CASE REPORT

Complex Regional Pain Syndrome Type 1 Treated with Vitamin C

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ABSTRACT

Complex regional pain syndrome, formerly reflex sympathetic dystrophy or causalgia, is a chronic progressive disease characterized by severe pain, swelling, and changes in the skin. It often affects an arm or a leg and may spread to another part of the body and is associated with dysregulation of the autonomic nervous system resulting in multiple functional loss, impairment, and disability. It occurs mostly in of adolescence. Though treatment is often unsatisfactory, we report a young girl treated satisfactorily with vitamin C.

Keywords: CPRS, autonomic nervous system, vitamin C

The condition currently known as complex regional pain syndrome (CRPS) was originally described during the American Civil War by Silas Weir Mitchell, who is sometimes also credited with inventing the name "causalgia."[1] also known as reflex sympathetic dystrophy (RSD). It is a chronic progressive disease characterized by severe pain, swelling, and changes in the skin. It often affects an arm or a leg and may spread to another part of the body and is associated with dysregulation of the autonomic nervous system resulting in multiple functional loss, impairment, and disability. It is usually seen in adolescent girls but has been described in children.[2,3] Vitamin C may have a therapeutic role related to its antioxidant properties; vitamin C deficiency has not been implicated as cause of CRPS. We are reporting scurvy and CRPS in the same patient.

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CASE

A 3-year-old female child presented with inability to walk, pain in both lower limb and she was bedridden from 6 months. After a minor accident, patient developed swelling of both knees and ankles, she had excessive pain to light touch and excessive sweating. Diet was adequate in proteins and calories. Developmental milestones were normal.

On examination patient is conscious, irritable vitals were stable. Patient was pallor, spongy gums with bleeding along with petechiae and hyperkeratosis on lower limbs. There was no lymphadenopathy or hepatosplenomegaly. Central nervous system examination revealed normal tone, power and reflexes in all limbs. There was hyperesthesia in both lower limbs.

On local examination she was not moving her lower limbs both lower limbs were in flexed attitude. Temperature of the swollen part was raised.

As given in Gerald Fenichel's Clinical Pediatric Neurology, we immersed the affected limbs in warm water. Wrinkling of the skin of toes was absent as compared to wrinkling of hands. As wrinkling of fingers and toes requires intact sympathetic innervations this manure is helpful in diagnosing.

Hemoglobin was 6 g/dL, total leukocyte count (TLC) of 11,000/mm^3 and platelet count was 6,12,000/mm^3. Peripheral smear revealed microcytic, hypochromic anemia; there were no abnormal cells. Creatine phosphokinase (CPK) and venereal disease research

laboratory (VDRL) were normal. X-ray hip and knee joints revealed pencil thin cortex, decreased bone density and white line of Frankel, suggestive of scurvy. A provisional diagnosis of scurvy and CRPS type1 was made.

Hundred milligram of oral vitamin C was given daily. Hyperesthesia started improving and child was able to walk with support within 15 days of treatment. After 1 month duration, she was walking independently and was perfectly normal.

DISCUSSION

CRPS, formerly RSD or causalgia, is a chronic progressive disease characterized by severe pain, swelling, and changes in the skin. It often affects an arm or a leg and may spread to another part of the body and is associated with dysregulation of the autonomic nervous system resulting in multiple functional loss, impairment, and disability. Though treatment is often unsatisfactory, early multimodal therapy can cause dramatic improvement or remission of the syndrome in some patients.[4] The International Association for the Study of Pain has proposed dividing CRPS into two types based on the presence of nerve lesion following the injury.

- Type I, formerly known as RSD, Sudeck's atrophy, reflex neurovascular dystrophy, or algoneurodystrophy, does not have demonstrable nerve lesions.
- Type II, formerly known as causalgia, has evidence of obvious nerve damage.

CRPS can strike at any age, but the mean age at diagnosis is 42.5 CRPS has been diagnosed in children as young as 2 years old.[6] It affects both men and women; however, CRPS is 3 times more frequent in females than males.[5] The number of reported CRPS cases among adolescents and young adults is increasing.[7]

There may be bilateral involvement.[2] The pathophysiology of CRPS remains uncertain. It may be due to sympathetic dysfunction, central dysfunction or an inflammatory process. However recent research has suggested that oxidative damage (e.g., by free radicals) may play a role.[5]

The International Association for the Study of Pain (IASP) lists the diagnostic criteria for CRPS I (RSDS) as follows:

- The presence of an initiating noxious event or a cause of immobilization.
- Continuing pain, allodynia (perception of pain from a nonpainful stimulus), or hyperalgesia (an exaggerated sense of pain) disproportionate to the inciting event.
- Evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the area of pain.
- The diagnosis is excluded by the existence of any condition that would otherwise account for the degree of pain and dysfunction.

The IASP criteria for CRPS I diagnosis has shown a sensitivity ranging from 98-100% and a specificity ranging from 36-55%. Per the IASP guidelines, interobserver reliability for CRPS I diagnosis is poor. Two other criteria used for CRPS I diagnosis are Bruehl's criteria and Veldman's criteria which have moderate to good interobserver reliability. In the absence of clear evidence supporting one set of criteria over the others, clinicians may use IASP, Bruehl's, or Veldman's clinical criteria for diagnosis. While the IASP criteria are nonspecific and possibly not as reproducible as Bruehl's or Veldman's criteria, they are cited more widely the literature including treatment trials.[8]

According to Veldman, et al.[5] diagnosis of CRPS can be made clinically if:

- At least 4 of the 5 symptoms and signs are present: unexplained diffuse pain, altered skin color, altered skin temperature, edema and reduced active range of movements.
- Symptoms aggravated by activity of the extremity.
- Symptoms are present in an area much larger than and distal to primary injury.

All these features were seen in our patient. International Association for Study of Pain criteria are also similar, with nerve conduction velocity (NCV) and electromyography (EMG) required to distinguish between type 1 and 2, though the clinical validation of these criteria are still debated.[2] Veldman's criteria are most widely used.

No specific test is available for CRPS and diagnosis is primarily through observations of symptoms. However thermography, sweat test, X-ray and sympathetic blocks can be used to build up picture of the disorder.[9] EMG/NCV can help differentiate early phases of CRPS type 2. Scintigraphy and bone scan have a sensitivity of 72% and 50%, respectively.[10,11] Absence of abnormal tests does not preclude diagnosis of CRPS. Early diagnosis is the mainstay of successful treatment of RSD. Management consist of physiotherapy, sympathetic blocks, epidural

blocks, drug treatment (alpha blocker, calcium channel blocker, nonsteroidal anti-inflammatory drug (NSAID), calcitonin, corticosteroid, antidepressant) and surgical sympathectomy.[12,13]

Vitamin C could have some efficacy related to its antioxidant properties. One double-blind study showed that vitamin C given to patients with wrist fractures reduced the incidence of CRPS.[14]

In two placebo-controlled randomized clinical trials Zollinger et al. show that 500 mg vitamin C daily, reduces the chance for the occurrence of CRPS after wrist fractures.[14]

In teens and younger patients with CRPS, the prognosis is excellent. Most of the patients improve markedly without invasive therapy, 75% of children have full recovery. Long-term sequelae include shortening of limbs or foot because of prolonged immobilization and osteoporosis.[5]

Our patient had showed response to vitamin C administration, so there might be some association between scurvy and CPRS.

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