Sacral Nerve Dysfunction Plus Generalized Polyneuropathy in Herpes Simplex Genitalis

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Sacral nerve dysfunction consisting of urinary retention, anal sphincter paralysis, sacral paresthesias, and weakness and reflex changes in the lower extremities may complicate genital herpes simplex virus (HSV) infections [1, 2, 6]. Less often, generalized polyneuropathy has been linked to HSV [4, 7]. We describe a patient who developed both these disorders accompanying an attack of HSV vaginalis.

A 23-year-old woman noted numbness in the feet that over the next two weeks progressed to include mild lower leg weakness, accompanied by difficulty in urinating, constipation, and a burning sensation around the vulva. The entire perineal region became numb. Concurrently she experienced paresthesias in the mouth and developed facial weakness. Her sexual partner was said to have recurrent genital "herpes."

In hospital we observed a few ulcerated lesions on the vulva of the patient. She had bilateral facial weakness and mild symmetrical distal limb weakness with areflexia. There was mild distal reduction in responses to pinprick, temperature change, and light touch, plus anesthesia in the S3–S5 dermatomes. The anal sphincter was patulous.

The following investigations showed no abnormalities: electroencephalogram, cranial computed tomographic scan, visual evoked responses, SMA-12 autoanalysis, complete blood count, serum complement, protein electrophoresis and immunoglobulin quantification, mononucleosis spot test, throat swab for diphtheria, antistreptolysin-O titer, hepatitis-B antigen, urinalysis, and urinary porphobilinogen. Cerebrospinal fluid examination showed 60 lymphocytes per cubic millimeter, a protein level of 94 mg/dl, an IgG level of 10.1 mg/dl, and a glucose level of 53 mg/dl; a viral antibodies test was negative. Skin scrapings from the vulvar lesions grew HSV.

Nerve conduction studies showed marked slowing of distal and proximal motor conduction velocities, with prolongation of the distal F-wave latencies. Sensory action potentials were also markedly reduced in amplitude but had normal velocities. Blink responses showed delayed early and late ipsilateral and late contralateral responses. Electromyography of limb and facial muscles showed a reduction in the recruitment but no denervation.

By eight weeks after the onset of symptoms, the patient had regained strength and bladder and bowel control; four months later she had no abnormal neurological symptoms or signs.

This patient's flaccid weakness, mild distal sensory changes, areflexia, facial diplegia, and nerve conduction abnormalities were characteristic of acute polyneuropathy. The prominent urinary and anal sphincter disturbance, severe sacral hypesthesia, and cerebrospinal fluid pleocytosis are atypical of an acute polyneuropathy and more attributable to a localized sacral radiculopathy. The relatively rapid clinical recovery from both syndromes, as well as the results of the initial nerve conduction studies, suggests that the nerve damage was segmental demyelination rather than axonal destruction.

The pathogenetic mechanisms in localized sacral nerve dysfunction in HSV infections are unknown but are presumed to be direct invasion by these neurotropic viruses. Those in postinfectious polyneuropathy are similarly unknown but believed to be immune mediated. In humans, acute polyneuropathy has been linked only rarely to HSV infections, although in fowl an inflammatory demyelinating peripheral neuropathy can be caused by a herpesvirus (Marek's disease) [8].

A variety of neurological syndromes, including sacral and more diffuse [5] radiculopathies, myelopathy [3], and polyneuropathy, have been reported in association with herpes simplex genitalis, but to our knowledge this is the first report of a patient in whom sacral radiculopathy and generalized polyneuropathy occurred simultaneously.

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Late-Onset Pyridoxine-Dependency Convulsions

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A patient with late-onset pyridoxine-dependency seizures was reported in a recent issue [1]. I have followed a boy with a similar disorder.

A 6-year-old boy had single seizures unassociated with fever at 4 months, 5 months, and 6 months of age. On each occasion, the parents had given the boy whole milk, and the seizure reportedly occurred after he had drunk a single bottle of whole milk. After they had reverted to formula, the seizures ceased until the patient was brought to the hospital at 8 months of age in status epilepticus (on this occasion, he had not been given whole milk). The status epilepticus was unresponsive to large doses of phenytoin, phenobarbital, and paraldehyde. He was given 50 mg of intravenous pyridoxine, and the seizures stopped within minutes. He remained seizure-free for the next six days, when seizure activity recurred and again promptly stopped with intravenous administration of pyridoxine.

Results of extensive laboratory testing were normal. Over the ensuing six months the dosages of anticonvulsants were tapered and discontinued. The patient remained seizure-free on pyridoxine, 25 mg a day taken orally.

When the patient was 2½ years of age, pyridoxine was tapered and discontinued. Within a few days of discontinuing the pyridoxine, he began having occasional myoclonic jerks, particularly during sleep. Two days later, he again developed status epilepticus, which was promptly stopped by intravenous pyridoxine.

At age 6, he remains seizure-free on a daily dose of 25 mg of pyridoxine. His intellectual development is entirely normal. Interictal electroencephalograms have been normal. Ictal electroencephalograms were abnormal, with nonfocal seizure discharges.

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Long Latency Between the Onset of Motor and Vocal Tics in Tourette's Syndrome

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Most authorities regard the occurrence of involuntary vocal utterances as necessary for the diagnosis of Tourette's syndrome. Most patients develop vocal tics during childhood or shortly after the onset of the disorder, usually within one to three years [6].

A 74-year-old woman had a history of simple motor tics involving her face, neck, and shoulders dating back to before the age of 20. The tics were of variable intensity, increasing with emotion and decreasing with relaxation. She had noted no spontaneous waxing and waning of symptoms and no notable variability in distribution. She had not experienced any vocal or complex motor tics. The tics had interfered with daily living only minimally, until at the age of 72 she began shouting the names of friends and her husband involuntarily several times an hour, producing considerable marital discord. The frequency of the shouting fluctuated with changes in emotional state but rarely occurred during conversations. She said that she had first developed rare, involuntary coprolalia and echolalia at age 74, but these tics were not witnessed during her hospitalization. Over the year prior to admission she had suffered from a depressive disorder treated with amitriptyline, which had no influence on the tics and was discontinued one month before admission. Her history was otherwise unremarkable, except for hypertension controlled with propranolol. There was no history suggestive of a tic disorder in blood relatives.

Examination revealed an elderly, mildly depressed woman with frequent tics involving the left sternomastoid and pectoralis muscles. There was less frequent shoulder abduction, lip pursing, and grimacing of the left side of the mouth. She had no vocalizations during the initial interview, but subsequently she would frequently shout out the names of friends, her husband, and later the ward staff. The remainder of the neurological examination was normal. Pimozide [5], 3 mg daily, controlled both her vocal utterances and her motor tics.

This long delay between the onset of simple motor tics and complex vocalizations is most unusual. Shapiro and coworkers state that vocal tics usually appear within the first three years of the onset of the disorder, but coprolalia may be delayed up to 24 years [6]. Until the age of 72 this patient would have been classified as having a chronic simple motor tic disorder [1]. This late occurrence of vocalizations is evidence for the need to consider tic syndromes on a continuum [3] rather than as a series of separate entities.

Ziegler reported another woman in her seventies with Tourette's syndrome and essential tremor and emphasized the rare documentation of this lifelong disorder in the elderly [7]. Interestingly, the first well-described patient, reported by Itard in 1825 [4] and included in Tourette's original paper [2], was 85 years of age; this remains the oldest documented case to date. These and all other elderly Tourette patients reported have had the usual short latency between the onset of motor and vocal tics.

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