

Complex Regional Pain Syndrome

Background

1. Definition

- Chronic pain condition
- Not necessarily confined to specific nerve distribution
- Usually affecting one limb or portion of one limb
- Often characterized by allodynia
 - Pain in response to non-noxious stimuli
- Complex regional pain syndrome I
 - No demonstrable nerve damage
 - About 90% of cases
 - Previously known as reflex sympathetic dystrophy
- Complex regional pain syndrome II
 - Demonstrable nerve damage to specific nerve
 - Previously known as causalgia

2. General info

- Stage I
 - Occurs shortly after an injury or with no apparent trigger
 - Burning/throbbing pain, vasomotor instability, hypesthesia, sensitivity to heat or cold, possibly edema
- Stage II
 - Progression of soft tissue edema, skin thickening & color changes, muscle wasting, early bone demineralization
- Stage III
 - Contractures, significant osteopenia, waxy skin, brittle nails

Pathophysiology

1. Pathology of disease

- Not entirely understood
- Disease of the sympathetic nervous system
- Multiple theories including:
 - Sympathetic nervous dysfunction related to nociception
 - Peripheral pain receptors hypersensitive to circulating catecholamines
 - Changes in pain sensing in dorsal horn of spinal cord

2. Incidence, prevalence

- 1-5% of patients who have sustained peripheral nerve damage
- 30% in patients post-Colles fracture
- Many cases resolve spontaneously over 9-12 weeks

3. Risk factors

- Precipitating event often unknown
- Inactivity can increase risk, especially after trauma
 - Trauma:
 - Sprain, fracture, nerve injury
 - Iatrogenic:
 - After carpal tunnel release or arthroscopic surgery, venipuncture, IM injection
 - Medical conditions:
 - Diabetic neuropathy, malignant invasion of nerve plexus, etc

- Psychosocial stress is NOT a risk factor, but can develop as a result of dz
4. Morbidity
- Osteopenia (from inactivity), muscle atrophy, substantial psychosocial stress related to pain management

Diagnostics

1. Dx based on Hx & PE (SOR:B)
 - May take weeks to months to Dx
2. History
 - Chief complaint
 - Severe dysesthesia
 - Spontaneous occurrence of pain in absence of painful stimuli
 - Allodynia
 - Painful response to thermal or mechanical stimuli that would not normally cause pain
 - Hyperesthesia
 - Prolonged/exaggerated response to painful stimuli
 - Pain
 - Throbbing, burning, constant, or aching
 - Often begins days or weeks after triggering incident, and lasts much longer than anticipate for normal healing
 - Often triggered by change in temperature, light touch, movement, or psychosocial stress/excitement
 - Can progress proximally to involve entire limb
3. Physical exam
 - Compare affected & unaffected limb for:
 - Color
 - Warmth
 - Sensitivity to touch
 - Edema
 - Atrophy
 - Progressive changes may occur
 - Initially affected area warm, erythematous and dry, with accelerated hair and nail growth
 - Hair then becomes sparse, nails grooved and brittle
 - Skin becomes cool, cyanotic and moist
 - Swelling and vasomotor changes can lead to skin dystrophy
 - Soft puffy edema changes to tight, shiny swelling and loss of skin creases
 - Atrophic limb develops
 - Decrease in fat pads, digits thin & pointed, muscle spasms & wasting, joint thickening
 - Eventually marked bone and muscle atrophy, weakness and flexor tendon contractures
4. Diagnostic testing
 - No definitive testing available
 - Diagnostic imaging
 - X-ray: may show osteopenia (69%)³
 - Bone scan may be helpful if x-ray non-diagnostic

- Other studies
 - Immersion in ice water triggers substantial pain similar to pain patient describes as characteristic
- Diagnostic criteria
 - Listed below are the International Association for the Study of Pain diagnostic criteria^{4,5}
 - Note: other acceptable criteria include Bruehl's & Veldman's
 - **Complex regional pain syndrome type I**
 - Presence of initiating noxious event or cause of immobilization (this criterion not necessary for dx)
 - Continuing pain, allodynia or hyperalgesia with which pain is disproportionate to inciting event
 - Evidence at some time of edema, changes in skin blood flow or abnormal pseudomotor activity in painful region
 - Dx excluded by existence of conditions that would otherwise account for degree of pain and dysfunction
 - **Complex regional pain syndrome type II**
 - Continuing pain, allodynia or hyperalgesia after nerve injury, not necessarily limited to distribution of injured nerve
 - Evidence at some time of edema, changes in skin blood flow or abnormal pseudomotor activity in painful region
 - Dx excluded by existence of conditions that would otherwise account for degree of pain + dysfunction

Differential Diagnosis

1. Key DDx

- Musculoskeletal:
 - Sprain, fracture, bursitis, myofascial pain syndrome, rotator cuff tear
- Neurologic:
 - Postherpetic neuropathy, diabetic neuropathy, radiculopathy
- Infectious: cellulitis
- Psychosomatic:
 - Munchhausen's, malingering (esp. when 2^o gain issues exist, e.g., workers' compensation)

2. Extensive DDx

- Vascular: Raynaud's, vasculitis
- Rheumatic: SLE, RA

Therapeutics

1. Develop clear treatment plan, mutual goals and expectations for therapy

- Spontaneous resolution is frequent
- Watchful waiting and psychological support appropriate early in course
- Initiate medical and physical therapy simultaneously for optimal effect
- Expect some effect over 2-3 weeks
 - Gradual resolution of Sx over months
- Educate patient that pain does not represent ongoing injury, that mobilization & desensitization of affected area are keys to successful recovery
- Medical:
 - Listed therapies may have some effect, and are unlikely to cause harm

- Topical DMSO
 - Particularly helpful for analgesia during therapy (SOR:B)
- IV bisphosphonates
 - Requires 4-8 weeks of therapy, good long term effect (SOR:A)
- Calcitonin (SOR:B)
- Oral corticosteroids (limited course) (SOR:B)
- Vitamin C 500 mg daily for 50 days may reduce the risk of CRPS for pts with a wrist fracture
- Physical therapy
 - Mobilization of affected limb important, but painful

Follow-Up

1. Return to office
 - Follow-up by phone and/or in person to provide psychological support and assess efficacy of watchful waiting and/or interventions such as medical and physical therapy
2. Refer to specialist
 - Not indicated unless diagnostic uncertainty
3. Admit to hospital
 - Not indicated unless unable to manage pain as outpatient (rare)

Prognosis

1. Not well documented
2. Most patients do experience lessening of symptoms with combined medical, physical, and psychological therapy
3. Many eventually reach level of remission that allows for normal daily activities

Prevention

1. Early mobilization after trauma (SOR:C)^{1,2}
2. Vitamin C 500 mg PO qD for 60-90 days reduces the risk of CRPS in elderly patients with wrist fractures (SOR:A)

Patient Information

1. Handout from American Academy of Family Physicians
 - <http://familydoctor.org/handouts/238.html>
2. Handout including organizations for more information from National Institute of Neurological Disorders and Stroke (NINDS)
 - http://www.ninds.nih.gov/disorders/reflex_sympathetic_dystrophy/reflex_sympathetic_dystrophy.htm

References

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2. Quisel AM, Gill JMM, MPH, Witherell PM. Complex regional pain syndrome underdiagnosed: CRPS type 1 is an under-recognized problem in limbs recovering from fracture or immobilized post-stroke. Journal of Family Practice. 2005;54(6):524-532.

3. El-Khoury G, Bennett D, Dalinka M, et al. Chronic Foot Pain. American College of Radiology. Available at:
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PURLs

1. Give Vitamin C To Avert Lingering Pain After Fracture

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