Complex Regional Pain Syndrome (CRPS) encompasses a myriad of injury-induced pain conditions. CRPS Type I (formerly known as reflex sympathetic dystrophy) elicits in absence of any obvious nerve injury, whereas CRPS Type II (previously termed causalgia) is associated with injury to a major peripheral nerve. However, the clinical presentations of the two types are normally indistinguishable. Limb fracture is the most common traumatic event that precedes CRPS. Lesions of the central nervous system, e.g. spinal cord injuries and stroke, can also trigger CRPS. The risk of developing CRPS following fracture is about 1-2% [1-4].

Pathophysiology of CRPS:

No single mechanism can explain the constellation of symptoms in CRPS. There is growing evidence of the role of genes in predisposing certain individuals to CRPS [e.g. 5]. Many of the symptoms, at least locally, and particularly in the acute stages, can be explained by the expression of inflammatory agents [6]. There have been reports of altered central representation in the thalamus and cortex in CRPS patients [7, 8]. Interestingly, in one CRPS patient, the symptoms almost disappeared after a cerebral contusion of the left temporal lobe, which underscores a key role of impaired brain function in sustaining CRPS-related impairments [9].

Clinical symptoms of CRPS:

The symptoms of CRPS are disproportionate in both magnitude and duration to the severity of the initiating event (where known). The distal parts of the limbs are predominantly affected. However, the proximal regions may also be affected during the latter stages. The symptoms are a combination of sensory, motor and autonomic impairments (*see* following text), in addition to the presence of a neglect-like attribute wherein the patients describe the affected limb as strange, uncontrollable, imperceptible and/or not belonging to them. The symptoms are often fairly generalised, i.e. not limited to individual spinal segments or the region of inciting lesion. For example, a fracture of the radius bone may result in the whole hand being affected. The pain can also spread to the proximal regions, especially in the chronic stages.

Sensory impairment: Almost all patients suffer from persistent pain which is described as burning/stinging in quality. It is perceived in deep tissues and is accompanied by allodynia (pain due to a normally non-painful stimulus) and hyperalgesia (increased pain from a painful stimulus). Typical features of inflammation (swelling, redness and warmth) are commonly found in the affected body part.

Motor impairment: Joints/bones are often affected resulting in joint stiffness and pain during joint movement and/or when pressure is applied to the joint. Furthermore, weakness, tremor, dystonia and coordination deficits may occur.

Autonomic impairment: Changes of skin blood flow are frequently noted, which manifest as skin temperature and skin colour asymmetries. In addition, sweating abnormalities are a common occurrence in the CRPS-state. Trophic changes such as abnormal hair and nail growth in the painful swollen region, atrophy of skin and muscle, connective tissue fibrosis, and osteoporosis may be present, especially in chronic stages.

Diagnosis of CRPS:

CRPS is a clinical diagnosis. There is no single lab test that can provide a firm diagnosis of CRPS. Plain radiographs (only positive during chronic stages), quantitative sensory testing (temperature, temperature-pain and vibratory thresholds) and temperature changes during sympathetic stimulation can be used to aid the diagnosis of CRPS. CT and MRI examinations show poor specificity for the diagnosis of CRPS. However, triple-phase bone scintigraphy is an important diagnostic tool wherein a pathological uptake of a radioactive tracer in the bones of the affected extremity is thought to be highly sensitive and specific for CRPS, albeit only during the subacute stages (up to 1 year).

Treatment of CRPS:

Pain relief is provided first and foremost to the patient that may involve the use of the following pharmacological agents: traditional nonsteroidal anti-inflammatory drugs; COX-2 inhibitors; metamizol or opioids; and corticosteroids. Adjuvants may include anticonvulsants and tricyclic antidepressants. Rest and immobilisation of the affected limb is advised during the early stages of treatment. Next to pain management, physical therapy is used to restore movement and normal function of the limb. However, if symptoms aggravate or recur, it is recommended to cease physical therapy immediately. In recent years, mirror visual feedback (visual input from a moving unaffected limb) has been shown to be effective in relieving pain and improving movement [10]. An invasive therapy such as selective sympathetic blockade is indicated when there is intractable pain and/or allodynia. Other treatments may include spinal cord stimulation and intrathecal drug pumps to introduce analgesics and anaesthetics directly into a fluid-filled space surrounding the spinal cord.

Prognosis of CRPS:

Spontaneous remission from symptoms may occur in some patients. However, the prognosis is generally poor with complete recovery of the affected limb occurring in only 25-30% of patients. The indicators of poor prognosis include

joint stiffness, low skin temperature at disease onset, contracture in early stages, dystonia, tremor, oedema and psychological comorbidity [1, 2, 4, 6].

Further information and support:

International Research Foundation for RSD/CRPS
www.rsdfoundation.org

Reflex Sympathetic Dystrophy Syndrome Association
www.rsds.org

International Association for the Study of Pain
www.iasp-pain.org

Chronic Pain Australia
www.chronicpainaustralia.org.au

Australian Pain Management Association
www.painmanagement.org.au

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