Posttraumatic Complex Regional Pain Syndrome (CRPS)

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Complex regional pain syndrome (CRPS) is a painful condition that most often affects limbs. Upper extremity is affected more frequently than lower extremity. The character of the pain is continuing, spontaneous, usually burning and unexpectedly severe pain considering any inciting cause. Pain does not follow the anatomical distribution of a peripheral nerve. Diagnosis is still clinical. The precipitating causes have been fractures, surgery, prolonged immobilization, nerve lesions etc. The symptoms and signs of CRPS may be difficult to recognize because of similarities in the normal early phase posttraumatic or postoperative findings. Therefore it is important to remember to suspect CRPS, if severe pain continues. Physiotherapy is the cornerstone and the first line treatment with adequate pain medication.

CRPS Classification

Complex Regional Pain Syndrome (CRPS) is a troublesome, regional, usually posttraumatic, painful condition that most often affects limbs. Previously, CRPS has been known by several names, most commonly as reflex sympathetic dystrophy (RSD), causalgia, shoulder-hand syndrome and algodystrophy. Nowadays this disorder has been named CRPS by the International Association for the Study of Pain (IASP). Two types of CRPS have been recognized. Type 1 is not associated with a specific nerve damage and Type II, although having similar symptoms and signs to those of type I, is associated with a definable nerve damage (1).

Characteristics of CRPS

CRPS usually occurs in an arm or in a leg or part of it. It occurs mainly in the distal extremity, but it can spread proximally, rarely to other areas of the body. Upper extremity is affected more frequently than lower extremity. The character of the pain is continuing, spontaneous, regional usually burning pain that does not follow anatomical distribution of a peripheral nerve. Pain is unexpectedly severe considering any inciting cause. Symptoms and signs may begin gradually within a few hours, in days or a few weeks after an injury (2–5). During the early phase after the trauma, CRPS may be difficult to recognize because of similarities in the injury-related symptoms (6). CRPS varies from relatively mild to chronic. Symptoms and signs show considerable temporal variation in degree and timing, also within the time of evaluation (7).

Incidence and etiology

The reported incidence of CRPS ranges from 5.5 to 26.2 per 100,000 person-years (8,9). Females were affected three times more often than males (9). CRPS may occur at all ages although the highest incidence is found between 50 and 70 years (9). More than 50% of CRPS cases were related to trauma. A fracture was the most common precipitating event accounting for 44% of the CRPS cases (9). Prospective studies have repeatedly demonstrated a higher incidence than retrospective studies. The incidence in the studies of distal radius fractures has been 1–2% when retrospectively reported and up to 38% when prospectively reported (10), perhaps due to the lack of uniformity and specificity of diagnostic criteria. Other initiating events
included sprains (18%) and elective surgeries (12%) (9). Also prolonged immobilization, peripheral nerve injury and nerve entrapment can be the precipitating causes (2). Other illness, such as infections, hemiplegia and myocardial infarction, can lead to CRPS as well (11). Any precipitating event may not be identified in 8-10% of the cases (12,13).

Pathophysiology

The pathophysiology of CRPS is not yet completely understood. It has been suggested that trauma or less frequently illness may cause the symptom or disorder of central nervous system. CRPS could be a regulation disorder of sensory, autonomous and motor function. Also inflammation, neuroinflammation, immunological, peripherical factors and genetic predisposition may be involved (11,14–17). Changes in the somatotrophic map in the cerebral cortex of patients with CRPS have been found. Juottonen et al. found that patients with CRPS 1 have altered central sensomotor response to tactile stimulation of the fingertip (18). The cortical representation of the painful hand was significant smaller than that of the contralateral healthy hand in the study by Pleger et al. (19). Maihöfner et al. found a significant shrinkage of the cortical hand representation contralateral to the CRPS-affected painful hand due to reorganization of the primary somatosensory cortex (20). With recovery, cortical reorganization returned normal (21). However, cortical reorganisation is not specific to CRPS. Similar findings have been found in other pain states as well (7).

Symptoms and signs

Symptoms and signs include sensory, sudomotor disturbances, trophic changes and impaired motor function (22). Symptoms of sensory dysfunction are burning pain, hyperalgesia, hyperesthesia and allodynia. Motor dysfunction may present as impaired voluntary movement, decreased range of motion, weakness, tremor, joint stiffness, coordination problems, dystonia and muscle spasms. Sudomotor changes are manifested by edema and loss of or excessive perspiration, vasomotor dysfunction by changes in temperature, discoloration of the skin. Trophic changes may occur in the skin, nails and hair, such as change in nail and/or hair growth and skin atrophy. (2,22)

Diagnosis

CRPS is a clinical diagnosis and diagnosis is still a bit controversial. There is no specific test or examination to diagnose CRPS. The surgeon has to recognize these specific symptoms and differentiate them from normal posttraumatic or postoperative findings. Recent modifications in the diagnostic criteria have improved identification of this disorder. The new diagnostic criteria was proposed in workshop in International Association for the Study of Pain (IASP) congress in Budapest in 2004. The former IASP criteria were sensitive enough, but specificity was not adequate. (23–25). According to the new criteria the clinical diagnosis of CRPS requires following symptoms and signs shown in Table 1 (24,26).

Diagnostic methods other than clinical are rarely helpful, but are usually necessary to exclude other illnesses. It is important to find other possible illnesses behind CRPS symptoms, such as tumors or entrapment syndromes, Thoracic Outlet Syndrome, compartment syndromes, discogenic disease, thrombosis, lymph edema, arthritis, infections or conversion/self-harm and dis-/non-use. However, differentiation of CRPS from other clinical conditions may be difficult. CRPS is not accompanied by abnormalities on conventional blood tests. Neuropsychological tests may be needed to exclude radiculopathy or peripheral nerve lesion. Plain radiographs may or may not show osteoporosis, which can be focal or quite widespread, but those changes are non-specific (7). MRI or CT are not diagnostic, but they may reveal other causes for symptoms. (7,11)

Treatment and rehabilitation

The main treatment objective is functional restoration and pain reduction. Scientific evidence for any superior treatment is still lacking. It is based on case reports, clinical experience, consensus panels of CRPS specialists and researchers and retrospective studies. Randomized controlled trials are few. There is evidence that the earlier the specific treatment is started, the more successful it is. Thus, the first diagnosis of CRPS is usually made in many patients weeks or months after the onset of symptoms (23). Symptoms and severity of pain determine the treatment level and rehabilitation method used, because symptoms fluctuate from relatively mild to difficult, chronic disease (27). The treatment and rehabilitation consists of pain medica-
Rehabilitation time is longer than that of without CRPS symptoms. One of the treatment methods are "training the brain"- methods, which include mirror therapy (mirror image of unaffected limb gives an illusion of affected limb functioning normally and reduces pain in early CRPS), graded motor imaginery therapy and mind integration program (34–37).

Blockades should be used mainly in situations of failure to progress or even cannot begin rehabilitation in spite of adequate pain medication. Intravenous administration of ketanserine can relieve pain in CRPS (25,31). There is no scientific evidence of intravenous or percutaneous sympathetic blocks (38). Spinal cord stimulation is effective (in carefully selected chronic patients) for pain reduction and improving quality of life, but does not improve function (11,39). An operation intended to remove underlying factor that may be responsible for CRPS symptoms (neurolysis/ nerve repair) may speed up for recovery of CRPS symptoms (40).

Secondary prevention

Netherlands guidelines recommends that surgery of the existing or previously affected extremity should be postponed until the signs and symptoms of CRPS have almost disappeared. It also recommends to keep the operation as short as possible with adequate pre-, per- and postoperative pain control to prevent sensitisation of CNS (11). Perioperative blockades of the stellate ganglion or i.v. regional blockades using clonidine 1 μg/kg (not guanethidine) can be considered, when operation is needed in the patients who previously suffered from CRPS. The use of regional anaesthesia with a sympathicolytic effect (epidural/spinal analgesia, brachial plexus blockade), either alone or in combination with general anaesthesia, can be considered as well. (11,41–42)

Prognosis

In mild or acute cases, normal function of the limb and ability to work usually recover. In moderate or subacute form the normal function and ability to work is possible, but in the severe or chronic cases the normal function and ability to work is very unlikely. Chronic CRPS accompanied by severe pain and serious disability can occur in approximately 10% of CRPS cases (13).

The right diagnosis made early enough and func-
References:


36. Moseley L: Is successful rehabilitation of CRPS due to sustained attention to the affected limb? A randomised clinical


