Antiretroviral Drugs (principally zidovudine) have been used with success in the treatment of myelopathy associated with human T-lymphotrophic virus 1 (HTLV-1) (tropical spastic paraparesis–HTLV-1–associated myelopathy). In clinical reports and murine experiments, the retrovirus HTLV-1 has been implicated as a causative agent of Sjögren syndrome (SS). Moreover, a recognized complication of primary SS is a myelopathy, which has been shown in case reports to respond to immunosuppressive treatment.

We report the case of a patient with a rapidly progressive myelopathy in whom HTLV-1 infection and SS (probably secondary to HTLV-1) were treated successfully with combined immunosuppressive and antiviral therapy.

A 48-year-old Haitian woman living in Switzerland for the past 20 years was admitted to investigate a rapidly progressive spastic tetraparesis. Her medical history began 6 years earlier with a dryness syndrome. Three years later, she noticed that her sense of smell was impaired. Six months prior to admission, she started having difficulties walking. Her gait rapidly deteriorated, and by the time of admission, she could just stand and make a few broad-based spastic steps without assistance. She also had arthralgias in her shoulders and knees and had monocular blurred vision of the right eye. She did not have any voiding problems.

Clinical examination revealed anosmia (Sniffin’ Sticks test kit, Threshold Discrimination and Identification measures score = 8; rhinoscopy results were unremarkable) with partial preservation of taste, a spastic tetraparesis with generalized limb hyperreflexia, bilateral extensor plantar responses, and intermediate uveitis of the right eye.

Laboratory investigations showed the presence of anti-glycosaminoglycan and anti-envelope HTLV-1 protein in the serum and cerebrospinal fluid. In addition, quantitative DNA polymerase chain reac-
tion results were positive for HTLV-1 (but not for HTLV-2) in the serum and cerebrospinal fluid. Human T-lymphotrophic virus 1 reverse transcriptase was undetectable. There were strongly elevated levels of blood antinuclear antibodies (1/1280; normal value <1/80), SS antigen A antibodies (153 U; normal value <20 U), and SS antigen B antibodies (155 U; normal value <20 U); cerebrospinal fluid SS antigen A antibodies were also strongly positive (85 U). The cerebrospinal fluid was mildly inflammatory, with 20 cells/µL (94% lymphocytes) and 60 mg/dL of cerebrospinal fluid proteins. An increased IgG index with an intrathecal production of HTLV-1 antibodies was detected by dot-blot analysis.

Flow cytometry and T-cell receptor rearrangement analysis showed a discrete CD4+ T-lymphocyte monoclonal population in peripheral blood. Typing of HLA-AB was performed by microlymphocytoxicity analysis, HLA-DRB1 generic typing by reverse-transcription polymerase chain reaction with sequence-specific oligonucleotide probes, and DQ generic typing by polymerase chain reaction with sequence-specific primers. The complete HLA typing of our patient was A33, A34; B18, B44; DRB1*1302; DQB1*02, DQB1*06.

Accessory salivary gland biopsy showed lymphocytic sialadenitis suggestive of SS.

Magnetic resonance imaging of the spine showed extensive contrast-enhanced lesions in cervical and thoracic regions (Figure, A). Magnetic resonance imaging of the brain showed multiple non–contrast-enhanced lesions of white matter (right internal capsule, left and right centrum semiovale).

The patient received a high dose of corticosteroids (500 mg of methylprednisolone intravenously) for 6 days followed by oral prednisone (1 mg/kg) associated with antiviral bitherapy of lamivudine and tenofovir. After 1 month, prednisone was tapered off over 1 month to a maintenance dose of 5 mg/d, and treatment with mycophenolate mofetil was introduced to a dose of 1500 mg/d.

The clinical evolution was spectacular: after 1 month of treatment, spasticity and weakness improved; the patient was able to walk on her own for more than half of a mile and could climb stairs. Anosmia and the dryness syndrome remained unchanged. Cerebrospinal fluid and magnetic resonance imaging results of the brain and spine normalized. After 48 months of follow-up, minimal spasticity remained, tendon reflexes normalized on the right side but remained slightly brisk on the left side, and plantar reflexes were flexors. The patient was able to walk and run, and everyday activities were normal. Magnetic resonance imaging results of the spine normalized (Figure, B). In addition, quantitative HTLV-1 DNA polymerase chain reaction showed a 3-fold reduction in peripheral blood while the CD4+ T-cell clone level remained stable.

**COMMENT**

Infection with HTLV-1 has been associated not only with TSP/HAM but also with SS.2,3 Human T-lymphotrophic virus 1 was found in the biopsy specimens of salivatory glands of patients with SS, and 25% of the patients with TSP/HAM also showed concomitant SS.8

Our patient’s symptoms and deficits could be compatible with TSP/HAM: a tetraparetic syndrome predominant in the inferior limbs and associated with arthral-
Correspondence: Fabienne Perren, MD, Department of Neurology, University Hospital and Medical School of Geneva, Micheli-du-Crest 24, CH-1211 Geneva 14, Switzerland (fabienne.perren@hcuge.ch).

Author Contributions: Study concept and design: Pot, Landis, and Perren. Acquisition of data: Pot, Vokatch, Tieryc, and Ribi. Analysis and interpretation of data: Pot, Chizzolini, Vokatch, and Perren. Drafting of the manuscript: Pot, Vokatch, Tieryc, and Ribi. Critical revision of the manuscript for important intellectual content: Chizzolini, Landis, and Perren. Administrative, technical, and material support: Pot, Vokatch, Tieryc, and Ribi. Study supervision: Chizzolini, Landis, and Perren.

REFERENCES


