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TREATMENT OF JUVENILE CHRONIC GRANULOCYTIC LEUKEMIA*

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Although the most consistent feature of reported cases of juvenile chronic granulocytic leukemia (JCGL) is the poor response to chemotherapy, the success of treatment with sequential subcutaneous cytarabine and oral mercaptopurine is encouraging[1]. Here we would like to report a patient with the longest survival thus far, treated with cytosine arabinoside (Ara-C) and 6-mercaptopurine (6-MP).

Case Report

A 9-month-old boy came to the Hacettepe Children's Hospital in April 1978 with generalized lymphadenopathy and abdominal swelling. Physical examination revealed a chronically ill child with delayed development (weight 5300 gm). He had an eczamatoid facial rash. The spleen was palpable 10 cm below the left costal margin and the liver 5 cm below the right costal margin. Moderate enlargement of the cervical lymph nodes was noted. The hematological and clinical findings on admission and subsequent follow-ups are summarized in Table I. Bone marrow aspirates were hypercellular with a predominance of granulocytic series in all stages of maturation and an absence of megakaryocytes. The hemoglobin F concentration was 2% and leukocyte alkaline phosphatase value was markedly reduced (12 U, normal 50-150 U). Because of a technical failure, results of the chromosomal analysis were not available. Treatment with intravenous Ara-C 3 mg/kg weekly and oral 6-MP 1.5 mg/kg daily was started. Since clinical improvement was noted and the WBC count returned to normal, chemotherapy was stopped in September 1978. In January 1979 the WBC count was found to be increased and the liver and spleen had become enlarged. Combined chemotherapy was resumed with the same schedule and was given continuously for 30 months after the start of therapy (Table I). During this period the patient gained weight and developed normally. His clinical and hematological findings were improved, and he is still in complete remission 18 months after the cessation of therapy.

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TABLE I

Hematological and Clinical Findings in a Patient with Chronic Granulocytic Leukemia on Admission and Subsequent Follow-Ups

Date	Hb g/dl	WBC/ml	Peripheric Blood Smear %							Platelet /μl	Liver cm	Spleen cm	Treatment Ara-C	Treatment 6-MP
			P	L	MY	ME	M	E	BL					
April 4, 1978	3.45	36,600m	65	15	14	3	1	2	2	20,000	5	10	+	+
June 5, 1978	11.67	4,400	16	62	18	–	–	4	–	N	4	5	+	+
July 7, 1978	9.5	19,100	40	35	20	3	2	–	–	N	3	5	+	+
September 14, 1978	10.2	8,600	36	54	–	4	–	6	–	N	3	4	–	–
January 31, 1979	8.4	32,300	50	37	6	3	3	1	–	44,000	5	5	+	+
January 4, 1980	12.7	9,800	82	15	2	–	–	1	–	N	1	0	+	+
September 4, 1980	10	12,270	57	24	14	2	–	3	–	N	0	0	–	–
December 11, 1980	12.3	8,500	70	24	4	–	–	2	–	N	0	0	–	–
December 2, 1981	12.79	18,900	80	8	4	–	8	–	–	N	–	–	–	–
March 23, 1982	13.11	14,700	78	22	–	–	–	–	–	N	1.5	–	–	–

P: Polymorpholeukocytes
L: Lymphocyte
MY: Myelocyte
ME: Metamyelocyte
M: Monocyte
E: Eosinophil
BL: Blast
N: Normal
O: Nonpalpable
–: Not administered

Discussion

Chronic myeloctic leukemia of the juvenile type is a rare form of childhood leukemia. Under the age of three years, splenomegaly and hyperleukocytosis suggest the diagnosis of JCGL. Lymphadenopathy, hepatosplenomegaly and skin rashes are also predominant features as seen in our patient.

JCGL is clearly distinguished from the adult type by well described clinical and hematological findings[2]. However, it has a poor prognosis with a median survival of less than 2 years[2,3]. In previously reported cases chemotherapy with either single or multiple agents has been tried with limited success in inducing remission and prolonging survival. Since there is some experimental evidence that suggests that JCGL should be classified as a variant of myelomonocytic leukemia[4], therapeutic approaches with a combination of subcutaneous cytarabine and oral mercaptopurine exerted some control over the disease. However, prolongation of survival was short[1]. In our patient we obtained the longest survival (48 months). More recently successful treatment of JCGL with marrow transplantation was reported in a 46-month-old boy[5]. Because a marrow transplant can be used only in a small number of patients who have a suitable donor, we believe that even though one case, it is worthwhile to note that our protocol of treatment seems to be more convenient and promising.

Summary

A 9-month-old boy with juvenile chronic granulocytic leukemia was treated with cytosine arabinoside and 6-mercaptopurine. Although in these patients the median survival time is very short, our patient has survived for 48 months since the diagnosis and is still free of disease. Since effective treatment still remains a problem in JCGL, our protocol of treatment seems to be promising.

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