Rheumatology Quiz

Tippawan Onchan

Department of Medicine, Faculty of Medicine, Khon Kaen University, Khon Kaen, Thailand.

A 37-year-old healthy female with a history of surgically repaired right index finger fracture 2 years ago presented with a 17-month history of right forearm pain. She reported recurrent episodes of redness, swelling, warmth, and burning pain that began in her right index finger and gradually progressed to involve the right hand, forearm and elbow. Symptoms worsened with heat exposure and improved with cooling. Examination revealed erythema, mild edema, moist skin, and warmth over the right fingers and forearm (Figure 1). There was tenderness at the proximal interphalangeal joints and metacarpophalangeal joints with a flexion deformity of the index finger. The pain was disproportionate to light palpation and did not improve after a complete course of antibiotics. The physical findings and hand radiographs are shown in Figures 1 and 2, respectively.

Questions

What is the most likely diagnosis?

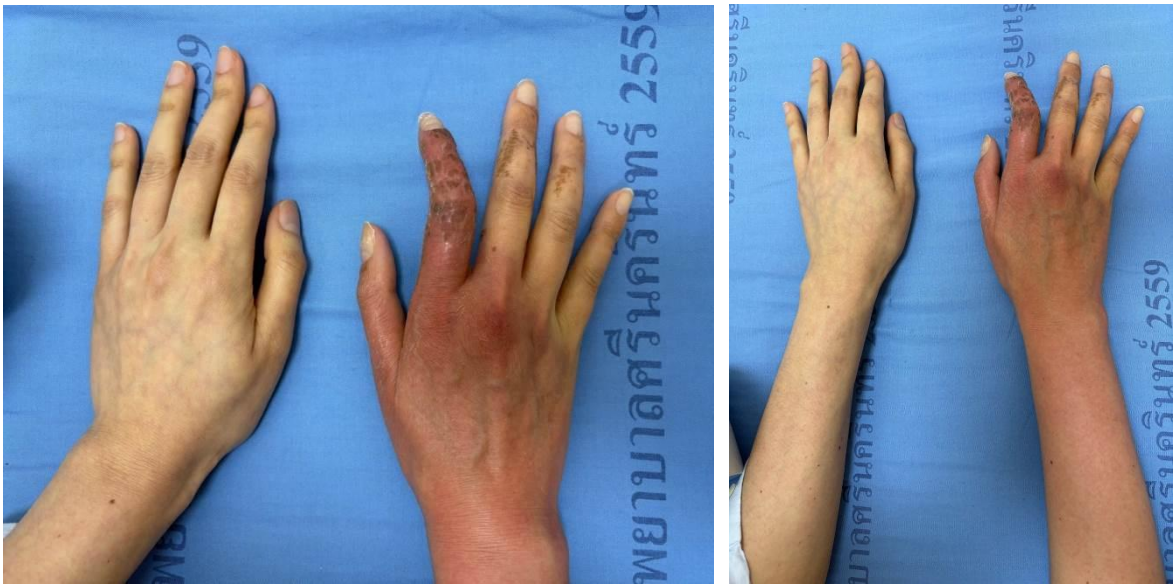

Figure 1 Physical findings of the hands and forearms.

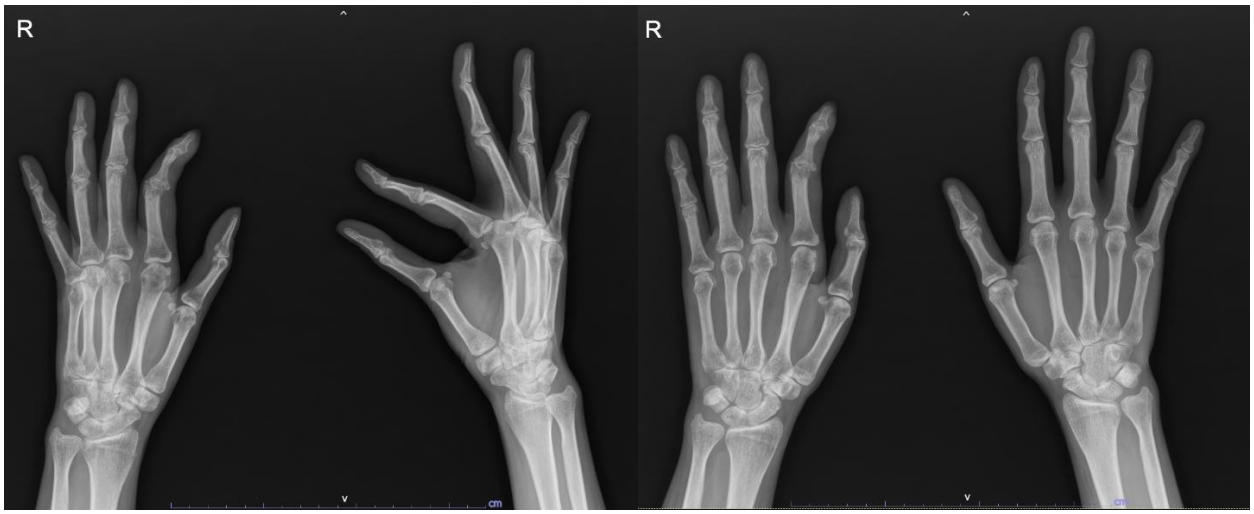

Figure 2 Hand radiographs.

Answers

1. **Diagnosis:** Complex Regional Pain Syndrome of right forearm

This patient had a history of trauma to the right index finger, which preceded the development of progressive, disproportionate pain and vasomotor symptoms. Plain radiography revealed post-traumatic subluxation at the proximal interphalangeal joint of the right index finger with osteoarthritic changes. She was diagnosed with complex regional pain syndrome of the right forearm and was treated with nonsteroidal anti-inflammatory drugs and short course corticosteroids, and supportive medication such as anticonvulsants and opioids.

Short Review

Complex Regional Pain Syndrome (CRPS), formerly known as *reflex sympathetic dystrophy or causalgia*, is a rare, chronic neuropathic pain disorder with an incidence of 6.3-26.2 per 100,000 person-years. It occurs more frequently in women than men (3:1). The exact etiology remains unclear, but its pathophysiology is multifactorial, involving inflammation, immunologic responses, central and peripheral sensitization, and autonomic dysregulation. CRPS typically develops after fracture, trauma or surgery and persists beyond normal healing. It is characterized by pain, allodynia or hyperalgesia disproportionate to the inciting event, along with edema, temperature or skin color asymmetry, abnormal sweating, and trophic changes of nails or hair-most often affecting upper extremities or distal limbs. Symptoms typically begin within 4-6 weeks after the inciting event, and 80% of cases develop within 12 weeks.

CRPS is classified into two types: Type I (without nerve injury, more common), and Type II (with known nerve injury)[1]. Conditions that may mimic CRPS include peripheral arterial disease, deep vein thrombosis, Guillain-Barre syndrome, and multiple sclerosis. Diagnosis is clinical and based on the Budapest criteria (also known as ISAP criteria)[11], which require exclusion of other causes:

I. Continuing pain, disproportionate to the inciting event.
II. Patient reports at least one symptom in all four categories
    1. Sensory: allodynia, hyperalgesia
    2. Vasomotor: temperature asymmetry, skin color changes/ asymmetry
    3. Sudomotor: change in or asymmetrical edema/ sweating
    4. Trophic: decrease range of motion, weakness, tremor, dystonia, changes of hair, nail, skin
III. Physician observes at least one sign in at least two categories
    1. Sensory: allodynia (to light touch, temperature, pressure, or vibration), hyperalgesia (to pinprick)
    2. Vasomotor: temperature asymmetry of >1-degree Celsius, skin color changes/ asymmetry
    3. Sudomotor: change in or asymmetrical edema/ sweating
    4. Trophic: decrease range of motion, weakness, tremor, dystonia, changes of hair, nail, skin
IV. No other diagnosis explains the symptoms and signs

CRPS generally progresses through three stages[2]:

1. **Acute stage** (6-12 months): pain, swelling, color changes, painful movement, and early osteoporosis on X-ray.
2. **Dystrophic stage** (1-2 years): persistent pain, hard edema, muscle atrophy, cooler skin, and progressive osteoporosis.
3. **Atrophic stage** (chronic): persisting or diminishing pain, stiffness, smooth and shiny skin, advanced osteoporosis or bony ankylosis.

Since there is no single standard diagnostic test, plain radiography, magnetic resonance imaging (MRI) can be useful for excluding musculoskeletal disorders, particularly osteonecrosis. Bone scintigraphy is more specific for CRPS than MRI and helps in diagnosis. Doppler flow studies assist in excluding venous thrombosis. Infrared thermometry and thermography typically show higher temperatures in the affected limb compared to the contralateral side.

Management of CRPS is multidisciplinary and includes pharmacologic, physical, psychological and interventional approaches. Pharmacotherapy options include nonsteroidal anti-inflammatory drugs, steroids, gabapentin, bisphosphonate, and ketamine.[1,3] Additional agents such as botulinum toxin A, vitamin C, and immunotherapy (e.g. interleukin-1 receptor antagonists, intravenous immunoglobulin, and Tumor necrosis factor-α inhibitors)[4] may be considered. Physical therapy focuses on multimodal physiotherapy, aerobic exercise, and transcutaneous electrical nerve stimulation.[5] Psychological support is essential for coping and functional recovery. Invasive options, including sympathetic blocks, spinal cord or dorsal root ganglion stimulation, and amputation, may be considered for refractory cases.

CRPS complications may involve cognitive executive dysfunction, neurocardiogenic syncope, chest wall muscle dystonia, adrenal insufficiency, increase urinary frequency, urgency, or incontinence, gastroparesis, irritable bowel syndrome, lethargy, weakness and sleep disturbance.[6–9] The prognosis of CRPS is variable. Early diagnosis and treatment may improve outcomes.[10]

To cite: Onchan T.
Thai J Rheum. 2025;2(4):72-76. Available from: https://he04.tci-thaijo.org/index.php/tjr

Reference

1. Abd-Elsayed A, Stark CW, Topoluk N, Isaamullah M, Uzodinma P, Viswanath O, et al. A brief review of complex regional pain syndrome and current management. Ann Med. 56(1):2334398.

2. Aboul-Fettouh A, Goree J. Complex Regional Pain Syndrome. In: Banik RK, editor. Anesthesiology In-Training Exam Review: Regional Anesthesia and Chronic Pain [Internet]. Cham: Springer International Publishing; 2022 [cited 2025 Nov 10]. p. 267–71. Available from: https://doi.org/10.1007/978-3-030-87266-3_51

3. Doan HN, Chang MC. Comprehensive Mechanisms and Non-Invasive Treatment for Complex Regional Pain Syndrome: A Narrative Review. J Pain Res. 2025 Sept 11;18:4699–726.

4. Kaye AD, Nguyen A, Boullion J, Blue MEF, Allen DWH, Kelkar RA, et al. Efficacy of Immunotherapy for Complex Regional Pain Syndrome: A Narrative Review. Curr Pain Headache Rep. 2025 Jan 4;29(1):4.

5. Smart KM, Ferraro MC, Wand BM, O'Connell NE. Physiotherapy for pain and disability in adults with complex regional pain syndrome (CRPS) types I and II. Cochrane Database Syst Rev. 2022 May 17;2022(5):CD010853.

6. Libon DJ, Schwartzman RJ, Eppig J, Wambach D, Brahin E, Peterlin BL, et al. Neuropsychological deficits associated with Complex Regional Pain Syndrome. J Int Neuropsychol Soc JINS. 2010 May;16(3):566–73.

7. Park JY, Ahn RS. Hypothalamic-pituitary-adrenal axis function in patients with complex regional pain syndrome type 1. Psychoneuroendocrinology. 2012 Sept;37(9):1557–68.

8. Irwin DJ, Schwartzman RJ. Complex regional pain syndrome with associated chest wall dystonia: a case report. J Brachial Plex Peripher Nerve Inj. 2011 Sept 26;6:6.

9. Chancellor MB, Shenot PJ, Rivas DA, Mandel S, Schwartzman RJ. Urological symptomatology in patients with reflex sympathetic dystrophy. J Urol. 1996 Feb;155(2):634–7.

10. Guthmiller KB, Dua A, Dey S, Varacallo MA. Complex Regional Pain Syndrome. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 [cited 2025 Nov 1]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK430719/

11. Ferraro MC, O'Connell NE, Sommer C, Goebel A, Bultitude JH, Cashin AG, et al. Complex regional pain syndrome: advances in epidemiology, pathophysiology, diagnosis, and treatment. Lancet Neurol. 2024 May;23(5):522–33.