 HTLV-1 infection: the clinical spectrum widens

A neurological condition causing spastic paraparesis has long been recognised in the West Indies but it is only in recent years that the association between tropical spastic paraparesis (TSP) and human T-cell lymphotropic virus type I (HTLV-1) has been confirmed.1,2 Serological tests for HTLV-1 can now support a diagnosis of TSP in patients with atypical clinical features.

Patient 1 was born in Jamaica and came to the United Kingdom at the age of 38 years. At the age of 61 years she presented with pain in her left shoulder, a three year history of difficulty raising her arms above her head, fatigue and inability to walk long distances. On examination the only sign of both scapulae with weakness in the deltoids, triceps and biceps, without fasciculations. Distal upper limb muscle atrophy was normal. The biceps and supinator jerks were absent and there were no Babinski signs. There was a mild spastic paraparesis with increased knee jerks, diminished ankle jerks and extensor plantar responses. Sensory testing was normal.

The patient had been mildly elevated and muscle histology revealed neurogenic changes. Myelography and CSF examination were normal. There was a polyclonal increase in the serum immunoglobulins.

At first a diagnosis of motor neuron disease was considered but there was no change in her condition during the following four years and reinvestigation revealed serum antibodies against HTLV-1 in a titre of 1 in 6400. In TSP, pain is usually a precurser or preceded weakness but is characteristically confined to the lower limbs and lumbar spine. This patient is also unusual in the severity of the muscle wasting, which in TSP is seldom prominent and usually confined to the intrinsic hand muscles.

Patient 2 was born in Jamaica and came to the United Kingdom aged 42 years. He was first seen aged 63 years following a single generalised convulsion; his right plantar response was extensor but there were no other neurological signs. CT brain scan and EEG were normal. Treponemal serology was positive (VDRL 1:16, Treponema pallidum haemagglutination 1:4; TPHA positive; FTA IgG positive, IgM negative) without a history of previous venereal disease or yaws. Lumbar CSF was acellular with normal protein concentration and the tests for syphilis were all negative; Link's IgG index was 0-78 (upper limit of normal 0-58) suggesting intrauterine immunoglobulin synthesis. He was treated with a course of intramuscular procaine penicillin and regular phenytoin.

At the age of 70 years he was seen again for reassessment of his epilepsy. Only on direct questioning did he admit that his legs had become weak several years before. He had been rising from a low chair but otherwise his gait was normal.

There was a mild spastic paraparesis with brisk lower limb reflexes and the right plantar response was extensor as before. A CT brain scan was again normal. MRI scan showed several small punctate white matter lesions in both cerebral hemispheres but no abnormality of the spinal cord. Antibodies to IFTLV-1 were detected in the serum in a titre of 1 in 8000.

Patient 3 was born in British Guyana and came to the United Kingdom at the age of 34 years. At the age of 59 years she developed bilateral uveitis and was found to have positive treponemal serology (VDRL negative; TPHA positive; FTA IgG positive, IgM negative) without a past history of venereal infection. CSF contained 32 lymphocytes/mm³ and the protein concentration was raised (0·85 g/l); CSF tests for VDRL, TPHA and FTA were negative. Her vision remained steady and she took a one month course of oral doxycycline. Six months later she developed weakness and sensory loss in the legs which progressed over a year. At that time she had a spastic paraparesis with a sensory level at T10 and normal position sense. The upper limbs and cranial nerves were normal. Myelography was normal but was followed by urinary retention requiring catheterisation. The CSF on this occasion contained no cells but the protein concentration remained raised (1·0 g/l).

A diagnosis of neurosarcoidosis was considered but the patient showed no response to oral corticosteroid therapy; subsequent bronchoscopy, bronchial biopsy and Kveim test were found to be normal.

At follow up two years later her gait had deteriorated; she then developed mild bilateral nerve deafness and there was muscle wasting in both hands. Antibodies against HTLV-1 were detected in the serum in a titre of 1 in 64000. The CSF film was normal but there was a polyclonal increase in the serum immunoglobulins.

The initial detection of HTLV-1 antibodies was by gel particle agglutination assay and confirmed by more specific methods (ELISA, indirect immunoenphroetosis, IgG antibody capture radio-immunnoassay and Western blot techniques).3,4 These patients illustrate many of the recognised features of TSP. This unusual illness in each of them had been tentatively attributed to another cause: motor neuron disease (patient 1), parasagittal tumour or neurosyphilis (patient 2), neurosarcoidosis or nevritis (patient 3). The routine neurological presentations were pectoral pain and amyotrophy (patient 1), late-onset epilepsy (patient 2) and uveitis (patient 3). The diagnosis of HTLV-1 infection became available, early diagnosis will be necessary to identify patients before severe, irreversible neurological damage has occurred. This will require greater awareness of the diverse ways in which TSP may present. The full spectrum of HTLV-1 infection remains to be defined.
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