Sensory Perineuritis and Non-Hodgkin’s T-Cell Lymphoma

Sensory perineuritis is a rare disorder that develops mainly in cutaneous nerves [1-4]. A pathomechanism underlying the perineuritis is unclear, although an immune-mediated process affecting a perineurium has been suggested. Various types of neuropathies are known to be associated with lymphomas [5, 6]. Here we describe a patient with the sensory perineuritis who developed a non-Hodgkin’s T-cell lymphoma.

A 57-year-old man first noticed numbness in both of his feet. The numbness gradually got worse. The patient was examined 4 months after the onset. There was no lymphadenopathy. Dysesthesia and impairment of perception of light touch, pinprick, and temperature were found bilaterally in the distribution of the sural nerve and to a lesser extent in the distribution of the plantar nerve. Tinel’s sign was elicited when the sural nerve was tapped. There was no weakness. The tendon reflexes were bilaterally diminished at the ankles. The cerebrospinal fluid protein was 50 mg/dl without pleocytosis. Blood cell count, urinalysis, blood chemistry, serum M protein, immunoglobulins, complements, autoantibodies, tumor markers, serum antibodies to human T-cell lymphotropic virus type 1 (HTLV-1) and human immunodeficiency virus (HIV), and chest X-ray and CT were normal or negative. Nerve conduction studies were normal except for the bilateral sural nerve in which no sensory nerve action potential was obtained. Sural nerve biopsy revealed a perineuritis with a marked loss of the myelin...
Fig. 1. Transverse sections of the sural nerve. Fibrotic thickening of the perineurium (asterisks) is found with nearly total loss of myelinated fibers (a, b). Infiltration of mononuclear cells is focally observed in the epineurium adjacent to the thickened perineurium with no evidence of necrotizing vasculitis (b). A very few remaining myelinated fibers presented with axonal degeneration as demonstrated in the teased preparations (data not shown). Electron microscopically, the thickened perineurium was composed of normal or degenerative perineurial cells, irregularly thickened and interrupted basement membrane, fibroblasts, collagen fibers, and microfilamentous and amorphous materials (data not shown). The density of unmyelinated fibers was 21,552/mm². Semithin sections of the resin-embedded sural nerve stained with toluidine blue, a x 97, b x 256.

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ed fibers (fig. 1). Most of the mononuclear cells infiltrating in the epineurium were CD8+ lymphocytes. No significant deposition of immunoglobulin or complements was found in the nerve. Serum immunoglobulins from the patient did not react with frozen nerve sections from a normal control (data not shown). The patient was diagnosed as having the sensory perineuritis, and was treated with steroid pulse and oral prednisolone therapies, resulting in improvement of the symptoms. Twenty months after the onset of the perineuritis, the patient developed swelling of the cervical, axillary, and inguinal lymph nodes. There was no exacerbation in the neurologic findings. The biopsy of the lymph node revealed non-Hodgkin’s malignant lymphoma (diffuse, mixed type). The lymphoma cells were CD4+ T-cell type as demonstrated in immunohistochemical and flow cytometric studies (data not shown). Southern blot analysis of DNA prepared from the lymph node tissue revealed rearrangements of the T-cell receptor (ß and ë chain genes, but not of the immunoglobulin genes, confirming the T-cell nature of the lymphoma (data not shown). Combination chemotherapy composed of pirarubicin, cyclophosphamide, vinblastine, and prednisolone diminished the enlarged lymph nodes. Our patient with the sensory perineuritis developed non-Hodgkin’s T-cell lymphoma. The infiltrating cells in the biopsied nerve were mainly CD8+ T cells, but not the lymphoma cells that had a CD4+ T-cell nature. There was no evidence of infection of viruses such as HTLV-1 which could be involved in the causation of T-cell lymphomas.

Perineuritis is found to occur sometimes with systemic diseases [7]. A patient with perineuritis and disseminated carcinoma was described [4]. However, the association of the perineuritis and lymphoma has not been reported as yet. In addition, pure sensory neuropathy complicating lymphomas is very rare [6]. The relation between the perineuritis and the lymphoma in our case could not be clarified. However, excess risk of lymphomas has been reported in populations with altered immunity including a variety of systemic or tissue-specific autoimmune states [8-10]. We suggest the presence of the immunological derangement that may be related to the pathogenesis of both the perineuritis and lymphoid neoplasia. The perineuritis, a unique tissue-specific autoimmune state, may be caused by disturbance in the T-cell regulatory system, which could also result in a predisposition to the emergence of T-cell lymphoma.
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References
