Reflex Sympathetic Dystrophy Syndrome

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Abstract
Reflex Sympathetic Dystrophy (RSD) or Complex Regional Pain Syndrome Type-I (CRPS-I), a disease of unknown prevalence, complicates any minor trauma, stroke, myocardial infection, colle’s fracture, peripheral nerve injury and in one-fourth of cases without any precipitant factor. An awareness of RSD and the injuries, illnesses and drugs that can provoke it is the first step to learn for an early treatment and better outcome. Here we present a neglected case of RSD following minor trauma who presented to us after 6-7 months of onset of disease. Delay in treatment resulted in partial recovery of the patient.

Key Words
Reflex sympathetic dystrophy (RSD), Complex regional pain syndrome-I (CRPS-I), Complex regional pain syndrome-II (CRPS-II), Causalgia

Introduction
Reflex sympathetic dystrophy (RSD) is a syndrome of extremity pain, swelling, stiffness and discolouration often leading to disability, usually occurring after trauma or in association with a disease or a drug. Different names are used for RSD, including algodystrophy, Sudeck’s atrophy, shoulder-hand syndrome, Post infarction sclerodactyly, sympathetic trophoneurosis. However, at a consensus in 1993, the International Association for the Study of Pain (IASP) coined the term complex Regional Pain Syndrome type-I (CRPS-I) for RSD and CRPS type-II was defined as causalgia, a painful dystrophy following injury of a peripheral nerve. CRPS type-I is felt to be a spectrum of entities, with pain as the primary characteristic of the illness, “Complex” refers to its variability over time and from person to person and also refers to the plethora of inflammatory, autonomic, motor, cutaneous and dystrophic signs that accompany the neuropathic pain, “Regional” reflects the involvement of an area beyond the area initially injured (1). Nevertheless, for historical reasons, RSD is used more commonly. Lack of awareness of this entity leads to delay in diagnosis and treatment & thus residual deficits and deformities. We report a similar experience where late reporting, delay in treatment and poor follow up could not give complete relief to the patient.

Case Report
A 65 year old female, non-diabetic, non-hypertensive, non-smoker, non-alcoholic presented with the chief complaints of pain and swelling at left wrist joint for last 6-7 months; stiffness and restriction of movement at the same joint for last 2-3 months. Six months back, patient was apparently alright when she started with pain in the left dorsum of the hand which was severe in...
intensity, continuous in nature, burning and without any
diurnal variation. Patient gave a history of minor trauma
teen days before the onset of pain. There was an
aggravation in the intensity as well as duration of pain
by any motion of the extremity or by dependency and
used to get relief whenever she used to keep her extremity
elevated or wrapped in a moist cloth or wet dressing.

Pain was associated with swelling and was pitting in
nature and in the early stage was soft but as the illness
progressed, the swelling became hard, brawny and non-
pitting. Since last 3 months, there was history of
limitation of movement at wrist joint with sparing of
movements at elbow joint. On examination, the wrist
joint was flexed with flexion deformity at small joints
of hand; skin overlying the hand was shiny, edematous,
smooth and glossy with fewer wrinkles. However, no
abnormal pigmentation, pustular lesions, telangiectasia,
ulcers and cellulitis was seen. No motor abnormalities,
trauma, myoclonus etc. could be documented. All the
peripheral pulses were palpable and the extremity of the
opposite side was normal without any deformity.

On investigations, complete blood count, renal
function test, liver function test, sedimentation rate,
C-reactive protein, urine examination and basic
autoimmune profile were normal and non-conclusive.
Chest x-ray (PA view) and ultrasound abdomen was
normal. X-ray of the hands including wrist joints typically
showed patchy or spotty osteopenia with irregular
resorption of trabecular bone on left side (Fig. 5). Patient
was subjected to bone scanning with technetium and
findings suggestive of a diagnosis of RSDS of left hand
or are depicted in different figures 6 to 8.
Findings of bone scan in different phases are depicted below showing more rapid accumulation of radionuclide in left hand (Fig. 6 & 7) and increased radionuclide in the delayed scan (Fig. 8).

**Fig. 5**: X-ray of the hand and wrist showing osteopenia of the left wrist joint.

**Fig. 6**: Phase-I

**Fig. 7**: Phase-II

**Fig. 8**: Phase-III

Treatment was started with a multimodal and coordinatd pattern. Patient was advised to elevate the limbs and was given psychological support. Steroids were started and as per Kozin recommendation, 28 day course was given with 15 mg qid x 4 days, then 10 mg qid x 4 days, then 10 mg tid x 4 days and finally 5 mg qd x 4 days. In addition, analgesics, calcium and bisphosphonates were also prescribed. Besides drugs, physiotherapy was also advocated. Patient had lot of relief in the pain and swelling but did not get complete recovery in the movements & deformity. She remained on follow up for 2 months & did not report after that.

**Discussion**

Prevalance of RSD (CRPS type-I) is not known and can be associated with a number of diseases, drugs or injuries. Among adults, women are slightly more prone to RSD than men. More than 25% of the time, no associated precipitant is found. Tauma is the most common inciting factor, but the trauma can be very minor. Our patient did suffer from minor trauma. Reported incidence ranges from 5% to 20% with myocardial infarction, 12% to 21% with hemiplegia, 0.2% to 35% after colles fracture and 2% to 5% after peripheral nerve injury (2), certain stressful life events like depression, anxiety etc. increase the risk of RSD (3).

Steinbrocker and coworkers described three clinical stages of RSD (4). The first, acute stage, is characterised
by soft edema, pain, paresthesia, decreased motion, and increased sweating. The second, dystrophic stage, from 3 to 9 or 12 months after the initial symptoms, is marked by continued pain, stiffening and brawny edema and vasospasm with cooling of hand. The third, atrophic stage, begins at 9 to 12 months and is characterised by stiffness and flexion contractions in the hand with continued pain on attempted motion. However, there is no clear cut progression from one stage to another and our patient was most probably in second, dystrophic stage. Dividing RSD into warm onset and cold onset based on the observed or recorded temperature in the extremity at the onset of illness is useful prognostically (5).

RSD is bilateral in at least 25% of cases clinically (6), bone scanning and dolorimeter testing indicate a degree of bilateral presentation in a far higher percentage and involvement of three or more limbs has been reported (7). However, in our patient only one limb was involved.

Resting sweat output has a 94% specificity for RSD but have a poor sensitivity. The sensitivity of sonography ranges from 54% to 100% and the specificity ranges from 85% to 98%. The delayed images seem to be the most sensitive in diagnosing RSD (8).

Awareness of RSD and the injuries, illnesses and drugs that can provoke it is the first step in the treatment of RSD. It is thought that early mobilization after myocardial infarction, trauma and stroke can lessen the likelihood of developing RSD. Early treatment leads to a better outcome. Rosen and Graham (9) found an excellent result in 43% with symptoms less than 6 months, but only 20% with longer duration did as well. A number of drugs like α-blockers, x2 agonists, calcitonin, bisphosphonates, anti emulsants, steroids and various regional blockade have been tried with variable results (10). Elevated resting skin temperature, edema, disease duration of less than 6 months and warm type are the best predictors of a good response. Our patient had a warm type, disease duration of 6-7 months, edema, raised temperature and some flexion deformity at wrist and was treated with steroids, calcium, bisphosphonates. Physiotherapy was also advocated. There was a marked improvement in pain and swelling. However, the flexion deformity did not show much improvement as expected. It is stressed that RSD should be discussed more so that there is awareness amongst the treating doctors to pick up and treat these patients at the earliest and avoid any morbidity.

References