Paraplegia **28** (1990) 48–54

0031–1758/90/0028–0048$10.00

Paraplegia

Reflex Sympathetic Dystrophy in Central Cord Syndrome: Case Report and Review of the Literature

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Summary

Reflex sympathetic dystrophy (RSD) has been reported in incomplete spinal cord injury patients, most often occurring unilaterally; however to our knowledge, bilateral RSD has not been reported in patients with a central cord syndrome. We report a case of bilateral RSD in a patient with incomplete cervical myelopathy and the clinical picture of central cord syndrome. Diagnosis of RSD was based upon clinical, roentgenographic and scintigraphic findings. Management of RSD included elevation of forearm and hands, gentle active and passive range of movements of all upper extremity joints and systemic corticosteroids. With treatment, pain subsided, the range of motion of the joints improved and the patient achieved good functional recovery.
Key words: *Reflex sympathetic dystrophy; Spinal cord compression; Cervical spondylosis; Central cord syndrome; Rehabilitation*

Reflex sympathetic dystrophy (RSD) is characterised by severe burning pain, oedema of extremity, limited range of motion of the involved joints, vasomotor instability, trophic changes and delayed functional recovery. RSD has been only rarely reported in patients with myelopathy, most often symptoms are unilateral and infrequently symptoms may be bilateral. In a retrospective study of 125 patients with RSD, Subbarao and Stillwell (1981) reported 2 patients with unilateral RSD following neck injury, however they made no mention as to whether or not these patients had myelopathy. Wainapel (1984) reported 2 patients with incomplete cervical myelopathy and unilateral RSD. There was no mention of roentgenography or scintigraphy in these patients. Kozin *et al.* (1976) found that scintigraphy was more specific than roentgenography (86% vs 71%), whereas sensitivity of these tests was identical (68% vs 69%) in the

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diagnosis of RSD. We report a case of bilateral RSD in a patient with incomplete cervical myelopathy and clinical picture of central cord syndrome.

Case report

A 69-year-old female was admitted to the hospital with a 3-month history of progressive upper and lower extremity weakness and increasing difficulty with ambulation. Clinical diagnosis of cervical central cord syndrome secondary to cervical spondylosis was made. Diagnostic evaluation to confirm the clinical diagnosis included electromyography, myelography and computerised axial tomography of the cervical spine. One week after admission a decompressive laminectomy from C3 through C7 was carried out and 2 weeks later the patient was transferred to the rehabilitation facility for further management. She did not receive any physical therapy prior to the rehabilitation admission.

At the time of admission to the rehabilitation unit, neurological examination showed quadriparesis with significantly more weakness in the upper compared to the lower extremities. Muscle stretch reflexes were exaggerated in all the limbs, Babinski was positive bilaterally and ankle clonus was present on both sides. Sensory examination did not reveal any definite level, however, there was diminution of pain and two point discrimination in all extremities while lateralisation, vibration and position sense were intact.

About 4 months after the initial onset of symptoms and 4 weeks after surgery the patient began to complain of bilateral upper extremity pain and hyperesthesia that gradually worsened to the point of preventing participation in her rehabilitation programme and the use of her upper extremities for any functional activities. Neurological examination was unchanged. Inspection of the upper extremities revealed swelling, erythema and excessive sweating of both hands. Active and passive range of motion of the fingers and shoulders were limited and painful. The metacarpophalengeal joints were more tender than other joints of the fingers. The radial and ulnar pulses were normal bilaterally. X-rays of shoulders, wrists and hands as well as scintigraphy were done within 2 days of onset of bilateral upper extremity pain. The X-rays of the shoulders were normal while both wrists and hands showed moderate osteoporosis (Fig. 1). Scintigraphy demonstrated periarticular increased uptake in hands and wrists bilaterally with no other abnormal areas of tracer accumulation (Fig. 2). These findings were compatible with the diagnosis of RSD.

The treatment consisted of elevation of the forearm and hands, and gentle active and passive range of motion of all upper extremity joints. In addition she was started on 60 mgs of Prednisone a day in divided doses for 1 week and the dosage was tapered over the next 2 weeks. Within 5 days of starting treatment, the pain and hyperesthesia improved, the swelling of the hand decreased and there was increase in the range of movements of shoulders and fingers. She was able to participate in the rehabilitation programme. Seven weeks after admission to the rehabilitation facility she was discharged home. At that time she had significant return of upper extremity strength and function. She became independent in transfers and was walking short distances without assistive devices. One month after discharge she was seen in the outpatient clinic. There had been no recurrence of her symptoms and she was able to maintain her achieved functional status.

Discussion

The association of RSD with the central cord syndrome has been only rarely documented. Unlike other case reports of RSD following myelopathy our patient presented with a bilateral shoulder-hand syndrome. The clinical diagnosis of RSD was confirmed by roentgenography and scintigraphy. There was good therapeutic response to oral corticosteroids.

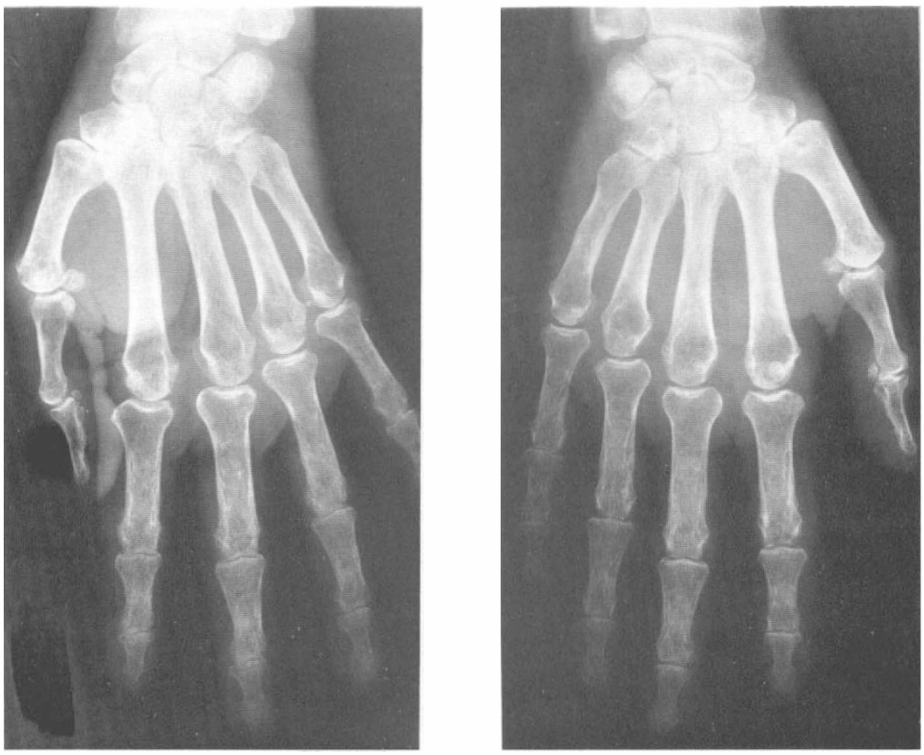

Figure 1 Roentgenograms of hands showing characteristic patchy demineralisation.

Since the description by Mitchell, Morehouse and Keen (1864), RSD has been reported in association with various clinical conditions, however, the pathogenesis remains poorly understood. It may be related to both peripherally and centrally mediated factors. Melzak and Wall (1965) in their gate control theory of pain transmission, postulated an inhibitory fine tuning mechanism in the substantia gelatinosa of the dorsal horn. They suggested that selective activation of large (A) fibres enhances the inhibitory tone in substantia gelatinosa, thereby 'closing the gate'. Predominant small (c) fibre stimulation could then result in the unchecked transmission of pain through an 'open gate'. Livingston (1943) suggested a vicious self-sustaining cycle where trauma produces chronic irritation of a peripheral nerve leading to increased afferent input and abnormal activity in the internucial neuronal pool leading to continuous stimulation of sympathetic and motor efferent fibres. Based on prior laboratory investigations, Doupe, Cullen and Chance (1944) postulated the formulation of artificial synapses between sensory afferents and sympathetic efferents leading to direct cross firing and cycle formation.

Physiological studies utilising oscillography, plethysmography, skin temperature measurements, venous gas determinations (Kozin *et al.*, 1976) and sweat test (Wainapel, 1984) have suggested increased blood flow or increased venous oxygen saturation in the affected extremity and the clinical manifestations of RSD may be due to local sympathetic nervous system overactivity. The scintigraphic studies probably reflect an increased vascular pool (McCarty *et al.*, 1970).

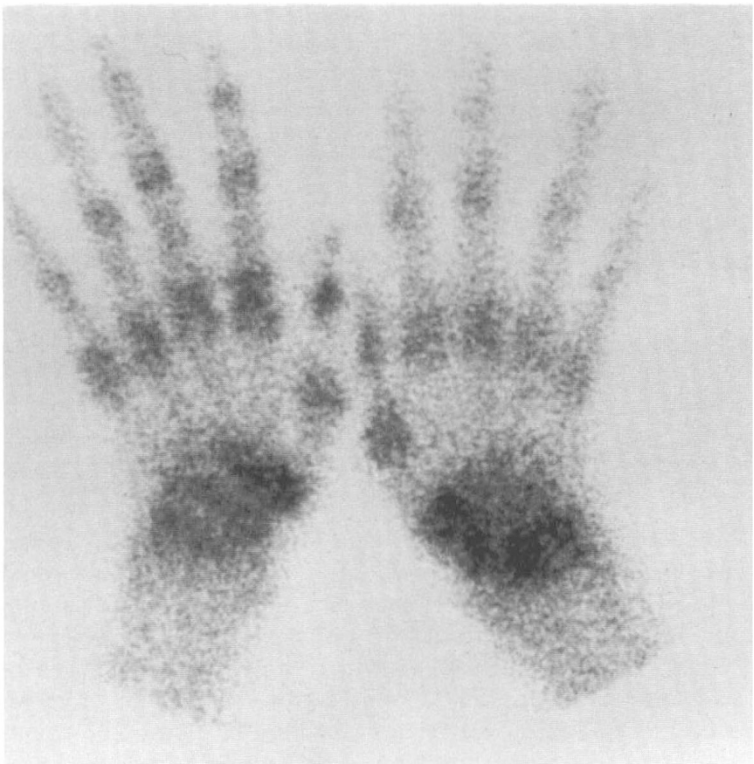

Figure 2 Scintigraphy using technetium 99m (^{99m}Tc) methylene diphosphonate (MDP) showing typical pattern of accentuation of uptake in wrists, MCP and IP joints.

Based on current experimental evidence, RSD may result from abnormal firing of peripheral nerves due to demyelination or sprout outgrowth of Schwann cells or axons (Devor, 1983). The injured or inflamed segment incorporates excessive numbers of sodium and calcium channels and alpha-adrenergic receptors. Thus ectopic pacemakers come into play and circulating catecholamines and those released from the sympathetic efferent sprouts activate these pacemakers and augment the discharge. This would result in spontaneous pain and hyperesthesia. The abnormal firing alters the responses by the neuronal pools in the spinal cord, which then responds abnormally to brain stem and cortex. These spinal and supraspinal mechanisms are likely responsible for the centralisation of pain in patients with RSD.

Steinbrocker (1968) in his classical description of RSD grouped the clinical syndrome into three phases, and one symptom may dominate the others (Table). The best clinical diagnostic indicator, as reported by Teperman *et al.* (1984), is metacarpophalangeal joint tenderness which presented a sensitivity of 85·7% and a specificity of 100%. It was also noted by them that scintigraphy is a useful diagnostic aid in patients suspected of having RSD.

Multiple treatment choices and modalities have been described. Several studies have stressed the importance of early treatment to prevent morbidity. Early range of motion exercises, splinting, contrast bath, paraffin bath, massage,

Table Clinical Evolution of Shoulder-Hand Syndrome in Reflex Neurovascular Syndrome of Upper Extremity *

Phase 1	Painful disability of the shoulder preceding, accompanying or following onset of pain, diffuse swelling, exquisite tenderness and vasomotor changes in the hand and digits of the same extremity; spotty demineralisation occurs in the bones of the hand and shoulder. There is bilateral involvement in 20% or more. This phase lasts from 3 to 6 months and sometimes may resolve spontaneously. TREAT AT ONCE.
Phase 2	Period of disappearance of pain and disability of the shoulder and swelling of the hand and fingers. Vasomotor changes and tenderness may remain in some patients. There is usually noticeable atrophy of the skin and muscles; sometimes in advancing cases thickening of the palmar fascia, which resembles Dupuytren's contracture, atrophy of the nails and impending contractures of the fingers occur. This phase lasts on the average from 3 to 6 months. Treatment is imperative during this phase to minimise residuals. Prognosis is poor.
Phase 3	Period of trophic changes with atrophy of skin of the hand, muscular weakness and atrophy, and limited articular function progressing to irreversible flexion deformities of the fingers and occasionally of the shoulder. Vasomotor changes and tenderness are generally absent in this phase. Prognosis is usually poor.

* From Steinbroker (1968).

transcutaneous electrical nerve stimulation (TENS) ultrasound over the stellate galglion and pneumatic compression were used. Kozin *et al.* (1976) reported 11 patients with RSD, diagnosis based upon clinical findings, synovial biopsy and scintigraphy. They reported improvement of symptoms with systemic corticosteroids. Ninety per cent of the patients with positive scintigraphy experienced a good or excellent response to oral steroids, however only 34% of patients with negative scintigraph reported any significant improvement. The response was not related to the duration of symptoms. These authors concluded that scintigraphy is useful both as a diagnostic aid and as a predictor of therapeutic response. Our patient had a positive scintigraph and responded well to oral steroids. The potent anti-inflammatory properties of corticosteroids may account for the therapeutic effect. Their destabilising effects on basement membranes, can reduce capillary permeability and therefore decrease the plasma extravasation that is commonly associated with the early stage of RSD (Kozin *et al.*, 1981).

Use of intravenous regional blocks of 1% lidocaine combined with Solu-Medrol (methyl prednisone sodium succinate) followed by immediate range of motion exercises has been described. Significant improvement was noted after several treatments (Poplawski *et al.*, 1983). The rationale is interruption of the abnormal reflex mediated by the autonomic nervous system. The blockade can be achieved by intravenous infusion of reserpine or guanethidine agents, regional sympathetic block or surgical sympathectomy. Usually several blocks are required. The current indication for surgical sympathectomy is a diagnosis of RSD in any patient who has obtained even partial relief from regional sympathetic blockade, but who had four such blocks without a permanent cure (Schutzer and Gossling, 1984). Hannington-Kiff (1977) described the technique of intravenous infusion of guanethidine which functions as a false transmitter, as being actively taken up by the sympathetic nerve endings and displacing norepinephrine from its storage sites. Benzon *et al.* (1980) reported successful intravenous block with reserpine which reduces storage vesicle re-uptake of catacholamines, thereby slowly depleting norepinephrine stores in sympathetic

nerve endings. Kleinert *et al.* (1973) reported that 80% of the patients who were resistant to physical therapy and medical management experienced pain relief from one or more stellate ganglion blocks. Eighty one per cent required no further treatment for 1 to 5 years of follow-up and 19% had only temporary relief and ultimately required surgical sympathectomy. Seventeen per cent of patients in this group showed no permanent improvement. Lankford and Thompson (1977) reported that 89% of their patients with causalgia received long term palliation with sympathetic blocks.

Examination of several studies suggests that patients with early RSD respond well to oral corticosteroids (76–90%), but later treatment may be required. Similar response (81–89%) has also been described with serial paravertebral ganglion blocks. When there are multiple recurrences, paravertebral ganglionectomy should be considered. Regional blocks with lidocane and corticosteriods or with reserpine or guanethidine can be effective when symptoms recur after sympathectomy. The early recognition and treatment are the most important factors in the effective management of RSD as patients with longstanding disease are less likely to recover. Early mobilisation facilitates recovery and reduces physical disability.

Reflex sympathetic dystrophy in myelopathy is not readily disagnosed or does not become apparent due to both early administration of steroids, which have a protective effect, as well as early mobilisation of the upper extremities (Wainapel, 1984). RSD must be entertained as a differential diagnosis in patients with incomplete quadriplegia presenting with unexplained painful syndromes of the extremities. In incomplete cervical myelopathy the combination of hand pain and shoulder as presenting symptoms of RSD is helpful in making the diagnosis. In some reported cases of RSD there was no shoulder joint symptoms and the findings were exclusively in the hands. Shoulder pain in patients with complete C5 quadriplegia is common. Often the cause of pain is attributed to forceful range of movements of the shoulder or to possible nerve root irritation due to the cervical injury. RSD must be considered as a differential diagnosis in patients with C5 complete quadriplegia and shoulder pain. In the absence of shoulder pain in C5 complete quadriplegia other findings as hand edema, vasomotor instability and reduced range of movements of fingers should alert the clinician to further evaluate for the possibility of RSD.

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