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Cytomegalovirus retinitis revealing a leukemic γ/δ t-cell lymphoma

Sir,

We report a case of γ/δ T-cell lymphoma (γ/δ TCL) in a patient treated for sarcoidosis, revealed by cytomegalovirus (CMV) infection. Clinical presentation was atypical with lymph node and leukemic involvement. To our knowledge, such a clinical history has never been reported.

Case report. A 44-year-old man was admitted to

Table 1.

	1994	1996
<i>Peripheral blood lymphocytes</i>		
Total count ($\times 10^9/L$)	0.88	1.0
% CD3	60	83
% CD4	33	3
% CD8	32	2
% CD56	19	Not done
% γ/δ T cells	0	74
<i>Lymph nodes</i>		
Morphology	Lymphoid hyperplasia B and T	Diffuse pleiomorphic lymphoid cell proliferation with irregular nuclei
Immunostaining	Normal	γ/δ TCR+, $\alpha\beta$ TCR-, CD3+, CD43+, CD45+, CD4-, CD8-, CD5-

hospital in May 1994, because of pruritic papulous skin lesions. Cervical and inguinal nodes were enlarged. Hematologic tests showed eosinophils $1.24 \times 10^9/L$; lymphocytes: $0.88 \times 10^9/L$; ESR: 10 mm per hour; serum angioconvertase activity: 266 IU/mL (N=45-145). The immunophenotypic profile of peripheral blood lymphocytes (PBCL) evidenced an increase in CD56+ without γ/δ T cells (Table 1). Skin biopsy showed a sarcoid-like granulomatous inflammation. The histologic characteristics of the cervical lymph nodes were lymphoid B and T hyperplasia. There were no atypical lymphoid cells. A diagnosis of sarcoidosis was made. Prednisolone was started, with effect on the skin lesions.

In September 1996 this patient developed sudden visual loss. Ophthalmologic examination and serological tests evidenced a CMV retinitis. There was generalized lymphadenopathy and splenomegaly, but not hepatomegaly. Lumbar puncture was normal. The HIV test was negative. The immunophenotypic profile of PBCL evidenced CD4 and CD8 lymphocytopenia with 74% of γ/δ T cells+. A CT scan revealed mediastinal lymph nodes and homogenous splenomegaly. The bone marrow biopsy was normal and CD3 immunostaining did not reveal neoplastic infiltrating. Treatment with foscarnet induced stabilization of the retinal lesions. Histopathologic examination of a lymph node showed a diffuse pleiomorphic CD3+, CD4-, CD5-, CD8-, CD43+, CD19-, CD56-, CD45+ lymphoid cell proliferation. The lymphoid cells were atypical with abundant cytoplasm and irregular nuclei. The cells were γ/δ TCR+, TiA-1+, granzyme B+. Epstein-Barr virus was detected (LMP-1+). Polymerase chain reaction revealed a clonal rearrangement of γ T cell receptor gene and heavy (IgH) chain immunoglobulins. Sequential chemotherapy was started with no improvement. Monoclonal gammopathy (MG) IgG λ appeared after four

months. The patient died 7 months later from systemic CMV infection.

A relationship between simultaneous sarcoidosis and lymphoma has been described as sarcoidosis-lymphoma syndrome¹ but no case of $\gamma\delta$ TCL has been reported. In this case, it seems that the patient really had sarcoidosis preceding $\gamma\delta$ TCL. Indeed lymph nodes, bone marrow and skin lesions were analyzed in May 1994 when they showed no lymphoma cells. Our patient also had a CMV retinitis. Such a complication may occur in the course of Hodgkin's and non-Hodgkin's lymphomas (NHL)² but has never been reported in $\gamma\delta$ TCL. This may be due to the rareness of this lymphoma and also to the short survival of affected patients.

The extrahepatosplenic manifestations seen in our patient have rarely been reported in $\gamma\delta$ TCL.³ Only three cases with lymph node involvement have been described^{4,5} with no initial leukemic involvement.

MG may be encountered in various lymphomas, but it has never been described with $\gamma\delta$ TCL. This abnormality could be secondary to the impaired immunity we observe in NHL, but the clonal rearrangement of IgH in this patient remains questionable.

Finally, the association between sarcoidosis, CMV retinitis and MG is frequent in NHL's patients. This case is original, given the characteristics of lymphoma with an extrahepatosplenic $\gamma\delta$ T cell tumor proliferation.

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Key words

$\gamma\delta$ lymphoma, sarcoidosis, lymph nodes, cytomegalovirus, retinitis

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