Motor Abnormalities in CRPS: A Neglected but Key Component

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HISTORICAL PERSPECTIVE

A review of the medical and scientific literature reveals that little attention has been paid to the motor abnormalities seen in patients with complex regional pain syndrome type 1 (CRPS-I), previously known as reflex sympathetic dystrophy (RSD). However, most authorities have remarked, albeit briefly, that motor dysfunction is indeed a common phenomena in patients with this condition. For instance, Bonica’s 1953 classic staging of RSD describes Stage 1’s symptomatology as “limitation of movement” and Stage 3 as “extreme weakness and limitation of motion” (Bonica 1990). Hannington-Kiff (1991) wrote: “reflex sympathetic dystrophy is as much a movement disorder as a painful condition.” In fact, several earlier diagnostic criteria did include motor symptoms as key components to diagnosis (Bonica 1990; Blumberg et al. 1994). At the 6th World Congress on Pain, authorities adopted an operational definition of RSD that included “abnormalities in the motor system” (Jänig et al. 1991). Yet motor disturbances are often ignored in the clinical evaluation and treatment of CRPS patients.

The current IASP guidelines do not include motor dysfunction as one of the diagnostic criteria for CRPS and only state that “impairment of motor function can include weakness, tremor, and in rare instances, dystonia” as “associated symptoms and signs” (Merskey and Bogduk 1994). Interestingly, the expert consensus groups that met at a 1993 workshop in Orlando to define these criteria reportedly had a heated discussion about including...
motor abnormalities in the criteria. Remarking on this decision, Boas (1996) later wrote that “weakness, tremor, and reduced movement are often present in CRPS, to the extent that some members of the workshop felt it should be an inclusion criteria in definition of the disorder … these changes can be as limiting as the pain.” Because this group felt that motor disturbance was not as well understood or documented as the “dysautonomic signs” and that it was a secondary symptom or sign under the patient’s volitional control, the current IASP diagnostic criteria exclude motor abnormality.

CLASSICAL TEACHING VERSUS NEW THEORIES

Classical teaching about the immobile “RSD limb” infers that the patient volitionally guards and protects the painful limb to minimize mechanical and proprioceptive stimulation so as to reduce the exacerbation of pain (allodynia). While some patients may indeed respond in this manner, recent interesting and novel data from CRPS patients strongly suggest that motor abnormalities (1) are one of the more common symptoms and signs observed in CRPS; (2) may be a distinctive and unique clinical feature of CRPS, or at least a subset of CRPS, that may allow for a more precise diagnosis of this condition; and (3) may be due to neuroplastic alterations in the central nervous system (CNS), specifically in the brain.

INTEGRATION OF THE MOTOR SYSTEM 
AND PAIN: THEORY AND NEUROANATOMY

Pain clinicians and researchers alike sometimes fail to recognize that a behavioral motor response is an integral component of all pain experiences. Pain causes us to immediately cease our behavior, pay attention to an important (and potentially tissue-damaging or even life-threatening) stimulus, and then react to the stimulus. Interestingly, recent scientific writings in the neurological “movement disorder” literature have hypothesized that in certain movement disorders, such as dystonia, the abnormality may not lie in the motor system but rather in the sensory system, and may constitute a disconnection between sensory input and motor output within the CNS (Dauer et al. 1998; Bara-Jimenez et al. 2000). Thus, it should be no surprise that a pain condition that results in chronic and persistent abnormal sensory input into the CNS can have associated symptoms and signs of motor abnormalities (Galer et al. 1995).

Functional neuroimaging studies of the human (and animal) pain experience are producing evidence that the networks for pain, attention, autonomic
regulation, and motor function are interrelated and share many brain structures (Galer et al. 1995) (Table I). Teleologically, survival of the organism will depend on the seamless cross-networking integration of these four interrelated and integral brain systems. An animal in pain must possess two automatic reactions, i.e., two innate “software packages.” One directs attention to the body region in pain so as to immediately decrease and then later increase blood flow to the region to aid healing and to guard and protect the region from further harm. The second reaction, occurring in the midst of battle, ignores the pain, and pays attention to the threat, and increases blood flow to all muscles to increase strength and quickness for escaping or fighting (“fight or flight”).

If these systems are interrelated, it follows that dysfunction in one brain network most likely will have direct effects on others, especially if the dysfunction persists. As we now know, the nervous system is plastic and is continuously adapting to both internal and external environmental changes. Thus, continuous increased input from peripheral nociceptors will result in neuroplastic changes at higher nervous system levels, not only within the well-described brain pain network, but also within other related brain networks, such as attention, autonomic, and motor regions, as hypothesized earlier (Galer et al. 1995). Thus, we can theorize about the underlying pathophysiological process of CRPS-associated motor abnormalities.

**EPIDEMIOLOGY**

Only recently have prospective studies evaluated how frequently CRPS (or RSD) patients complain of motor abnormalities and how often motor signs are present on examination. In the largest prospective study of signs and symptoms in 829 RSD patients, Veldman and colleagues (1993) reported that 95% of patients had “weakness” of the involved limb, 54% “muscular

<p>| Table I |
| Brain regions involved in pain, autonomic regulation, attention, and motor functions |</p>
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<tr>
<th>Brain Region</th>
<th>Pain</th>
<th>Autonomic</th>
<th>Attention</th>
<th>Motor</th>
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<tr>
<td>Anterior cingulate cortex</td>
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<td>Periaqueductal gray</td>
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<td>Posterior parietal cortex</td>
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<td>Hypothalamus</td>
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<td>Reticular system</td>
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incoordination,” and 49% “tremor.” Interestingly, this report also noted a subset of 15% of patients with “no active movement” of the RSD limb, where the investigators diagnosed a neurological disorder described as “pseudoparalysis” because electrophysiological testing was normal. A smaller study of 41 CRPS patients evaluated and cared for in a university tertiary pain center asked them to recall their initial symptoms (Rashiq and Galer 1999). Of note, 65% of these patients reported that they had significant motor abnormalities as one of their initial symptoms. An early descriptive study by Schwartzman and Kerrigan (1990) observed that “focal dystonias, weakness, spasms, tremors, difficulty initiating movement, and increased tone” are common features of RSD. Thus, evidence is clear that a significant proportion of CRPS patients suffer from an associated motor disturbance involving the painful limb.

NEGLECT-LIKE ABNORMALITIES

Knowledge of basic neurobehavioral concepts reveals that motor symptoms and signs in some CRPS patients are not attributable to the classical teaching of voluntary guarding and protection of the painful limb. Histories taken from these patients suggest that they are suffering from a subtle form of “neglect,” similar to that seen in parietal stroke patients. These patients stated that their inability to use their limb was “not just because of the pain” but rather because they “needed to focus all mental attention and look at the limb in order for it to move the way I want.” At times the patients admitted that “the limb feels disconnected from my body, like it doesn’t belong to me.” Unfortunately, many of these patients were (and continue to be) reticent to discuss these strange perceptions, fearing further labeling of “psychogenic” pain (Galer et al. 1995). These observations led to further study to document this phenomenon. However, in their earlier descriptive study, Schwartzman and Kerrigan briefly mentioned problems with “initiating movement” in RSD patients, but they hypothesized a spinal cord abnormality resulting in a “movement disorder” and not a neglect-like disorder, which would most likely arise from brain dysfunction (Schwartzmann and Kerrigan 1990).

Motor neglect is comprised of akinesia, bradykinesia, deficits in movement amplitude, and reduced frequency of movement. Cognitive neglect is present when the patient disowns a body part. The patient feels that the involved limb is not part of the body or is not connected to the body. A survey study of 242 CRPS patients found that 84% admitted to at least one neglect symptom and 47% reported at least one motor and one cognitive neglect symptom (Galer and Jensen 1999).
MOTOR ABNORMALITIES IN CRPS

Motor neglect is easily ascertained. The clinician must simply document on physical examination that the movement of the involved body part improves with direct visualization and verbal prompting. Improvement is assessed in movement initiation, frequency, and amplitude. Thus, for motor neglect of the hands or fingers, the clinician can ask the patient to make a fist (open and close the fingers). At first the patient is asked to look away, i.e., in the contralateral hemispace, and is given one command to “make a fist, open and close your fingers as fast as possible.” Then, the patient is asked to look directly at the involved hand or fingers, and the same command is repeated several times. For neglect testing of the foot, the patient’s task is to tap the foot.

MOTOR DYSFUNCTION AS A DIAGNOSTIC CRITERION

As described by Harden and Bruehl, several studies of the current IASP diagnostic criteria for CRPS-I have observed poor specificity (Galer et al. 1998; Bruehl et al. 1999; Harden et al. 1999). Recent published multicenter studies assessing the IASP criteria have reported that their specificity is statistically improved by including symptoms and signs of a motor disturbance (Galer et al. 1998; Bruehl et al. 1999; Harden et al. 1999). Based upon statistical analysis of data from a multicenter study, Harden and colleagues (1999) have formulated new CRPS diagnostic criteria with improved categories of signs and symptoms, including criteria for the presence of a “motor/trophic” factor. Thus, evidence is growing that motor abnormalities are not only a common feature in CRPS but also a unique feature that may be used to improve diagnostic specificity.

CONCLUSIONS

Over the past several years, many published reports have once again established motor abnormalities as an important component of CRPS. Studies have reported that motor dysfunction is among the most common symptoms and signs in CRPS and that motor signs and symptoms add specificity to the current IASP diagnostic criteria. Moreover, reports have shown that motor dysfunction may be due to “neglect” rather than to volitional guarding, giving further credence to the involvement of the CNS in some patients with CRPS.
REFERENCES


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