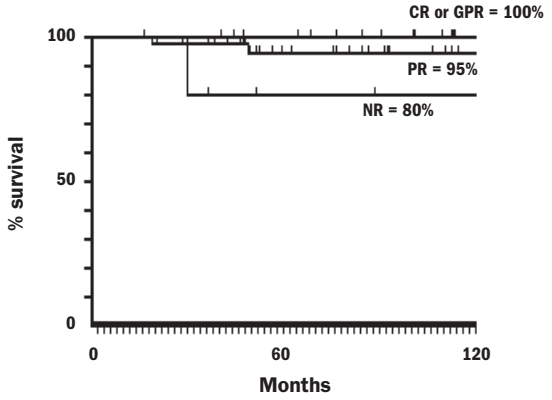

Figure 1. Survival according to type of response to first line IFN therapy.

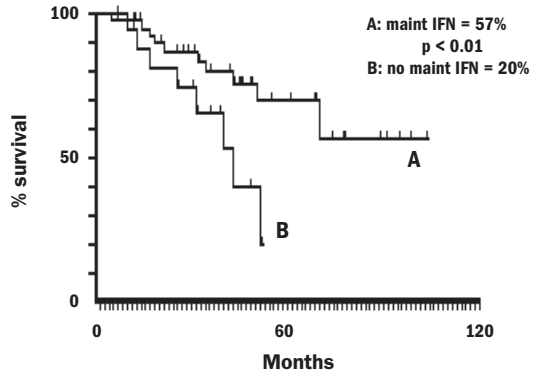

Figure 2. Progression free survival after first line IFN therapy, according to IFN maintenance therapy.

Zaja[9] also yielded similar results. A statistically significant lengthening of PFS was the result of IFN maintenance therapy, as already reported.[10]

Our experience, although limited, confirms the efficacy of purine analogs as second line therapy.

In agreement with Rai *et al*.[8] the very high projected survival probability of our series can be explained mainly by a two step therapeutic strategy made up of IFN first line therapy and purine analog salvage treatment for IFN resistant patients. Considering on the one hand our results and the reported good outcome of patients receiving IFN first,[7,8] and, on the other hand, the high CR rate and the prolonged survival obtained with front line purine analog therapy, the problem of defining a recommended first line therapy for HCL remains unsolved.

In conclusion, although the therapeutic emphasis in HCL has recently shifted to 2-CdA and DCF, IFN remains a therapeutic choice for this disease. We confirm that IFN is effective and tolerable and prolonged, reduced dosage administration may produce a long progression free period. We also confirm that achieving CR has no primary relevance in disease control and that good use of therapeutic resources may assure HCL patients a survival which is comparable to that of the normal, healthy population.

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Key words

Hairy cell leukemia, α-interferon

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Screening for hemoglobinopathies in neonates in Argentina

Sir,

So far there is very little published information about the distribution of hemoglobinopathies in Argentina.[1] Historically the Argentine population is predominantly composed of individuals of Spanish, Aboriginal and mixed ancestry. There was an important wave of Italian immigration between 1876-1925, when 2,145,000 Italians arrived in our country, and another between 1947-1951 when a further 400,000 arrived. These immigrants settled especially in the provinces of Buenos Aires, Santa Fe, Cordoba, Mendoza and Entre Rios.[2] The population under study comes from the city of Rosario, in the south of Santa Fe.

One thousand unselected, umbilical cord blood samples collected with EDTA from consecutive neonates (Hospital Provincial del Centenario), were submitted daily to: identification of unstable hemo-