Case report

Primary lymphoma of the central nervous system and HTLV-I infection

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Abstract—Only a few cases of AIDS-related primary lymphomas of the central nervous system (CNS) show a T-cell phenotype. We have recently studied two intravenous drug users with HIV infection who had primary CNS T-cell lymphomas. In both cases, the enzyme immunoassay (EIA) for HTLV gave a positive result. In the first case, study by western-blot (WB) and specific PCR confirmed the human T-cell lymphotropic virus type I (HTLV-I) infection and serological study by EIA for HTLV of his mother was negative. In the second case, analysis of ante-mortem serum samples by two different WBs showed an indeterminate pattern suggestive of HTLV-I infection, but adequate samples for PCR were not available. We speculate about the possibility that the horizontal transmission of HTLV-I infection could have facilitated the development of a primary CNS T-cell lymphoma in these HIV patients, although they cannot be strictly considered as ATLL cases.

Key words: Lymphoma; central nervous system; HTLV-I.

AIDS-related primary lymphomas of the central nervous system (CNS) are most frequently Epstein–Barr virus-positive B-cell lymphomas [1]. Only a few cases show a T-cell phenotype. We have recently studied two HIV patients who had primary CNS T-cell lymphomas.

Both cases involved Spanish men, aged 29 (case 1) and 34 years (case 2), who were intravenous drug users (IVDUs) and were HIV-infected. Neither was affected by lymphoma beyond the CNS. In both cases, the pathological diagnosis was T-cell lymphoma, CD45(+), CD45RO(+), CD20(−), made by biopsy (case 1)

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Table 1. Results of serological investigation for HTLV-I infection

<table>
<thead>
<tr>
<th></th>
<th>EIA</th>
<th>DBL 2.3-WB reactivity</th>
<th>CBS-WB reactivity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1 (serum)</td>
<td>Positive</td>
<td>p19, p24, p26, p28, p36, rgp21, rgp46-I</td>
<td>ND</td>
</tr>
<tr>
<td>Case 1 (cerebrospinal fluid)</td>
<td>Positive</td>
<td>p19, p24, p26, p28, p36, rgp21, rgp46-I</td>
<td>ND</td>
</tr>
<tr>
<td>Case 2 (serum)</td>
<td>Positive</td>
<td>p24, p28, rgp21</td>
<td></td>
</tr>
</tbody>
</table>

EIA: enzyme immunoassay (Cambridge Biotech, Ortho, Raritan, USA).
DBL 2.3-WB: DBL version 2.3 western blot (Diagnostic Biotechnology Ltd., Singapore).
ND = not done.

and at necropsy (case 2). In both cases, the enzyme immunoassay (EIA) for HTLV (Cambridge Biotech, USA) gave a positive result. Neither case had neurological symptoms associated with tropical spastic paraparesis/HTLV-I-associated myelopathy.

In case 1, serological study by western blot (WB) confirmed the human T-cell lymphotrophic virus type I (HTLV-I) infection; the presence of the retrovirus was detected in peripheral mononuclear cells by means of a polymerase chain reaction (PCR) with specific primers (SK 43–44/45; SK 54–55/56). PCR analysis of paraffin tumour tissue samples yielded negative results. However, WB analysis of the cerebrospinal fluid revealed the same reactive bands as in the serum (Table 1). Serological study by EIA for HTLV of his mother was negative.

In case 2, analysis of ante-mortem serum samples obtained by two different WBs showed an indeterminate pattern suggestive of HTLV-I infection (Table 1). Adequate samples for PCR were not available in this case. We could not study his parents.

HTLV-I is the causal agent of adult T-cell leukaemia/lymphoma (ATLL). This process has been described mostly in endemic areas, but also sporadically in the US and European countries, including Spain [2]. Clinically, 75% of cases of ATLL are leukaemia and 25% are lymphomas without peripheral blood involvement. The CNS is frequently affected in ATLL, although no cases of primary CNS lymphomas have been reported.

According to the criteria proposed by Levine et al. for the diagnosis of ATLL [3], case 1 could well be considered as possible ATLL. However, we have not been able to demonstrate the presence of clonally integrated HTLV-I in neoplastic cells, a feature that is specific for ATLL. Failure to demonstrate the presence of HTLV-I in tumour tissue could be related to the lower sensitivity of PCR from paraffin samples.

It is difficult to ascertain whether HTLV-I infection may have played a role in the development of CNS lymphoma. The casual association of primary CNS T-cell lymphomas and HTLV-I infection is possible in our first case, although not very likely. Primary CNS T-cell lymphomas are rare, representing approximately 4%