

**49th ANNUAL DIAGNOSTIC SLIDE SESSION, 2008  
DIAGNOSES AND REFERENCES**

**MODERATOR: Anthony T. Yachnis, M.D.**

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**Case 2008-4**

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**Diagnosis: Human T-lymphotropic virus type 1 (HTLV-1)-associated myelopathy/tropical spastic paraparesis (HAM/TSP)**

**Comment:** The cord had diffuse, active myelitis, with degeneration of lateral, anterior, posterior and subpial white matter. Demyelination was proportional to loss of axons. Anterior horn cell bodies were largely unaffected despite the severe inflammatory and degenerative changes. The thoracic cord was the most severely affected portion. Inflammatory cells in the cord were predominantly CD8-positive T cells; there were few CD4+ or CD20+ cells. The predominance of CD8+ cells in the cord was not due to systemic depletion since CD8+ and CD4+ cell populations were normal in the spleen. CD68 and CD133 stains showed diffuse microglial activation; GFAP staining revealed diffuse gliosis. There was also marked perivascular fibrosis and inflammation in dorsal root ganglia. The brain showed minimal changes including focal inflammation only. Immunohistochemistry for HTLV-1 was negative in the cord.

From the presenter: CSF and serum serology were positive for HTLV-1 in 1989 (no further history was available). The patient thought that he had acquired HTLV-1 through unprotected sexual contact while stationed in Okinawa during the Korean War (1950-51), 54 years prior to his death. He had also traveled to the Caribbean. The patient had received treatment with interferon, IVIG, and prednisone earlier in his course, without benefit. The flattened caudates had old infarcts.

**References:**

Iwasaki Y, Koichi Sawada K, Aiba I, Mukai E, Yoshida M, Hashizume Y, Sobue G: Widespread active inflammatory lesions in a case of HTLV-I-associated myelopathy lasting 29 years. *Acta Neuropathol* 2004; 108:546-551.

Matsuura E, Umehara F, Nose H, Higuchi I, Matsuoka E, Izumi K, Kubota R, Saito M, Izumo S, Arimura K, Osame M. Inclusion body myositis associated with human T-lymphotropic virus-type I infection: eleven patients from an endemic area in Japan. *J Neuropathol Exp Neurol* 2008; 67:41-49.

Moore GRW, Traugott U, Scheinberg LC, Raine CS: Tropical spastic paraparesis: a model of virus-induced, cytotoxic T-cell-mediated demyelination? *Ann Neurol* 1989; 26:523-530.