

Disease Profile: Transverse Myelitis

(from Synapse magazine, 2008)

Transverse Myelitis is an autoimmune disorder attacking the spinal cord, that presents with a wide variety of symptoms involving sensory, motor and autonomic dysfunction. These symptoms may develop very quickly over a few hours or gradually over a period of several weeks, complicating diagnosis for GPs and Casualty staff. TM is not common – estimated 300 cases p.a. in UK – but it is both debilitating and treatable in the acute phase. Therefore early diagnosis and referral to a specialist neurologist is important. Primary-care and rehabilitation specialists also bear the main role in managing after-care for TM patients, two thirds of whom suffer from long-term sequelae.

Variable Presentation, Diagnosis and Treatment

Some patients encounter TM with rapid onset of weakness and/or bladder dysfunction. This group are more likely to visit Casualty. But other patients start with subacute onset of sensory symptoms and little motor dysfunction. These patients usually visit their GPs, who should note that the symptoms listed below occur in varying combinations and sequence.

Sensory dysfunction: 80-94% of patients will have numbness, paresthesias or bandlike dysesthesias, although these may not all be present initially. A tight, uncomfortable banding sensation around the trunk is a particularly good indicator of TM. Adults are more likely to present with numbness or paresthesias (ie burning, tingling) with a midthoracic sensory level, whereas children show a higher frequency of cervical spine involvement. Other sensory symptoms include heightened or diminished sensitivity to temperature and allodynia – pain caused by non-painful stimuli such as light touch or even wearing clothes.

Autonomic dysfunction and weakness: Acute urinary retention is typical at the outset, and most TM patients suffer bladder and bowel dysfunction due to loss of sensation, which is often complete in the acute phase. Most patients develop leg weakness. At the maximal level of deficit, 50% of patients have lost all leg movement. Onset of paralysis tends to be rapidly progressive; complete paralysis can occur within hours.

Timely MRI imaging, CSF analysis and lab tests are used for diagnosis, as well as history and physical examination. TM must be distinguished from compressive lesions first of all, and then from other conditions such as Guillain-Barre Syndrome and MS. Rheumatological symptoms must be looked for, as TM may also be a presentation of systemic autoimmune disease eg lupus, sarcoidosis etc.

First-line treatment is high-dose IV corticosteroids for 3-5 days to reduce inflammation, followed by oral steroids taper. Plasma exchange should be considered for severe TM that is refractory to corticosteroids, or for patients with suspected antibody-mediated disease eg NMO. When TM is believed to

be recurrent (NMO-IgG blood test), immunosuppressant drugs should be considered.

Prognosis and Long-Term Management

Most people with TM experience some degree of neurologic recovery but are also left with neurologic deficits. Though recovery is more rapid in the first 6 months after onset, patients can experience some improvement for up to 2 years and physiotherapy should not be terminated prematurely.

Approximately one third recover with little or no sequelae, one third are left moderately disabled, and one third are left severely disabled. Bad prognostic indicators include back pain at onset, rapid progression to maximal symptoms within hours, spinal shock, and sensory involvement up to the cervical level. Lack of any improvement in the first 3-6 months makes significant recovery less likely.

TM is not always monophasic as previously believed; recurrence affects a small percentage of patients. In all cases of recurrence, the potential for an underlying disorder should be investigated. Patients with lesions over 3 vertebral segments may go on to be diagnosed with NMO. In some patients TM may be first manifestation of MS – brain MRI and oligoclonal CSF bands are usually indicative of MS. Treatments for MS, NMO and TM are different, emphasising the need for good diagnosis.

Once the acute phase of TM subsides, most patients are left with sequelae that greatly affect their lives and whose course depends on early physical and occupational therapy:

- Physiotherapy is essential not only to improve strength, mobility and gait but also to manage spasticity and improve sensation.
- Chronic neuropathic pain responds poorly to narcotics. Persistence and attention to side effects by primary-care doctors are required to identify the right remedy(s) in each individual case.
- Anti-spasticity drugs should not be limited to baclofen, and should be used in combination with active stretching exercise to maintain flexibility.
- Most patients continue to have some degree of bladder and bowel dysfunction. All patients should see a urologist for long-term surveillance. Urodynamic studies should be used to diagnose type(s) of bladder dysfunction. Sexual dysfunction often parallels bladder dysfunction.
- 25% of patients with TM become clinically depressed, *irrespective of their level of disability*. Patients should be routinely screened for signs and symptoms of clinical depression. Fortunately, patients with TM seem to respond favourably to medications and counselling.

Long-term TM sequelae are often subtle but very disabling. Neurologists do not monitor TM patients closely unless MS is suspected, and it is up to GPs and rehabilitation specialists to work together to manage chronic pain, continence, spasticity, fatigue and depression.