

Complex Regional Pain Syndrome Type 1 and Scurvy

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A 5 year old female developed features of complex regional pain syndrome (CRPS) i.e excessive pain to touch, decreased sweating and edema of left ankle 2 years after fracture of left tibia. Gum bleeding, petechiae and pseudoparalysis and suggestive radiograph characterized scurvy. Hyperesthesia improved and child walked with support following administration of vitamin C.

Key words: Complex regional pain syndrome, Reflex sympathetic dystrophy, Scurvy.

Complex regional pain syndrome (CRPS), formerly known as Sudeck's dystrophy or reflex sympathetic dystrophy (RSD) is characterized by pain, sensory and vasomotor disturbances, trophic changes and impaired motor function(1). 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.

CASE REPORT

A 5 yr old female child presented with inability to walk, pain in left lower limb and gum bleeding of four months duration. After minor trauma to the left lower limb, patient developed swelling of left ankle, excessive pain to light touch and excessive sweating. The child had an accidental fracture of left tibia 2 yr back, following treatment she had a normal walking pattern. Diet was adequate in proteins and calories. Developmental milestones were normal.

The girl was conscious, apprehensive and irritable. She was of average built and nutrition. She had 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. Bulk of left thigh muscles was less as compared to right thigh (mid thigh circumference 24.5 vs 26.0 cms). Left lower limb was shorter by 2 cms than the right. There was hyperesthesia in both lower limbs but left lower limb was more sensitive to light touch.

Hemoglobin was 7.2g/dL, TLC of 12,200/mm³ and platelet count was 3,12,000/mm³. Peripheral smear revealed normocytic, normochromic anemia; there were no abnormal cells. Her serum calcium level was 12.9 mg/dL, serum alkaline phosphatase was 368.2 U/L, CPK19 IU /L and VDRL was nonreactive. X-ray knee joint revealed pencil thin cortex, decreased bone density and white line of Frankel, suggestive of scurvy. A provisional diagnosis of scurvy and CRPS type 1 was made.

The child was administered 100 mg of oral vitamin C daily. Hyperesthesia started improving and child was able to walk with support after 4 days of treatment. After 15 days, she was walking independently with some limping due to shortening of left lower limb.

DISCUSSION

CRPS is disease of adolescents and adults, its incidence in children is low(2,4). In CRPS type I there is no nerve lesion. There is a definite nerve lesion in type 2 (causalgia). Precipitating factors includes trauma (including surgical), CNS disorders, inflammatory arthropathy, visceral lesions (myocardial infarction) or the manifestation may be idiopathic, especially in children(2). The onset of symptoms may follow a trivial injury such as simple twisted ankle or sprain or it may not be associated with a definite event(3). The onset is heralded by severe pain and exquisite tenderness to light touch, including that of clothing. Symptoms are intensified by weight bearing and relieved by keeping the involved area as motionless as possible(3).

CRPS most commonly affects the extremities with hand, wrist, knee, ankles and foot being the commonest. Occasionally the whole limb is involved. 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).

According to Veldman, *et al.*(5) diagnosis of CRPS can be made clinically if (i) 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 (ii) symptoms aggravated by activity of the extremity; and (iii)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 NCV and 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(6). EMG/ NCV can help differentiate early phases of CRPS type 2.

Scintigraphy and bone scan have a sensitivity of 72% and 50%, respectively(7,8). 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, NSAID, calcitonin, corticosteroid, antidepressant) and surgical sympathectomy(9,10). 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(4).

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).

Association of CRPS and scurvy in this case suggests that vitamin C deficiency may have a therapeutic role in its management.

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