

but chromosome 7 abnormalities, which are frequently found in MDS and AML/TMDS,⁷ have not been reported. Del 7q implies loss of genes, some of which play a major role in myeloid differentiation and have putative tumor suppressor function.¹⁰ We did not find 17p aberrations in any metaphases.

In conclusion, this case shows that ACC is a sign of dysplastic maturation affecting not only mature and semimature cells of all hematopoietic lineages, but also stem cells.

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Key words

Chromatin clumping, acute myelogenous leukemia, trilineage leukemia

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Familial hairy cell leukemia: a HLA-linked disease or farmers-linked disease?

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In the 16 cases of familial HCL published, different HLA have been found. Although specific HLA antigens were found to have the same structure in some cases that suggests a genetic predisposition to HCL. Environmental factors, specifically farming labours, were implicated in too. We add two cases (father and son, both farmers), Their HLA haplotype has not been described, but the type A2Bw4Bw6 and Bw6, presented in father and/or son, have been.

A 70-year-old man was admitted to hospital for excessive sweating. The patient worked as a farmer. The physical examination revealed splenomegaly. The man's leukocyte count was $5.6 \times 10^9/L$, platelet count was $96 \times 10^9/L$, and he had a hemoglobin concentration of 11.8 g/L. The differential count showed 20% hairy cells CD19, CD20, CD25, CD11c. Bone marrow aspirate and biopsy were diagnostic of hairy cell leukemia (HCL). He was treated with α_2a -interferon, and remains in remission for five years since the diagnosis. Three years after his father's diagnosis, the 27-year-old son of the patient described above, was found to have thrombocytopenia. His physical examination revealed splenomegaly with bilateral axillary lymphadenopathy. The son was also a farmer. His leukocyte count was $10.3 \times 10^9/L$, with a platelet count of $83 \times 10^9/L$ and a hemoglobin of 14 g/L. The differential count revealed 15% of hairy cells CD19, CD22, DR, CD20, CD23, CD25, CD11c, CD103 and FMC7 positive. Bone marrow aspirate and the biopsy were again diagnostic for HCL. The son was given 2-deoxycoformycin, and has now been in remission for two years since his diagnosis. HLA typing was carried out with standard procedures using a microcytotoxicity assay in both cases (Table 1).

Table 1. HLA typing.

Son	A80	Cw6	B45	Bw6	DR9	DR53	DQ2
	A32	C-	B67	Bw6	DR3	DR52	DQ2
Father	A80	Cw6	B45	Bw6	DR9	DR53	DQ2
	A2	C-	B51	Bw4	DR14	DR52	DQ5

Exposure to chemicals, occupational factors, ionizing radiation^{1,2} and heritability³ have been reported

to be risk factors for HCL. Recently, an elevated risk for developing HCL has been reported in farmers.^{1,2} Familial HCL, defined as the occurrence of HCL among numerous first degree family members, has previously been reported in only seven families.⁴⁻¹⁰ Analyzing the HLA typing in these families, similarities has been found in the HLA types, raising the possibility that HCL is an HLA-linked disease. In contrast, no specific HLA antigens have been found in unrelated cases of HCL, but an increased frequency of B17 and DR11 antigens compared to those found in the normal Caucasian population, has been recorded.¹¹ In familial cases, there were different haplotypes reported to be specific for the disease: type A1 A3 B8 B149; type A1 B78; type A3 A9 B7 Cw610; type A3 B3 B7 DR2 7; type A2 Bw4 Bw62(15)Cw1DR4 DRw53 DQ33; and type A3 B7 or A2 Bw4 and Bw64. It is interesting that the type A3 B7 and A2 Bw4 Bw6 were reported in various cases and the type Bw6 was common in all the cases reported by two authors.^{4,5} The hypothesis that HCL is an HLA-linked disease was, therefore, considered, but not proven.

We present two patients (father and son); HLA typing of our patients showed the haplotype A80 B45 Bw6 Cw6 DR9 DR53 DQ2. This haplotype has not previously been seen in reported cases of familial HCL. The father's HLA showed an interesting association of HLA type A2Bw4Bw6, previously reported.^{4,5} We feel that our findings strengthen the possible association between these antigens and the development of HCL. However, environmental factors could play a role in the genesis of HCL within families. It has recently been published that the most frequent occupation among 48 Swedish men and women with HCL was farming or gardening (31%) (39% of the men).² Our two patients were farm workers exposed to suspected carcinogens. The existence of some HLA antigens in common suggests a familial predisposition to the disease,³ however, familial HCL is not associated with a specific HLA haplotype; the role of an environmental risk factor, to which the affected members of the families were exposed, cannot be excluded either.

Key words

Familial hairy cell leukemia, hairy cells, HLA typing, HLA-linked disease

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Tumor burden and serum level of soluble CD25, CD8, CD23, CD54 and CD44 in non-Hodgkin's lymphoma

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We studied the value of soluble CD25, CD8, CD23, CD54 and CD44 serum levels as tumor burden markers in lymphoma. Soluble CD25 compared with the others sCD and the usual serum factors (albumin, lactate dehydrogenase, β_2 -microglobulin, uric acid and C-reactive protein), showed the strongest correlation with the Ann Arbor stage and the number of affected localizations. sCD25 level is the most sensitive serum marker for tumor burden in lymphoma.

Tumor burden is an important prognostic factor in lymphoma.¹ Tumor burden is roughly approximated by physical examination, bone marrow biopsy and imaging techniques. Estimating tumor burden by a non-invasive method is an old interest in oncology. Over the last decade, the measurement of soluble receptors levels has been explored as an additional tool for the assessment of tumor burden and prognosis in patients with lymphoma.^{2,3} Several investigations in different histologic subtypes of non-Hodgkin's lymphoma (NHL) have demonstrated the good association between soluble serum interleukin-