AN IN-DEPTH LOOK AT C.R.P.S – FROM DIAGNOSIS TO TREATMENT

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DIFFERENTIAL DIAGNOSIS

- Diabetic and small-fiber peripheral neuropathies
- Entrapment neuropathies
- Thoracic outlet syndrome
- Discogenic disease
- Deep vein thrombosis
- Cellulitis
- Vascular insufficiency
- Lymphedema
- Costochondritis
- Brachial Plexopathies

THE DISEASE

The pain is profound, chronic and widespread. It can migrate to all parts of the body and vary in intensity. The pain has been described as stabbing and shooting pain and deep muscular aching, throbbing, and twitching. Neurological complaints such as numbness, tingling, and burning are often present and add to the discomfort of the patient. The severity of the pain and stiffness is often worse in the morning. Aggravating factors that affect pain include cold/humid weather, non-restorative sleep, physical and mental fatigue, excessive physical activity, physical inactivity, anxiety and stress.
Additional symptoms may include: irritable bowel and bladder, headaches and migraines, restless legs syndrome (periodic limb movement disorder), impaired memory and concentration, skin sensitivities and rashes, dry eyes and mouth, anxiety, depression, ringing in the ears, dizziness, vision problems, Reynaud's Syndrome, neurological symptoms, and impaired coordination, sleep disturbance and fatigue.
The preceding two slides are directly from the National Fibromyalgia Website
In MY opinion Fibromyalgia IS CRPS
REFERENCES

Is Fibromyalgia a Generalized Reflex Sympathetic Dystrophy?-

Fibromyalgia and the Complex Regional Pain Syndrome: Similarities in Pathophysiology and Treatment
Richard J Wurtman – “Metabolism Clinical and Experimental”- 59-2010 -837-840
MULTIPLE SCLEROSIS

I have studied nine patients, all female, who have been diagnosed with BOTH MS and CRPS. Treatment was initiated to the MS by neurologists and ALL nine showed improvement of the MS symptoms while seven showed improvement of the CRPS.
SYMPTOMS

- Sensory
  - Burning pain
  - Skin, sensitivity to touch
    - Skin, sensitivity to cold or heat

- Vasomotor
  - Abnormal skin color changes
  - Abnormal skin temperature changes
Symptoms

- **Sudomotor / Edema**
  - Abnormal sweating
  - Abnormal swelling

- **Motor / Trophic**
  - Limited movement
  - Weakness
  - Tremor
  - Dystonia
  - Neglect
  - Abnormal hair and/or nail growth
Vasomotor Changes
Abnormal Sweating
MOTOR DISTURBANCE-DYSTONIA
NEUROGENIC EDEMA
ERYTHEMA
EDEMA & ERYTHEMA
ORGAN INVOLVEMENT
CARDIAC
The patient whose EKG strip appeared on the previous slide was a 45 y/o female who presented in the ER following a syncopal attack. She had a history of Type II diabetes but blood sugar was 160. She had a history of thyroid dysfunction but TFS were all WNL. She had no other medical history save for her CRPS.
ARRHYTHMIAS

BRADYARRHYTHMIAS – Usually in female patients over 40

TACHYCARDIAS – Usually in female patient in their 20’s

ATYPICAL RHYTHMS
Many patients complain of chest pain that, following a comprehensive cardiac work-up proves to be a result of CRPS. The etiology appears to be sensitization of the Intercostobrachial nerve in patients with a brachial plexus injury.

A 50 year old male has a massive M.I. necessitating quadruple bypass surgery. The operation goes flawlessly and he awakens in more pain than when the surgery began – that pain accelerates and does not subside 12 years later.
VISUAL DISTURBANCE

DOUBLE VISION
BLURRED VISION
OCCULAR MIGRAINES
VISION LOSS
PHOTOPHOBIA
BURNING OF THE EYES
HEADACHES

MIGRAINES(?)

TENSION HEADACHES

GREATER OCCIPITAL NEURALGIA
Patients report significant otophobia.

Recently there has been an increase in individuals reporting significant discomfort from vibration, specifically the bass of stereos even through walls and from adjacent motor vehicles while travelling.

Intermittent and transient hoarseness comes from the effect of the disease on the branchial plexus and is frequently misdiagnosed as immune compromise.
I have one patient who in addition to pain etc. began to stutter uncontrollably as the CRPS worsened.

After one week of IV Ketamine he stopped stuttering altogether.
Unfortunately dental disease is rampant in patients with CRPS.

Part of this stems from dietary indiscretions, part from immune system compromise and part from the disruption of the dental nerve roots.

Perhaps the greatest reason is that the side effects of common medications prescribed for chronic pain lead to a change in lifestyle, poor oral hygiene, poor nutrition and a loss of saliva (dry mouth) that result in decay, periodontal disease and ultimately tooth loss.
The most common finding apart from dry skin or hyperhidrosis is neurodermatitis. This can occur randomly on any area of the body. Lesions have the appearance of small acne-type eruptions that itch for hours to days and disappear spontaneously. There is no specific etiology apart from the CRPS and no treatment save for topical low potency steroids or anti-histamines to reduce the itch. If scratched they will scar.
Dercum's disease, is a rare condition characterized by multiple, painful lipomas. These lipomas mainly occur on the trunk, the upper arms and upper legs. The understanding of the cause and mechanism of Dercum's disease remains unknown. Possible etiologies include: nervous system dysfunction, mechanical pressure on nerves, adipose tissue dysfunction and trauma.
LIVIDO RETICULARIS
Apart from the obvious acid peptic and irritable bowel symptoms, we have to deal with intractable nausea and vomiting. Endoscopically there may be some mild gastric irritation but generally the findings are minimal. Conventional treatment is rarely effective. The etiology is clearly gastroparesis and objectively identified via gastric emptying studies.

We have had great success with the endoscopic administration of Botox into the pyloric sphincter. In many instances one to three such injections have stopped the vomiting for prolonged periods of time.
Our only two failures with botox were both females with intractable pain, nausea and vomiting and severe malnutrition and weight loss.

They both ultimately underwent fundal plication surgery which was successful in reversing the GI abnormality.

THEY ARE SISTERS!
Another interesting finding is a number of patients with clinical and laboratory confirmed pancreatitis with no other etiology evident save for their CRPS.

Other symptoms include:
- Dysphagia, indigestion
- Diarrhea / constipation (opioid induced)
- Biliary dyskinesia
Urinary

Commonly, patients experience urinary incontinence, dysuria or inability/difficulty voiding. The condition is usually misdiagnosed as Interstitial Cystitis. The problem has responded marginally to conventional medications. Bladder pacemakers have been somewhat useful. Again, Botox injections into the pelvic floor have helped a great number of sufferers. Additionally, I have found that lumber epidural infusions of bupivacaine over a 5 day period works very well. Ketamine has resolved this to a small degree as well.
Menstrual dysfunction of all types is noted:

- Polymenorrhea
- Dysmenorrhea
- Menometorrhagia
- Secondary amenorrhea
An interesting finding is that patients in the third trimester of pregnancy (and some earlier in their pregnancy) become dramatically less symptomatic and many become asymptomatic. This lasts into and after childbirth and seems to be further extended by breast feeding. I currently have data on nine such cases and am exploring the hormonal shift that may be common to all of these individuals.
Morton’s neuroma is a mechanically induced degenerative neuropathy predominantly affecting the second and third common digital nerves. It is not actually a tumor but a thickening of the tissue that surrounds the nerves leading to the toes. It is eight to ten times more likely to occur in women than in men and most prevalent in middle aged women.
Vertigo is common as an early symptom. Sometimes it is positional but mainly it is movement related.

Treatment is based upon reducing the CRPS symptoms and occasionally meclizine helps make the vertigo tolerable pending the improvement of the disease process.
“Syncope is common in patients with CRPS especially with lower limb involvement. Autonomic dysregulation of the lower extremities leads to sympathetic vasoconstriction and venous pooling which can predispose these patients to syncope.”

Syncope in Complex Regional Pain Syndrome – Smith et al. – *Clinical Cardiology* 34.4; 222-225 (2011)
A 44 year old female with long standing history of CRPS is involved in a MVA which accelerates her symptoms. She further injures her brachial plexus in the accident and has classic symptoms of that sub-division of CRPS. However she begins to have “drop attacks” with increasing regularity.

Comprehensive work up with brain MRI, EEG, laboratory testing and carotid ultrasound all prove negative.

The solution proved to be immobilization in a soft cervical collar. Here is how that transpired……..
COGNITIVE DYSFUNCTION

There have been an increasing number of CRPS patients with cognitive issues. Mostly these are STML, word retrieval & difficulty with expression. It has been theorized that this is medication related but it occurs in individuals who take virtually no meds. Current thoughts abound with no single answer surfacing as being definitive.
CONSTITUTIONAL

- Lethargy
- Fatigue
- Weakness
- Sleep disturbance
OTHER SYMPTOMS

- Shortness of breath
- Inability to take a deep breath
- Neurogenic edema
- Muscle weakness/atrophy
- Endocrine dysfunction – adrenal, thyroid, hormonal imbalance
- Gardner Diamond Syndrome – spontaneous bruising in uninjured areas

“Systemic Complications of Complex Regional Pain Syndrome”
Robert J. Schwartzman - Neuroscience & Medicine, 2012, 3, 225-242
ANXIETY & DEPRESSION

- Chronic pain = Depression
- Depression begets anxiety
- Benzodiazepines worsen depression
- SSRI’s do not work
- SNRI’s works marginally

Currently my belief lies in the balancing of neurotransmitters
A 37 year old female casino worker is struck by a “money cart” in the left lateral thigh and subsequently develops CRPS in that limb. It later migrates to the left arm.

One year later, her sister, a 35 year old police officer was broadsided in her patrol car while driving. The door handle impacts her left lateral thigh and SHE develops CRPS in the left leg which within months migrates to the left arm!
Currently I treat 18 families with more than one member who has CRPS.

There is one article in the literature that has studied genetics and CRPS – 31 families were studied, two families had five afflicted members, four families had four, eight families had three and 17 had two.

_Familial occurrence of complex regional pain syndrome –_  
THERMOGRAPHY

“HEAT PICTURE”
A hallmark of CRPS is an excessive vasoconstriction of blood vessels that can cause cold hands and feet.

Because CRPS produces these neurovascular changes, infrared imaging, in a number of studies, has demonstrated a high degree of accuracy picking up these symptoms.
Validation of Thermography in the Diagnosis of Reflex Sympathetic Dystrophy

Dec. 1996, pp. 316-325

Long term skin temperature measurements – A practical diagnostic tool in complex regional pain syndrome

Krumova et al – Pain 140 (2008) 8-22
What Causes (Cold) Emissions? (Hypothermic)

Vasoconstriction
Tendons
Less Muscle Mass
Nerve Atrophy & Nerve Dysfunction
Sympathetic Stimulation
Increased Fibrous Tissue
Chronic Injuries
Chronic Spasm
Disuse & Atrophy
Muscle Inactivity
Chronic Scarring
A 40 y/o female was seated on the third row of a football stadium at a charity event when one of the players kicked a ball into the crowd. A fan threw the ball down attempting to reach the field but instead impacted the patient who, in an attempt to prevent getting hit in the face put her left arm up and was struck by the tip of the ball, fracturing her hand.
Max Temp: 29.0°C 26.7°C 27.6°C 25.6°C 29.0°C 25.4°C
Avg Temp: 29.3°C 26.1°C 26.7°C 24.7°C 28.1°C 25.1°C
Min Temp: 27.3°C 24.3°C 20.4°C 24.0°C 27.1°C 24.6°C
Delta Avg: 2.2°C 2.0°C 3.0°C
Shape: Circle
Color: Circle

21.8°C
It has long been my opinion that thermography has a “predictive” value in the evaluation of patients with CRPS.

The following is one example.
CASE HISTORY

A 43 year old female was an airline passenger wearing sandals. In the landing process she cut her foot on a piece of wire or metal that was sticking up from the plane.

When examined in my office 2 ½ years later she complained of foot and ankle pain with a “squishy feeling.”
Each fact is suggestive in itself. Together they have a cumulative force.

Sherlock Holmes