Cytomegalovirus retinitis revealing a leukemic γδ-TCR+ T-cell lymphoma

Sir,

We report a case of γδ T-cell lymphoma (γδ-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 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 angiotensinogen activity: $266$ IU/mL ($N=45-145$). The immunophenotypic profile of peripheral blood lymphocytes (PBCL) evidenced an increase in CD56+ without γδT 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 γδ 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 pleomorphic CD3+, CD4+, CD5+, CD8+, CD43+, CD45+ lymphoid cell proliferation. The lymphoid cells were atypical with abundant cytoplasm and irregular nuclei. The cells were γδ TCR+, TIA-1+, granzyme B+. Epstein-Barr virus was detected (LM-P-1+). Polymerase chain reaction revealed a clonal rearrangement of γ T cell receptor gene and heavy (IgH) chain immunoglobulins. Sequential chemotheraphy was started with no improvement. Monoclonal gammopathy (MG) IgG λ appeared after four months.

### Table 1. Peripheral blood lymphocytes

<table>
<thead>
<tr>
<th></th>
<th>1994</th>
<th>1996</th>
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<tbody>
<tr>
<td>Total count ($x10^9/L$)</td>
<td>0.88</td>
<td>1.0</td>
</tr>
<tr>
<td>% CD3</td>
<td>60</td>
<td>83</td>
</tr>
<tr>
<td>% CD4</td>
<td>33</td>
<td>3</td>
</tr>
<tr>
<td>% CD8</td>
<td>32</td>
<td>2</td>
</tr>
<tr>
<td>% CD56</td>
<td>19</td>
<td>Not done</td>
</tr>
<tr>
<td>γδT cells</td>
<td>0</td>
<td>74</td>
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</tbody>
</table>

### Lymph nodes

<table>
<thead>
<tr>
<th></th>
<th>Morphology</th>
<th>Lymphoid hyperplasia</th>
<th>Diffuse pleiomorphic lymphoid cell proliferation with irregular nuclei</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Normal</td>
<td>γδ TCR+, αβ TCR+, CD3+, CD43+, CD45+, CD4+, CD8+, CD5+</td>
<td></td>
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</table>

### Immunostaining

<table>
<thead>
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<tbody>
<tr>
<td></td>
<td>γδ TCR+, αβ TCR+, CD3+, CD43+, CD45+, CD4+, CD8+, CD5+</td>
</tr>
</tbody>
</table>

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References

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

A relationship between simultaneous sarcoidosis and lymphoma has been described as sarcoidosis-lymphoma syndrome, but no case of γδTCL has been reported. In this case, it seems that the patient really had sarcoidosis preceding γδTCL. 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) but has never been reported in γδTCL. This may be due to the rarity of this lymphoma and also to the short survival of affected patients.

The extrahepatosplenic manifestations seen in our patient have rarely been reported in γδTCL. Only three cases with lymph node involvement have been described, with no initial leukemic involvement. MG may be encountered in various lymphomas, but it has never been described with γδTCL. 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 γδ T cell tumor proliferation.

Key words
γδ lymphoma, sarcoidosis, lymph nodes, cytomegalovirus, retinitis

Correspondence
Prof. Pierre-Jean Weiller, Service de médecine interne, CHU Timone, 264 rue saint Pierre, 13385 Marseille cedex 5, France. Tel.: international +33-4-91386039 – Fax: international +33-4-91347401 – E-mail: pweiller@ap-hm.fr

References