

A case of post polio syndrome

HISTORY

A 43 year old lady was seen in clinic with progressive leg weakness. At the age of 9 weeks she had an illness causing weakness of both legs which kept her in hospital until the age of 6. A diagnosis of poliomyelitis was made at this stage and she had several foot and ankle operations. She had used callipers as a child and young adult, but had walked well and excelled at swimming. At the age of 36, she complained of right arm dysfunction with pain. Subsequent to this she had increasing weakness of her right arm and leg with some falls. This progressed slowly and severely limited her mobility to a few yards with crutches. Over the course of a few years she had progressive left leg weakness and the need for a wheelchair. More recently she developed some left sided hand parasthesiae in the mornings.

EXAMINATION

Cranial nerve examination was normal. She had no upper limb wasting. There was some weakness of the right triceps. Right triceps and supinator reflexes were absent, but the remainder were preserved. In the lower limbs, there was global weakness of the legs with bilateral wasting (more pronounced on the right and more distal than proximal). She was areflexic in the lower limbs with flexor plantars. There were no abnormal sensory findings and proprioception was normal.

INVESTIGATIONS

MRI brain was normal and an MRI of whole spine showed moderate thoracic scoliosis with minimal degenerative disease, but no focal compressive features or cord signal abnormality. Nerve conduction studies revealed a delayed left median nerve sensory conduction time, in keeping with a left carpal tunnel syndrome. Limited EMG studies showed evidence for chronic partial denervation in keeping with previous polio.

DIAGNOSIS

A diagnosis of post-polio syndrome was made.

DISCUSSION

Post-polio syndrome (PPS; also known as progressive postpoliomyelitis muscular atrophy) is a condition characterised by progressive weakness many years after recovery from acute poliomyelitis. Patients will have generally experienced neurological stability following the initial disease for at least 10 years, followed by progressive weakness in previously affected muscles. Sometimes previously unaffected muscles may also become weaker, although EMG examination in those muscles usually shows clear evidence of previous disease, suggesting that PPS is likely to be confined to those muscles initially affected by the acute disease.

The aetiology of PPS is unknown. Some patients have oligoclonal bands in the CSF and occasional lymphocytic infiltrates in muscles and spinal cord, raising the possibility of a persistent immune-mediated mechanism. There has been no definite documentation of persistent polio virus infection in patients with PPS. A further potential mechanism for the disease is the 'peripheral disintegration model'. This suggests that following acute motor neuron degeneration in the acute stages of poliomyelitis, there is oversprouting of new axon terminals which stabilises muscle strength. After many years, however, the increased metabolic demand on these resprouted motor neurons may lead to excessive metabolic burden and subsequent degeneration.

References

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- Farbu E, *Post Polio Syndrome: Diagnosis and Management*. ACNR 5(1): 10-11.



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