Tropical Spastic Paraparesis

There are two types of tropical myeloneuropathies that are different in aetiology and clinical features. They both occur predominantly in tropical countries, although tropical spastic paraparesis (TSP) has been described in temperate southern Japan.  

- TSP was initially described in tropical countries but has now also been identified in temperate countries (eg, southern Japan) as HTLV-1-associated myelopathy (HAM).  
- HAM/TSP predominantly affects the spinal cord, resulting in an upper motor neurone syndrome, mostly affecting the lower limbs.  
- Tropical ataxic neuropathy (TAN) is predominantly a sensory neuropathy. TAN is often seen in malnourished populations and where large quantities of cassava are present in the diet.  

HAM/TSP results in inflammation, demyelination and necrotic lesions in the spinal cord. It is a progressive disease involving the degeneration of neurons in the spinal cord, leading to a gradual paralysis of the lower limbs.

Pathogenesis

- HAM/TSP is associated with HTLV-1 infection. However, there have been some cases of TSP where evidence of HTLV-1 infection has not been found.  
- The signs of a chronic myeloneuropathy found in the West Indies had been recognised as a distinct entity for over a hundred years, it was not until 1985 that a link was first made with HTLV-1.  
- In a study in Martinique looking at the epidemiology of adult T-cell leukaemia, 59% of those patients who had TSP were found to have antibodies to HTLV-1, as opposed to 13% of controls.  
- Since then, several other studies have confirmed these findings, and a set of clinical criteria has been established to describe what is now referred to as HTLV-1-associated myelopathy/tropical spastic paraparesis (HAM/TSP).  
- Of the 10 to 20 million people in the world thought to have HTLV-1, only 1-4% will go on to develop HAM/TSP. In Japan that figure is lower at 0.25%. It is not exactly certain why this is. The outcome may be dependent upon the individual's immune response.  

Epidemiology

- Populations at risk include those in areas where the HTLV-1 virus is endemic such as the Caribbean, equatorial Africa, South America, the Seychelles and southern Japan.  
- It is estimated that 10-20 million people worldwide are infected with HTLV-1.  
- It tends to affect people from lower social classes.  
- There is a peak incidence in the third or fourth decade.  

Presentation

- Initial infection is usually asymptomatic.  
- Incubation period between infection and symptomatic disease may be many decades. Equally, symptoms can appear within months.

Symptoms

These may include:

- Gradual onset of leg weakness.  
- Loss of sensation and/or pins and needles.  
- Incontinence or urinary dysfunction. There is urinary frequency with detrusor instability.  
- Bowel dysfunction may be present.  
- There may be back pain with radiation to the legs.  
- Erectile dysfunction has been reported as a presenting feature.  
- Dermatitis or psoriasis may be present.

Signs

These are largely upper motor neurone signs:
Spastic paraparesis or paraplegia.
Hyperreflexia of lower and (sometimes) upper limbs, increased lower limb tone, clonus and extensor plantar response.
Lower limb muscular weakness (proximal weakness is usually most apparent).
Decreased touch and pinprick sensation, often in poorly defined thoracic areas.
Loss of vibration and position sense - again, more marked in lower limbs. Sensory loss appears to originate in the CNS (probably the spinal cord) rather than the peripheral nerves but it seems likely that peripheral nerves are also involved.[5, 6]

Less common features include:
- Cerebellar signs such as intention tremor.
- Optic atrophy.
- Nystagmus.
- Deafness.
- Cranial nerve lesions.
- Upper limb tremor.
- Absent or reduced ankle reflex.

Differential diagnosis
Includes:
- Lateral sclerosis
- Compressive myelopathy
- Syringomyelia
- Neurosyphilis
- Spinal multiple sclerosis

Investigations
Seek a full history with particular reference to place of birth, countries lived in, and social history. Perform a full neurological examination.
- MRI scan of the spinal cord is needed to exclude other causes of myelopathy. In this condition, MRI may show evidence of demyelination. Cord swelling or atrophy has been noted in a few cases.[1]
- Electrophysiological studies of the lower limb may show abnormalities.
- Lumbar puncture may show a mild lymphocytosis in the CSF of 25% to 60% of patients. Slightly more have mild protein elevation. Most patients have CSF oligoclonal bands.[1]
- A high proviral load in the CSF and peripheral blood may correlate with more severe symptoms.[1]
- Urodynamic studies may be required.

Associated diseases
The HTLV-1 virus is also associated with:
- Adult T-cell leukaemia/lymphoma (ATLL). About 4% of people infected with HTLV-1 develop ATLL.[4]
- Opportunistic infections (including Strongyloides stercoralis hyperinfection).[7]

Management
As with other forms of spastic paresis, patients will require support over a long period of time from many members of the healthcare team. Early introduction to all agencies should be established. This includes physiotherapy, occupational therapy and continence nurses.

There is no one specific drug treatment for the disease.[8] A number of therapeutic options have shown some response in trials:
- Oral methylprednisolone appears to give a good response in some patients.[9]
- Interferon alfa and interferon beta-1a have been reported to have given some good results.[9, 10, 11]
- Antiretroviral agents have also been trialled.[12, 13]
- Pentoxifylline has been used with apparent success but in an uncontrolled trial.[14]

Symptomatic treatment is also important. Spasticity may be treated with drugs such as baclofen. Detrusor instability may be helped by oxybutynin. Tricyclic antidepressants may help with neuropathic pain.

Prognosis
- The disease is a slowly progressive disorder. Although not life threatening in itself, death may occur as a complication of infection or immobility. For example:
  - Septicaemia from urinary infections or infected pressure sores.
  - Pneumonia and pulmonary emboli secondary to immobility.
Survival for 10 to 40 years is not uncommon.\[1\]

**Prevention**

Prevention is based on reducing risk of transmission of the HTLV-1 virus, eg safe sexual practices, screening blood and blood products.

**Further reading & references**

1. Culcea E et al, Tropical Myeloneuropathies, Medscape, Jan 2011

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