Short report

Tabes dorsalis: electrodiagnostic features

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SUMMARY Electrodiagnostic data have not been previously reported in tabes dorsalis. A patient with tabes dorsalis is described whose nerve conduction studies and median nerve somatosensory evoked responses (SEPs) were normal. H-reflexes were absent. SEPs of the tibial nerve suggested posterior column dysfunction. These electrodiagnostic findings correlate precisely with the known pathology of tabes dorsalis.

Although tabes dorsalis is the most common neurological expression of tertiary syphilis,¹ no reports exist of its clinical electrophysiology. Since the symptoms and signs of tabes dorsalis mimic those of severe peripheral neuropathy or myelopathy, patients with tabes dorsalis may be referred for electrodiagnosis. We describe the electrodiagnostic findings in this disorder.

Case presentation

A 54 year old woman was referred for poor balance, leg weakness and pain, recurrent left knee effusions, and a previous history of "polio". She developed weakness of both legs at age 7 years, diagnosed as "polio". Several years later, she began to experience diminished lower extremity sensation and gait deterioration.

Several years after onset of the disease, bladder distension and overflow incontinence further complicated her illness. Additional disabilities included lower extremity lightning pains, recurrent knee effusions, and chronic toe infections necessitating amputation.

Examination revealed an enlarged left knee, pes cavus and hammer toe deformities, and amputation of both small toes. Pupils were large and unreactive to light and accommodation. Dilute pilocarpine did not constrict them. Reflexes were normal in upper extremities and absent in the legs. Light touch, pinprick, and cold sensation were markedly reduced in the legs from 5 cm above the patella distally. Vibration sensation was absent distal to the pelvis. Proprioception was absent at the great toe and ankle, and markedly

reduced at the knee. Coordination was severely impaired in her legs. Her gait was wide-based and unsteady; she was unable to tandem walk. Romberg manoeuvre demonstrated profound instability.

Routine blood and urine studies were normal. Serum folate level was normal as was Schilling's Test, Part I. Serum VDRL was non-reactive (VDRL was reactive in a previous hospitalisation). Serum microhaemagglutination-*Treponema pallidum* (MHA-TP) was positive. Cerebrospinal fluid (CSF) analysis revealed no cells, a protein of 0.24 g/l (normal <0.45), and a non-reactive VDRL. Thoracic and lumbosacral spine myelography, CT, and MRI were normal.

Sural, median, and ulnar sensory and peroneal and posterior tibial motor nerve conduction studies were normal. Actual values (amplitude, distal latency, conduction velocity, respectively; normal values in parentheses) were: sural 15 μ V (≥ 6), 3.4 ms (≤ 4.2); median sensory 45 μ V (≥ 22), 2.5 ms (≤ 3.4), 62 m/s (≥ 53); ulnar sensory 30 μ V (≥ 10), 2.5 ms (≤ 3.2); peroneal motor 5 mV (≥ 2), 4.5 ms (≤ 6.1), 51 m/s (≥ 41); and posterior tibial 4 mV (≥ 3), 3.9 ms (≤ 6.2), 47 m/s (≥ 41). F-response latencies were normal: peroneal 45-6 ms (≤ 56) and posterior tibial 47-6 ms (≤ 58), but posterior tibial nerve H-reflexes were absent.

Median nerve somatosensory evoked potentials (SEPs) were normal, yet posterior tibial nerve SEPs demonstrated posterior column dysfunction (fig). EMG was unremarkable except for minimal spontaneous activity in both medial gastrocnemius muscles. Needle examination of lumbosacral paraspinal muscles was normal.

The patient's pupillary, neurological, urological, orthopaedic, and serological manifestations confirmed the diagnosis of tabes dorsalis. She denied previous syphilis injection, but subsequent questioning of relatives uncovered a previously-undisclosed history of syphilis in both maternal grandparents and her mother, in the latter case during the patient's childbirth. Because of the uncertainty of previous treatment, the patient received a full course of intravenous and intramuscular penicillin.

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Fig Somatosensory evoked potentials: Well-defined responses (two trials each) were recorded from Erb's point, the spine, and the scalp on left median nerve stimulation. Left tibial nerve stimulation produced recordable responses at the popliteal fossa, L_3 and T_{12} , but none over the scalp.

Discussion

This patient manifested the classic triads of symptoms and signs of tabes dorsalis described by Merritt, that is, symptoms (lightning pains, ataxia, and dysuria) and signs (tabetic pupils, areflexia, and proprioceptive loss).² Since her clinical and electrophysiological presentation was incompatible with previous polio myelitis, we hypothesise that she acquired syphilis congenitally and experienced her first symptoms of tertiary disease at age 7 years. Infants infected at birth

may be asymptomatic and may not manifest signs commonly associated with congenital syphilis. Even though most features of tabes dorsalis do not develop until 10–25 years after primary infection, this latency may be as short as 5 years in children.³

Serological findings are variable in tabes dorsalis depending on the clinical activity and duration of the disease.¹ Since MHA-TP detects antibodies specific for *Treponema pallidum*, reactivity persists regardless of disease duration, severity, or previous treatment. Conversely, VDRL is non-reactive in 25 to 57% of patients with late syphilis.^{1,4–6} CSF analysis may be similarly insensitive in tabes dorsalis. A nonreactive VDRL and acellular count is not unusual particularly in “burnt out cases”.¹ In Merritt's series of 100 patients with tabes dorsalis, CSF serology was non-reactive in 28 patients and a normal WBC count was observed in 53 patients.³

In addition to the classic symptom and sign triads, our patient displayed other features of tabes dorsalis. These included severe nociception loss in the legs resulting in painless trauma, recurrent toe infections, and amputation of several toes. Recurrent left knee effusions probably represented an early Charcot joint, although radiographs did not confirm destructive changes. Distal weakness and atrophy may be late manifestations of tabes dorsalis, attributed to extension of the syphilitic process to anterior horn cells or motor roots.³

Clinical electrophysiology has not been previously reported in tabes dorsalis. Dyck recorded normal amplitude and conduction velocities of A alpha, A delta, and C fibres in vitro from the sural nerve of a tabetic patient.⁷

A neurophysiological-pathological correlation emerges from the electrodiagnostic results in this case and the known pathology of tabes dorsalis. Abnormality in tabes dorsalis is concentrated in the dorsal roots, dorsal funiculi, and posterior columns of the lumbosacral and lower thoracic spinal cord.^{3,8} Usually spared are the anterior horn cells and ventral roots.⁸ The dorsal root ganglia are rarely affected to a significant degree and individual ganglion cells do not show features of degeneration.^{9,10} Stern identified inflammation in the dorsal root ganglia from only one of nine patients with tabes dorsalis.⁹

Normal sensory conduction studies verify integrity of the sensory nerves and dorsal root ganglia. Absent H-reflexes can be attributed to atrophy of the dorsal roots in the sacral area, preventing entry of the sensory arc of the H-reflex. Absent cortical SEPs on tibial nerve stimulation are explained by posterior column degeneration. Normal median nerve SEPs confirm integrity of the sensory pathway in the cervical area through the dorsal columns to the somatosensory cortex. Motor conduction studies are normal since

motor fibres are rarely affected in tabes dorsalis. Fibrillation potentials in the gastrocnemius muscles could be explained by superimposed S₁ radiculopathies, but absence of S₁ root abnormality in proximal and paraspinal muscles argues against this explanation. A more plausible interpretation would be partial anterior horn cell involvement known to occur in some cases of tabes dorsalis.³

Pupillary abnormalities excluded, tabes dorsalis mimics peripheral neuropathy, spinal cord disease, and lumbo-sacral polyradiculopathy. Tabes dorsalis should be considered in any patient manifesting the electrodiagnostic triad of normal nerve conduction studies, absent H-reflexes, and impaired posterior column conduction.

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