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Complex regional pain syndrome as a cause of acute painful limb swelling – a case report

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Abstract

Complex regional pain syndrome (CRPS) is a complex condition characterised by severe pain and various sensory and motor abnormalities. This case report is of an 18-year-old woman presenting with painful upper limb swelling for a few days, with skin mottling and dysesthesia, with significant disability. With negative investigations, a diagnosis of CRPS was made. The case emphasises the importance of early recognition and appropriate management of CRPS, as delayed diagnosis leads to poor quality of life and functional impairment. Healthcare professionals should maintain a high index of suspicion for CRPS in patients with unexplained limb pain and associated sensory and motor abnormalities, ensuring a comprehensive multidisciplinary approach to optimise patients' quality of life.

Key words: complex regional pain syndrome, CRPS, reflex sympathetic dystrophy, limb pain, neuralgia

Introduction

Complex regional pain syndrome (CRPS) is a condition characterised by persistent pain disproportionate to tissue injury, accompanied by various sensory, motor, and autonomic disturbances. (1) CRPS can be of two types with similar clinical features (I and II), but only type II is associated with an identified nerve injury. The condition affects all ages but is most prevalent in women of 60 to 70 years of age.(2) Several factors, including gender, race, income level, and comorbidities, are associated with CRPS.(2) The exact pathogenic mechanisms of CRPS remain complex and multifactorial. The condition is linked to worsened mental health, functional impairment, disability, and reduced quality of life.

This case report highlights a young girl's delayed diagnosis of CRPS, underscoring the significance of early recognition and management. This aims to increase the awareness about CRPS, consider it as a potential diagnosis in patients presenting with unexplained pain and associated sensory and motor abnormalities, and promote timely identification and appropriate treatment.

Case presentation

An 18-year-old, right hand-dominant woman presented with a three-day history of spontaneous onset swelling, burning pain in her right forearm and hand. The pain, rated as 10/10 in intensity, was diurnal, persistent, accompanied by allodynia, and worsened with mild tactile stimuli. She noticed cooler fingers with reddish mottling on the affected side. She attributed these to increased writing activities. Sleep was affected. She avoided the pain by allowing the affected limb to hang down without touching anything around. There was no fever, joint pain, weakness, tremors, or change in handwriting. She

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found it difficult to carry out most of the daily work and was concerned whether it will affect her studies. Oral acetaminophen and topical analgesics provided no relief.

Three months prior, she experienced cramping sensations in her right forearm, while writing and object manipulation. Despite seeking help from medical practitioners, no definitive diagnosis was made. The patient recalled a previous history of finger trauma at 14 years and forearm swelling at 16, both of which had resolved with analgesics. Her family history was unremarkable, and she had no atopy or allergy in the past.

On examination, the patient's vital signs and body mass index were normal. Anterior and posterior compartments of the right upper limb were swollen (figure 1), cooler, and had faint dusky mottling of skin (figure 2). Wounds, dystonic posturing, superficial infections, dyshidrosis, thrombophlebitis, or active synovitis were not observed. Arterial pulses were felt, with no blood pressure discrepancy. Pulse oximetry of all five fingers gave an on-air saturation reading of 98%. Infrared thermometry showed the affected limb's surface temperature to be lower (34^{0}C) than the contralateral side (37^{0}C). Allodynia, hyperesthesia, and restricted movements were present, but the rest of the motor, sensory, and cerebellar functions were normal. Systemic examinations were unremarkable.

Electrocardiogram, nerve conduction, and electromyographic studies, doppler studies of arterial and venous systems of the right upper limb, plain radiographs of the chest, cervical spine, and right upper limb (figure 3), and contrast-magnetic resonance imaging of the cervical cord, brachial plexus, and upper limb compartments were normal. Ultrasonography of the forearm confirmed mild soft tissue swelling (figure 4). The summary of the blood investigations is shown in table 1.

A final diagnosis of CRPS was established, as she fulfilled all four Budapest diagnostic criteria.(3) She had a continuous pain that was disproportionate to the inciting event, had no alternative explainable aetiologies, and displayed signs and symptoms of all four clinical categories – sensory (allodynia,

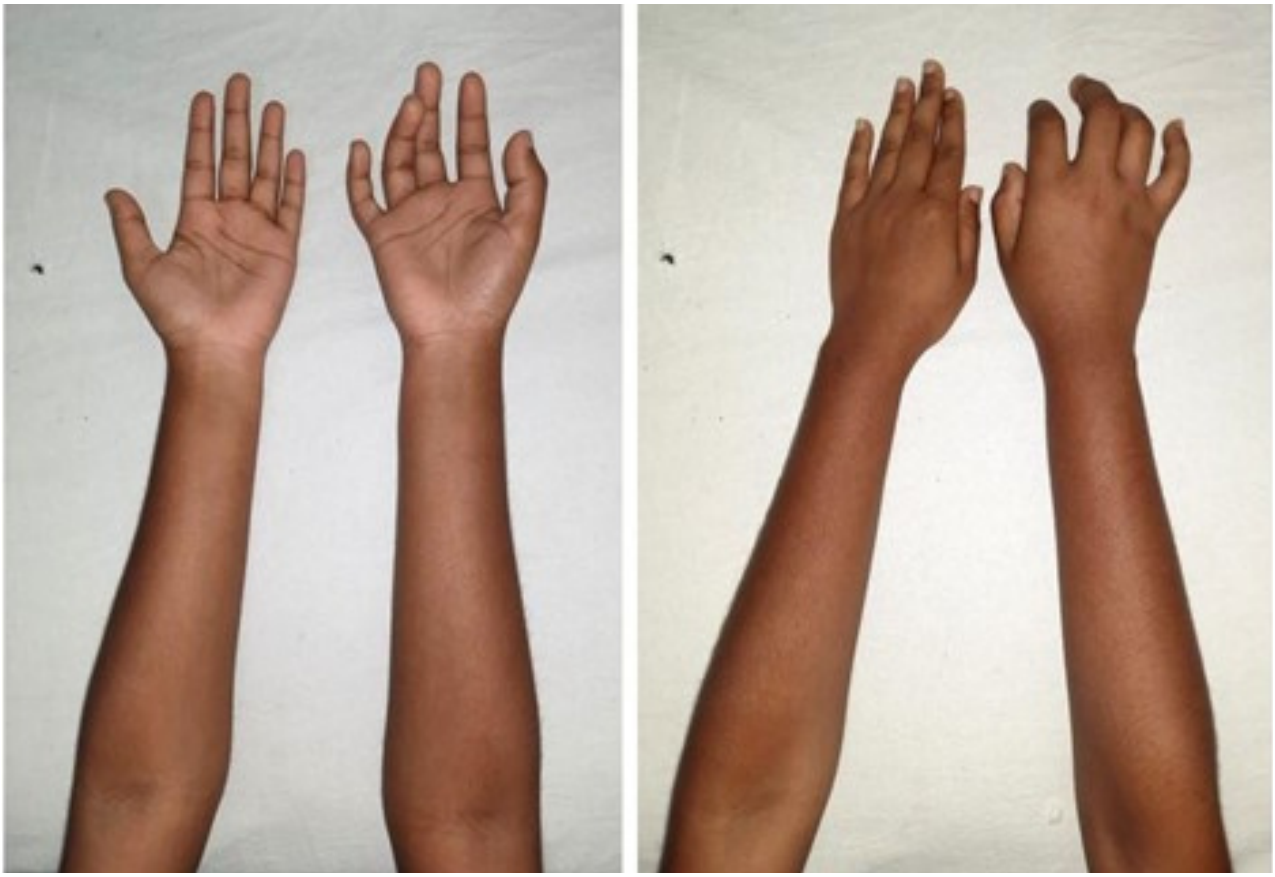

Figure 1 -Dusky-coloured swelling of the right hand and forearm, with limitation of finger extension

hyperalgaesia), vasomotor (temperature asynchrony, cutaneous colour change), sudomotor, oedema and motor (reduction in range of movement).

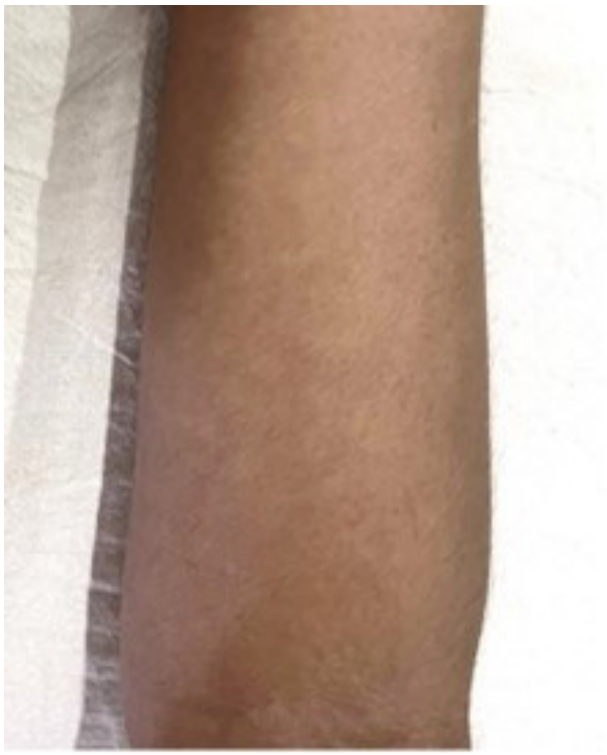

Figure 2 - Mottling of surface skin

The patient was educated about the condition. A combination of oral analgesics, including diclofenac sodium (50 mg twice daily) and gabapentin (100 mg twice daily) was given. Prednisolone was initiated at 25 mg with a tapering regimen over a few weeks based on evidence in a recent systematic review.(4) Calcium, vitamin D and famotidine were also prescribed. Outpatient physiotherapy was arranged. Upon discharge, the patient's pain score remained at 6/10. At a five-day follow-up, the patient's pain score remained unchanged, following which a pain specialist prescribed Panadeine, and low-dose amitriptyline, resulting in mild symptom relief after one month The patient declined the option of cervical sympathectomy. About 6 months later she was experiencing less pain, with a pain score of 2/10, and was managing her daily activities with low dose amitriptyline.

Discussion

Complex regional pain syndrome (CRPS) is often misdiagnosed or overlooked, presenting as a challenge in clinical practice. The lack of specific gold standard investigations for CRPS contributes to the diagnostic difficulty. However, the rarity of CRPS and its overlap with other conditions delay diagnosis. Timely recognition and consideration of CRPS as a potential diagnosis are vital to prevent poor quality of life and functional impairments in affected

Table 1 - Summary of investigations

Investigation	Patient's Value	Reference range
Total white blood cell count (x 10^3/uL)	12.75	4 – 10
Neutrophils percentage	60	50 – 70
Lymphocytes percentage	38	20 – 40
Haemoglobin (g/dL)	13.2	11 – 16
Platelet count (x 10^3/uL)	296	150 – 450
C –reactive protein (g/L)	2.3	<5
Erythrocyte sedimentation rate (mm/1st hour)	14	< 15
Serum creatinine (umol/L)	39	59 – 104
Creatine phosphokinase (U/L)	40.9	30-135
d-dimer	negative	

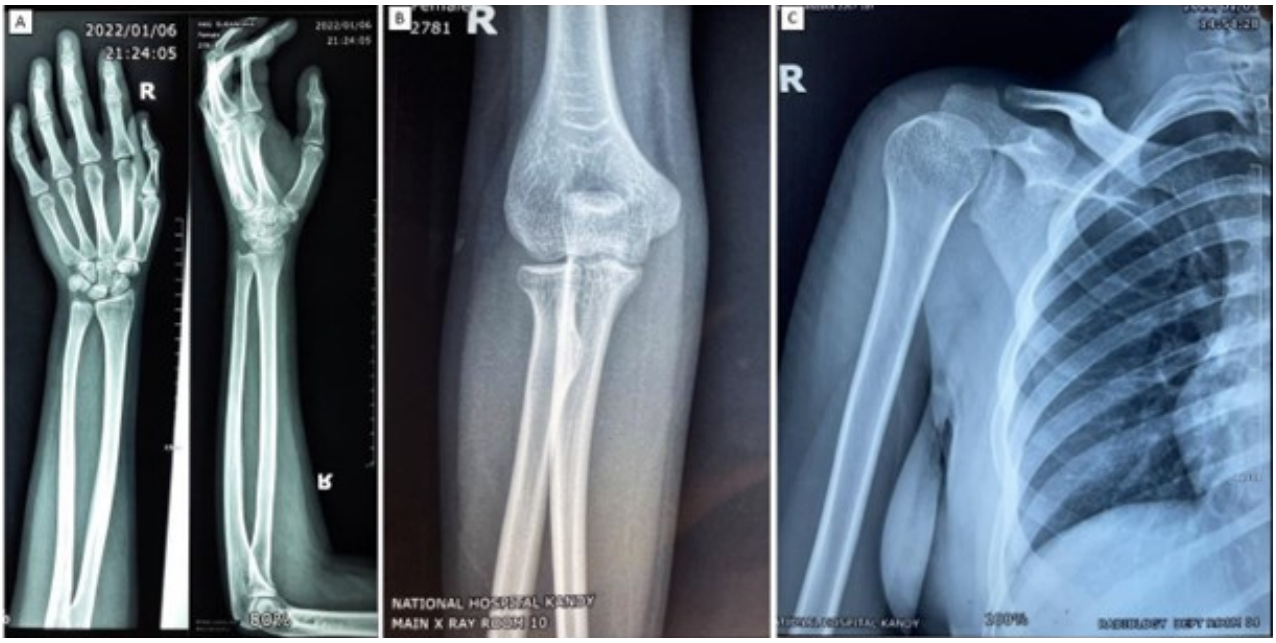

Figure 3 -Plain radiographs of the right forearm (A), elbow (B), and right shoulder (C) show no significant abnormalities

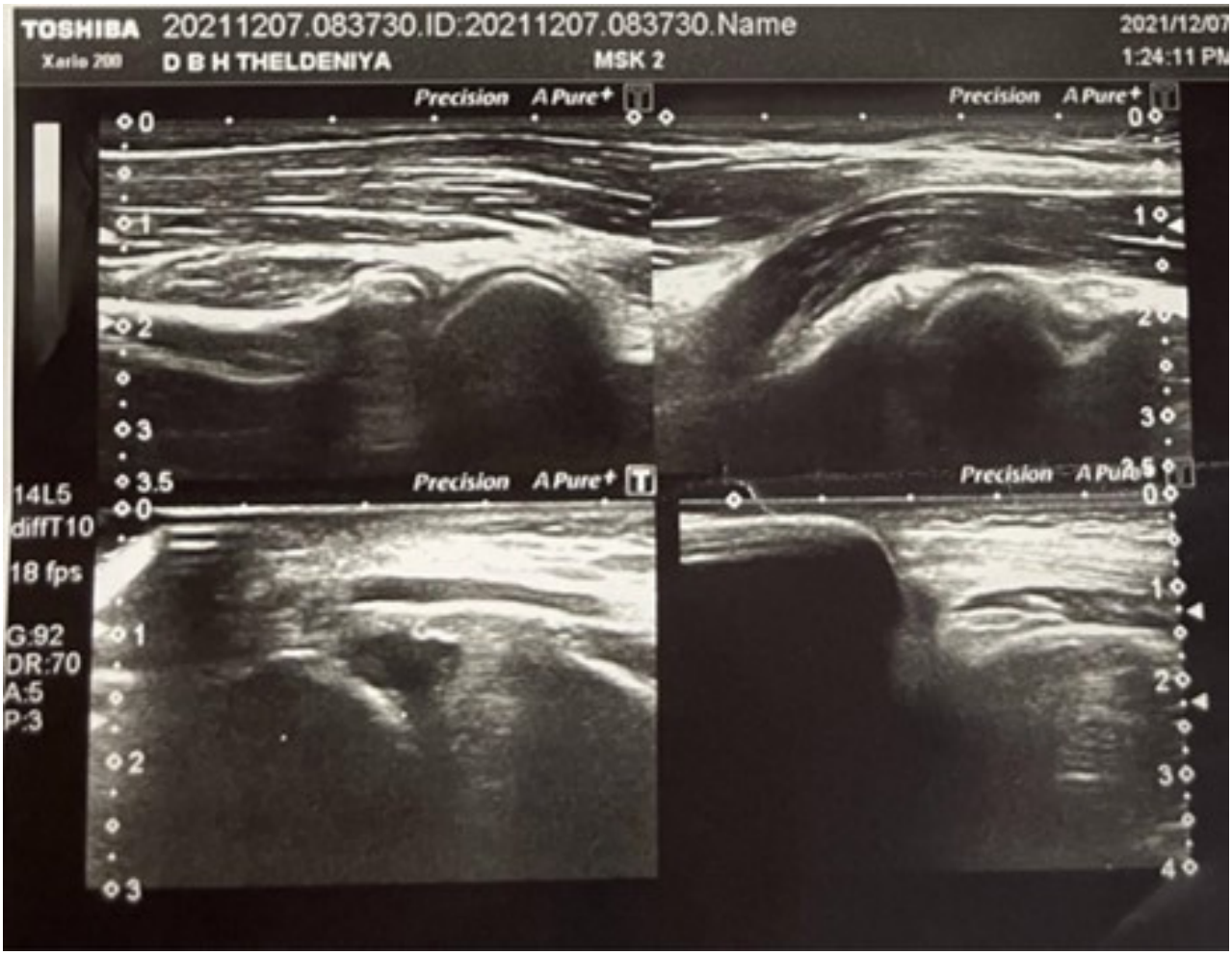

Figure 4 - Normal ultrasound of elbow and shoulder

individuals.(5) Differential diagnoses include sensorimotor neuropathy, arthropathy, peripheral arteriopathy, vasculitis, deep vein thrombosis, lymphoedema, Raynaud's phenomenon, scleroderma, and Dupuytren's contracture, making accurate identification challenging.(6)

The patient in this case exhibited severe diurnal pain, swelling, sensory abnormalities, allodynia, changes in skin temperature, mottling, and reduced range of motion in her right forearm and hand. A history of finger trauma, school-related stress, and forearm swelling raised suspicion for CRPS. The Budapest diagnostic criteria, which carries a sensitivity of 95% and specificity of 81%, was met in our patient, confirming the diagnosis.(3) In this case, imaging and electrophysiological studies yielded unremarkable results, emphasising the importance of clinical acumen in considering CRPS despite initial negative findings. Although bone scintigraphy supports the diagnosis of CRPS, it was not available in our setting.

The treatment of CRPS is challenging and lacks clear evidence for primary therapeutic choices. An interprofessional approach is recommended to improve symptoms, restore function, and to prevent disability, muscle atrophy, and demineralization. Management includes:

- Education about the condition
- Physical methods (manual therapy, bandages, and compression sleeves, exercises, transcutaneous electrical nerve stimulation, warm or cold application, contrast bath therapy, hydrotherapy methods, mirror therapy, graded motor imagery (GMI), occupational therapy, behavioural therapy, multimodal pharmacotherapy (anti-inflammatory

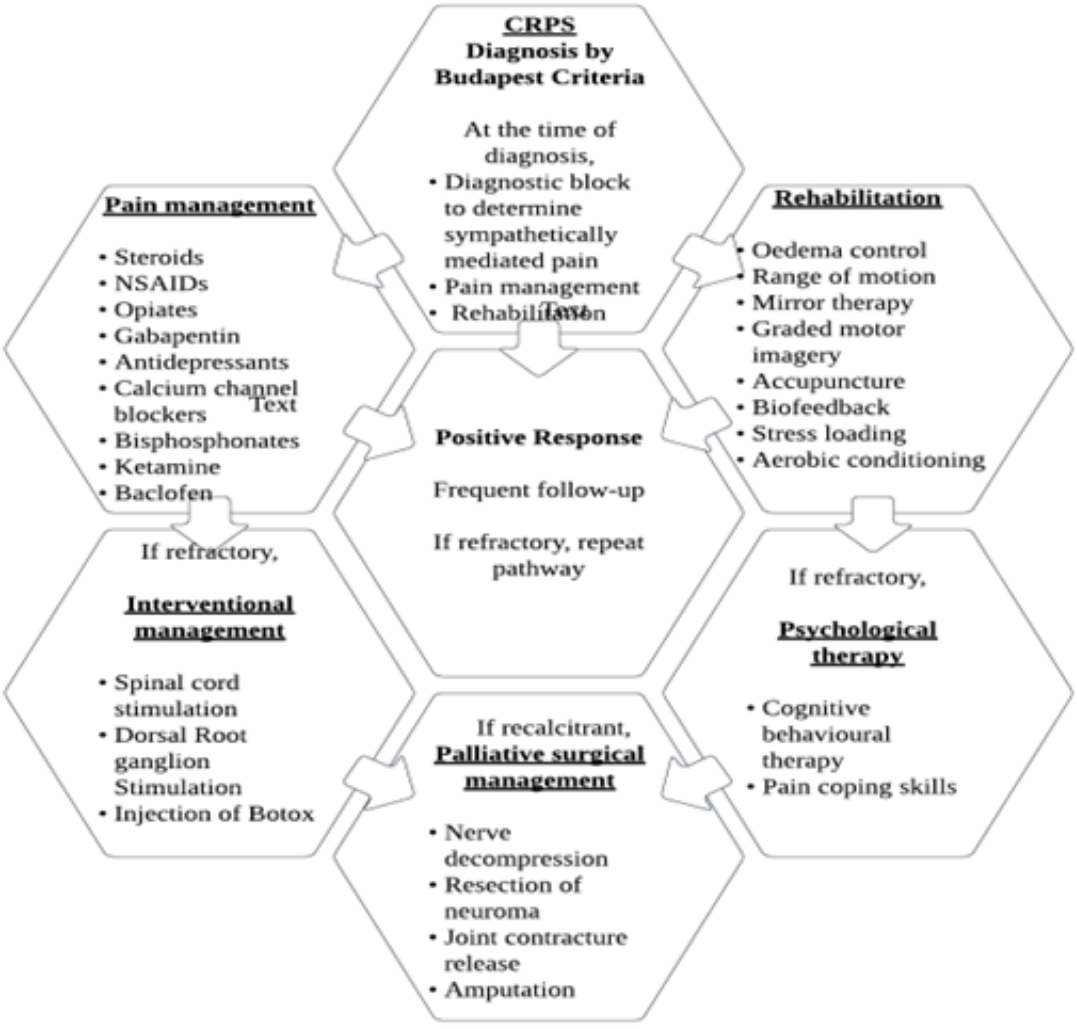

Figure 5 - Flow diagram of different treatment modalities in CRPS(7)

medications, anticonvulsants, antidepressants, transdermal lidocaine, opioids, NMDA antagonists, calcitonin, ketamine, clonidine, Vitamin C and bisphosphonates) and
- Interventional therapies (sympathetic blocks, dorsal root ganglion stimulation, spinal cord stimulation)

in resistant cases.(7)

A flow diagram of different treatment modalities in CRPS is given in figure 5.(8) Among the different physiotherapy modalities, mirror therapy and GMI may improve pain as well as function.(9) A multimodal pharmacologic regimen that combines several different classes may lead to superior outcome. A recent systematic review states that continuation of a short course of low-dose prednisone seems to be safe and effective in managing CRPS.(4) Steroids are more effective compared to NSAIDs.(10) Amitriptyline and gabapentin reduce pain intensity and disability, to a similar degree.(11) The mechanisms of bisphosphonates on CRPS include inhibition of bone marrow cell proliferation and migration, and inflammation modulation. As CRPS may progress to late-stage atrophy and contracture, a palliative approach may be considered, tailored to the patient's needs, including surgical correction of contractures or deformities. Relapses are generally common, with the median time for relapses being about two months, and about 80% of the relapses occur within six months of treatment completion.(12) Despite analgesics, our patient's pain remained minimally resolved, highlighting the complexity of managing CRPS when standard treatments prove insufficient.

This case report underscores the significant impact of CRPS on the patient's daily life and overall well-being, particularly in the context of academic struggles and disrupted activities due to pain. Healthcare professionals should maintain a high suspicion for CRPS in patients presenting with acute or chronic pain, sensory abnormalities, and motor dysfunction, especially after a history of trauma.

Conclusion

This case report sheds light on the diagnostic challenges of CRPS and emphasises the significance of early recognition and comprehensive management. It provides valuable insights for healthcare professionals on the complexities of diagnosing and treating CRPS. Increasing awareness and understanding of the condition among healthcare providers is crucial for timely diagnosis, timely specialist referrals, and multidisciplinary treatment approaches, effective treatment, and better outcomes, ultimately improving the quality of life for patients with CRPS.

Declarations

Author contributions
All authors were involved in the management of this patient

Conflicts of interest
No conflict of interest

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