

## Brief Report

# Somatosensory Phenomena in Huntington's Disease

Roger L. Albin and Anne B. Young

*Department of Neurology, University of Michigan, Ann Arbor, Michigan, U.S.A.*

---

**Summary:** Sensory symptoms are generally not associated with Huntington's disease (HD). We describe two patients with HD who had painful somatosensory symptoms. One patient also had auditory hallucinations. No other cause was found for these symptoms. Both patients also had significant depression and one patient committed suicide. Somatosensory symptoms may be a marker for depression in HD. **Key Words:** Huntington's disease—Somatosensory symptoms—Depression.

---

Huntington's disease (HD) is characterized by autosomal dominant inheritance of neuropsychiatric dysfunction, chorea, and other movement disorders. Sensory phenomena have not been associated with HD, although pathologic abnormalities have been documented in the thalamus and posterior columns of HD cases (1-3). Somatosensory evoked potentials (SSEPs) are also abnormal in HD (4). In Bruyn and Went's recent review of HD, sensory phenomena are not mentioned (5); Hayden, in his monograph, states that "there have been no reports of obvious sensory defects in Huntington's chorea" (6). Folstein et al. have reported two patients with auditory hallucinations and interpreted these aural symptoms as the manifestation of an associated psychiatric disturbance (7). We have encountered two HD patients with significant somatosensory symptoms and no other evident explanation for the occurrence of these symptoms. One of these patients also had auditory hallucinations. These two patients represent a small fraction of the approximately 150 HD patients seen in our movement disorders clinic over the past 8 years.

## CASE REPORTS

### Case 1

This 42-year-old right-handed man whose mother, brother, sister, half-sister, and half-brother have documented HD first noted involuntary movements in mid 1983. Over the next 3 years his chorea progressed. His coordination and gait gradually deteriorated and by

---

Address correspondence and reprint requests to Dr. R. L. Albin at Neuroscience Building, 1103 E. Huron Street, Ann Arbor, MI 48104, U.S.A.

mid 1986 he was no longer able to work as a janitor. He and family members denied cognitive changes.

In mid 1986 he began to notice an unusual sensory disturbance. He described this as an intermittent "bee-sting feeling." This sensation could occur on any part of his body, lasted for seconds to minutes, and was burning in quality. The discomfort was usually felt over an area ~1 cm in diameter. Initially infrequent and mild in intensity, the discomfort worsened over the subsequent months. By early 1987 the pain had become intense and was occurring many times per hour. Trials of diphenhydramine, haloperidol, and amitriptyline failed to relieve the pain.

He was initially seen at the University of Michigan Movement Disorders Clinic in February of 1987. Examination at that time revealed normal mental status except for a depressed affect. He admitted to occasional suicidal ideation. Examination of cranial nerves revealed slowed optokinetic nystagmus and saccades, intermittent blepharospasm, oral-buccal-lingual chorea, and mild dysarthria. He had normal power and bulk in all muscles tested, rapid rhythmic alternating movements were slowed, and there was moderate chorea and dystonia involving all extremities, trunk, and neck. Sensation was intact to pain and light touch with questionable impairment of position and vibration sense. Deep tendon reflexes were 3+ and symmetric. Both plantars were flexor.

His sensory symptoms continued to worsen and in March of 1987 he began to complain that the pain was so intense that he wished to end it by committing suicide. He was admitted to the inpatient psychiatric facility at the University of Michigan Medical Center for observation. He had a significant affective disturbance. Serum chemistries, complete blood count with differential, platelet count, serum folate and  $B_{12}$  levels, and thyroid function test results were normal. Nerve conduction studies, needle electromyography, and median nerve SSEPs were normal. Therapy was begun with carbamazepine, and on a dose of 200 mg p.o. t.i.d. (serum level 6.69; therapeutic range 4–8) he reported improvement in the intensity of his sensory symptoms, his affective disorder improved, and he was discharged. Several months later he continues to complain of his "bee-stings" but the intensity appears to be tolerable and his affective disorder has largely resolved.

## Case 2

This 42-year-old female elementary school teacher whose paternal grandfather, father, paternal uncle, and sister had HD first developed sensory symptoms in the spring of 1973. At that time she developed an upper respiratory infection with coryza, hyperacusis, vertigo, malaise, arthralgias, and myalgias. The coryza resolved but the other complaints persisted. She also began to experience uncomfortable paresthesias in her extremities, and intermittent, sharp, shooting pains in her legs. Coincident with this illness she began to notice infrequent, low amplitude, "jerky," involuntary movements of her extremities, and mild impairment of her ability to carry out fine coordinated movements.

She was first seen in the University of Michigan Neurology Clinic in August of 1974. Examination at that time was normal. Trials of diazepam, indomethacin, and doxepin failed to produce improvement of her symptoms. Her sensory complaints spontaneously receded over the next two years. In 1976 her symptoms returned. She again complained of myalgias, arthralgias, malaise, dysequilibrium, pain in her extremities, and hyperacusis. She was seen by several physicians and a diagnosis of polymyositis was made. A several-month course of prednisone did not relieve her symptoms. Her involuntary movements had gradually increased in frequency and amplitude from 1973 to 1976. She was treated with codeine, propoxyphene, perphenazine/amitriptyline, diazepam, and thioridazine without any change in symptoms.

She was seen in the Movement Disorders Clinic at the University of Michigan in February of 1980. She stated that her symptoms had progressively worsened over the past 4 years. She also complained of difficulty with walking and some problems with her memory.

Mental status testing revealed deficits of memory, calculations, and orientation. Examination of her cranial nerves showed slowed optokinetic nystagmus, and choreoathetoid movements of the tongue. Strength, tone, and muscle bulk were normal but there were mild choreoathetoid movements of all extremities and the trunk. Gait was narrow based with short, stuttering steps. Deep tendon reflexes were 3+ and plantars were flexor. Sensory examination revealed normal pain, temperature, light touch, and proprioception. Vibratory sense was minimally decreased in the lower extremities. Serial trials of phenytoin 300 mg/day, haloperidol 5 mg b.i.d., and thorazine 100 mg b.i.d. failed to relieve her symptoms. Her auditory symptoms worsened and she began to complain of aural hallucinations. These would often awaken her from sleep and consisted of choirs singing the last thought she had before falling asleep. She also described hearing hymns and an unidentifiable voice repeating her thoughts back to her. She continued to complain of severe, lancinating pain involving her extremities. She was admitted to the inpatient Neurology Service at the University of Michigan Medical Center in July of 1980 for evaluation of her auditory and somatosensory symptoms. A psychiatric consultant believed that the patient had an affective disorder. She was treated with amitriptyline, methadone, perphenazine, and ethchlorvynol. Her auditory and somatosensory symptoms continued. Over the following months she continued to complain of considerable difficulty with the auditory hallucinations and of severe lower extremity pain. She committed suicide in July of 1982.

Postmortem examination of her brain revealed no gross atrophy, but mild gliosis of the caudate nucleus was present on microscopic examination. No other gross or microscopic abnormalities were documented.

## DISCUSSION

We have described two HD victims with prominent somatosensory symptoms. One of these patients also had auditory hallucinations. Both had significant affective disturbances and one committed suicide. One patient had sensory symptoms coinciding with the onset of chorea. In the other, the sensory complaints began months after the onset of chorea. In the absence of any other identifiable cause it seems likely that these sensory symptoms were secondary to HD. Thalamic and posterior column abnormalities have been noted in HD (1-3) although their prevalence within the HD population is unknown. However, the documented thalamic abnormalities are restricted to the ventrolateral thalamus and seem not to involve the posterior portions of the thalamus involved in the relay of somatosensory input (3). SSEPs are known also to be abnormal in HD and the degree of abnormality seems to correlate with clinical disease status (4). However, in our two patients, SSEPs were normal in the one patient in whom they were examined, and no thalamic changes were noted in the brain of the patient who came to autopsy. The latter finding does not exclude thalamic pathology because such changes may only be detectable by quantitative morphometry (1).

One feature common to both of these cases is that both patients had significant affective disorders. Folstein et al. have shown that affective disorders are quite common in HD (8,9). Affective symptoms may be an intrinsic feature of HD and not a reaction to chronic illness. Somatic symptoms may be a manifestation of depression and treatment of depression may relieve somatic symptoms (10). One patient had a positive response to carbamazepine. This agent is commonly employed in the treatment of neurogenic pain but may also have some efficacy in the treatment of affective disorders (11). The sensory symptoms experienced by these two patients may be either a manifestation of depression or an associated symptom secondary to another process causing both sensory symptoms and affective disorder. Regardless, we recommend that HD patients with significant sensory complaints be carefully monitored for the development of depression.

**Acknowledgment:** The authors thank Suyin Liang for efficient word processing.

## REFERENCES

1. Dom R, Malfroid M, Baro F. Neuropathology of Huntington's chorea. *Neurology* 1976;26:64-8.
2. Forno LS, Jose C. Huntington's chorea, a pathological study. *Adv Neurol* 1973;1:453-70.
3. Roos RAC. Neuropathology of Huntington's chorea. In: Vinken PJ, Bruyn GW, Klawans HJ, eds. *Handbook of clinical neurology; Vol. 49; Extrapiramidal disorders*. New York: Elsevier, 1986:315-26.
4. Ehle AL, Stewart RM, Lellelid NA, Leventhal A. Evoked potentials in Huntington's disease: a comparative and longitudinal study. *Arch Neurol* 1984;41:379-82.
5. Bruyn GW, Went LW. Huntington's chorea. In: *Handbook of clinical neurology; vol 49; Extrapiramidal disorders*. New York: Elsevier, 1986:267-313.
6. Hayden MR. *Huntington's chorea*. New York: Springer-Verlag, 1981:72.
7. Folstein SE, Folstein MF, McHugh PR. Psychiatric syndromes in Huntington's disease. *Adv Neurol* 1979;23:281-90.
8. Folstein SE, Abbott MH, Chase GA, Jensen BA, Folstein MF. The association of affective disorder in Huntington's disease in a case series and in families. *Psychol Med* 1983;13:537-42.
9. Folstein SE, Folstein MF. Psychiatric features of Huntington's disease: recent approaches and findings. *Psychiatr Dev* 1983;2:193-206.
10. Sternbach RA. Pain and depression. In: Kiev A, ed. *Somatic manifestations of depressive disorders*. New York: Elsevier, 1974:107-19.
11. Post RM, Ballenger JC, Uhde TW, Bunney WE. Efficacy of carbamazepine in manic-depressive illness: Implications for underlying mechanisms. In: Post RM, Ballenger JC, eds. *Neurobiology of mood disorders*. Baltimore: Williams and Wilkins, 1984:777-816.