

# Refractory complex regional pain syndrome resulting in amputation with dermatosis neglecta: A case report

## Dermatozis neglekt'a'nın eşlik ettiđi ve amputasyon ile sonuçlanan dirençli kompleks bölgesel ağrı sendromu: Olgu sunumu

Hatice Ceylan , Kubra Unlu , Gizem Kiliñç Kamaci , Nurdan Korkmaz , Yasin Demir , Koray Aydemir 

Department of Physical Medicine and Rehabilitation, Health Sciences University, Ankara Gaziler Physical Therapy and Rehabilitation Training and Research Hospital, Ankara, Türkiye

### ABSTRACT

Complex regional pain syndrome (CRPS) is a painful condition which may develop after trauma. A 28-year-old male patient with severe hand pain that persisted despite long-term and multidisciplinary treatment was admitted to our clinic. The patient had severe pain and cutaneous findings. His extreme cutaneous findings were diagnosed as dermatosis neglecta (DN) associated with CRPS. To the best of our knowledge, this is the first reported case of DN occurring in association with CRPS. This case report highlights that extreme skin changes may occur in the advanced stages of complex regional pain syndrome, and when this stage is reached, treatment becomes challenging and can be controlled with irreversible treatments such as amputation.

**Keywords:** Amputation, analgesia, complex regional pain syndrome, dermatosis neglecta, hyperkeratosis, neglect-like symptoms, nerve injury.

Complex regional pain syndrome (CRPS) is a pain condition characterized by allodynia, hyperalgesia, sudomotor changes such as excessive sweating or dryness, vasomotor abnormalities, and trophic skin and nail changes. Thermography and bone mineral densitometry can support the diagnosis. Complex regional pain syndrome typically develops following a physical injury and is thought to have a multifactorial pathophysiology involving neurogenic inflammation, immunological mechanisms, and structural changes in the sympathetic and central nervous systems. Diagnosis is primarily clinical and is supported by radiological imaging, including conventional radiography, computed tomography, and magnetic

### ÖZ

Kompleks bölgesel ağrı sendromu (KBAS), travma sonrası gelişebilen ağrılı bir durumdur. Uzun süreli ve multidisipliner tedaviye rağmen devam eden şiddetli el ağrısı bulunan 28 yaşındaki erkek hasta kliniğimize başvurdu. Hastada şiddetli ağrı ve deri bulguları mevcuttu. Hastanın ileri derecede deri bulguları KBAS ile ilişkili dermatosis neglecta (DN) olarak tanılandı. Bilgimiz dahilinde bu, KBAS ile ilişkili DN'nin bildirildiđi ilk olgudur. Bu olgu sunumu, kompleks bölgesel ağrı sendromunun ileri evrelerinde belirgin cilt değışikliklerinin gelişebileceđini ve bu aşamaya ulaşıldığında tedavinin güçleşebileceđini, bazı durumlarda amputasyon gibi geri dönüşü olmayan tedavi seçeneklerinin gündeme gelebileceđini göstermektedir.

**Anahtar sözcükler:** Amputasyon, analjezi, kompleks bölgesel ağrı sendromu, dermatosis neglecta, hiperkeratoz, ihmal benzeri semptomlar, sinir yaralanması.

resonance imaging, as well as three-phase radionuclide bone scintigraphy.<sup>[1]</sup>

An integrated, multidisciplinary approach is recommended for CRPS treatment, encompassing patient education, pharmacological and interventional pain relief, physical rehabilitation, and psychological care. Initial treatments may include physical therapy modalities such as exercise, transcutaneous electrical nerve stimulation, and contrast baths, alongside pharmacological agents including nonsteroidal anti-inflammatory drugs, steroids, pregabalin, gabapentin, amitriptyline, and duloxetine.<sup>[2]</sup> Non-invasive options such as neuromodulation and psychological therapies should also be considered.

**Corresponding author:** Hatice Ceylan.

**E-mail:** drhaticeceylan@gmail.com

**Doi:** 10.5606/agri.2026.12

**Received:** May 13, 2025 | **Accepted:** May 29, 2025 | **Published online:** September 25, 2026

**Cite this article as:** Ceylan H, Unlu K, Kiliñç Kamaci G, Korkmaz N, Demir Y, Aydemir K. Refractory complex regional pain syndrome resulting in amputation with dermatosis neglecta: A case report. Agri 2026;38(4):292-297. doi: 10.5606/agri.2026.12.

For refractory cases, more invasive treatments, including amputation, may be necessary.<sup>[2-4]</sup>

Dermatosis neglecta (DN), a disorder of keratinization, is characterized by hyperpigmented, verrucous plaques and scales resulting from inadequate cleansing due to hyperesthesia, trauma, or inactivity. Lesions arise from insufficient cutaneous cleansing and exfoliation due to neglect or disability.<sup>[5]</sup> We report that hyperkeratinization, known as DN, may develop as a consequence of limb neglect in CRPS.

In this article, we describe a case of CRPS with DN who underwent amputation after six years of refractory allodynia following a peripheral nerve lesion in the right upper extremity.

## CASE REPORT

A 28-year-old male patient with a six-year history of right upper extremity CRPS secondary to a combat-related shrapnel injury was admitted to our hospital with pain and hyperkeratosis in the right hand and wrist.

### Medical history from 2018 to 2021

In September 2018, his right upper extremity, right lower extremity, and thorax were injured by an improvised explosive device. Electromyographic studies in November 2018 and March 2019 confirmed injuries to the right median, ulnar, and radial nerves. Shortly after the trauma, he developed

pain, burning sensations, hyperesthesia, skin color changes, and allodynia in the right hand. The skin of the hand was edematous and hyperemic. Complex regional pain syndrome was diagnosed according to the Budapest criteria.<sup>[6]</sup> The diagnosis was confirmed by bone scintigraphy.

Complex regional pain syndrome treatment was performed according to guideline recommendations. Medical therapy began in 2018 with non-steroidal anti-inflammatory drugs, systemic steroids, and vitamins B and C. Despite multiple oral agents, the patient showed no improvement. Pregabalin was initiated and titrated to 150 mg twice daily, and duloxetine was prescribed. In 2018, the patient started range of motion (ROM) exercises, physical therapy such as transcutaneous electrical nerve stimulation, psychotherapy, fluidotherapy, and contrast baths. He was also prescribed sertraline and olanzapine for post-traumatic stress disorder and sleep disturbances. However, the patient refused physical therapy due to intolerable pain. Rehabilitation included sensory reeducation with a desensitization kit and intermittent ultrasound treatment to the stellate ganglion. Stellate ganglion blockade was offered in 2019 and 2021, but the patient declined. Transcranial magnetic stimulation was recommended in 2021 but was discontinued due to lack of pain relief. The patient had no hospital admission for three years during the novel coronavirus disease 2019 (COVID-19) pandemic.



**Figure 1.** (a) Dermatitis neglecta in refractory complex regional pain syndrome. He had verrucous plaques on his right hand, hyperkeratized and hyperpigmented in appearance. (b) A photograph of the patient taken in the first days of hospitalization. (c) Two months after hospitalization, some of the lesions on the patient's hand have been removed. The patient reported greater sensitivity in non-lesional areas.

### Medical history from 2024 to 2025

The patient was admitted to the hospital again in January 2024. He had pain and verrucous plaques on his right hand, hyperkeratized and hyperpigmented in appearance (Figure 1). He was diagnosed with DN in 2024. Radiographs revealed shrapnel fragments and osteoporotic changes, as shown in Figure 2. The patient rated his pain as 10 on the Visual Analog Scale (VAS). He used pregabalin 300 mg twice daily for five years. Stellate ganglion blocks with steroids and local anesthetics were performed in January 2024 but failed to alleviate symptoms. Duloxetine was increased to 120 mg daily, and a spinal cord stimulator was implanted in February 2024, but

neither provided relief. Oxycodone was initiated and increased to 5 mg four times daily, yet the pain persisted. Table 1 summarizes the patient's clinical characteristics and treatment course.

Functional examination revealed no grasping or holding movements in the hand. The patient could not use the affected limb due to pain. Active movement in the right finger and wrist joints was absent, and ROM measurements were impossible due to severe pain. Right elbow ROM was 120 degrees of flexion and -90 degrees of extension, with complete limitation in supination and pronation. Manual muscle testing showed elbow flexion strength of 3/5 and extension strength of

**Table 1.** Summary of CRPS patient demographics, symptoms, and treatments

| Demographics                           | Patient's feature                                                 |
|----------------------------------------|-------------------------------------------------------------------|
| Sex                                    | Male                                                              |
| Age                                    | 28 years old                                                      |
| Injury cause                           | Combat injury with shrapnel                                       |
| Injury date                            | September 2018                                                    |
| Smoker                                 | No                                                                |
| <b>Symptoms</b>                        |                                                                   |
| Hyperalgesia                           | Yes                                                               |
| Allodynia                              | Yes                                                               |
| Range of motion                        | Decreased                                                         |
| Motor dysfunction                      | Weakness                                                          |
| Edema                                  | No, in the moment (It was in the early years.)                    |
| Trophic changes                        | Yes                                                               |
| Itchiness                              | No                                                                |
| Burning sensation                      | Yes                                                               |
| Numbness/Tingling                      | Yes                                                               |
| Erythema                               | No                                                                |
| <b>Treatments</b>                      |                                                                   |
| Physical therapy                       | Tried, the patient refused.                                       |
| ROM exercise                           | Tried, the patient refused.                                       |
| Load bearing the limb                  | No, the patient refused.                                          |
| Fluidotherapy                          | Yes, Non-effective                                                |
| Contrast bath treatment                | Yes, Non-effective                                                |
| Psychotherapy                          | Yes                                                               |
| Neurocognitive methods                 | No, transcranial magnetic stimulation tried, the patient refused. |
| Non-steroid antiinflammatory drugs     | Yes, Non-effective                                                |
| Systemic steroid                       | Yes, Non-effective                                                |
| Vitamin B                              | Yes, Non-effective                                                |
| Vitamin C                              | Yes, Non-effective                                                |
| Vitamin D                              | Yes                                                               |
| Antidepressant (duloxetine)            | Yes, Minimally effective                                          |
| Anticonvulsant (pregabalin)            | Yes, Minimally effective                                          |
| Narcotic analgesic                     | Yes, Minimally effective                                          |
| Stellate ganglion blockade             | Yes, Non-effective                                                |
| Spinal cord stimulator                 | Yes, Non-effective                                                |
| Dorsal root ganglion (DRG) stimulation | No, the patient refused.                                          |



**Figure 2.** X-ray graph in January 2024. X-rays showed osteoporosis and shrapnel. Although there was no arthrosis in the joints, range of motion was limited.



**Figure 3.** The patient's stump and use of a prosthetic hand. The patient underwent below-elbow amputation on 26 March 2024. The length of the stump is 22 cm from the right olecranon. Amputation was performed in the area, which was free of hyperkeratinization and suitable for prosthesis use. He was able to perform daily activities such as writing and drinking water using his prosthetic hand.

4/5. Right shoulder ROM was limited to 70 degrees abduction, 60 degrees flexion, 20 degrees extension, and 70 degrees internal and external rotation. Shoulder muscle strength was 4/5.

The patient was unable to move his right hand for six years and could not tolerate any contact, including water. He reported no benefit from long-term physical therapy and rehabilitation, severe sleep disturbances due to pain, and social isolation due to the skin's appearance and odor. Amputation was deemed necessary. The patient requested amputation, hoping to use a prosthetic limb. A multidisciplinary team, including physiotherapists, psychologists, pain specialists, orthopedic surgeons, hand surgeons, cardiovascular surgeons, and physical medicine and rehabilitation specialists, evaluated the case. After exhausting all treatment options, below-elbow amputation was performed on March 26, 2024, leaving a 22 cm stump from the olecranon, as shown in Figure 3.

Four weeks post-amputation, the patient's VAS pain score decreased to 4, and his quality of life improved. He was prescribed a myoelectric-controlled arm prosthesis, and the COVVI Hand (COVVI Ltd., Leeds, UK), a two-channel myoelectric-controlled multi-articulating prosthetic hand, was integrated into a myoelectric below-elbow prosthetic configuration for patient rehabilitation (Figure 3), and he began using his right hand functionally with the prosthesis. A written informed consent was obtained from the patient for publication of this case report.

## DISCUSSION

In this report, we present a patient with severe refractory CRPS who did not respond to conventional therapies. The patient was subsequently treated by amputation. In a study by Galer and Jensen,<sup>[7]</sup> 84% of patients confirmed the presence of at least one neglect symptom, and 47% reported both 'cognitive' and 'motor' neglect symptoms. In CRPS, inattention and avoidance of the affected limb is confirmed.<sup>[8]</sup> Although neglect of the painful limb has been described in CRPS, the association of DN with CRPS syndrome has not been previously reported. To the best of our knowledge, this is the first reported case of DN occurring in association with CRPS.

Skin changes are common in CRPS, and skin changes are also included in the diagnostic criteria of CRPS. It has been shown that skin findings

of vascular origin are most prominent in patients with CRPS.<sup>[9]</sup> Dermatological manifestations usually increase in severity in later stages of CRPS. Early diagnosis and management of CRPS are the main factors determining the chances of a favorable outcome with treatment.<sup>[10]</sup> Patients with significant concurrent psychological distress and/or poor coping mechanisms may exhibit pain-related behaviors and severe CRPS symptoms.<sup>[11]</sup> In this particular case, the psychological impact of the war injury may have been significant. The patient's non-compliance with early treatments due to psychological factors may have led to the progression of findings and the development of refractory CRPS.

The patient's multiple injuries may have caused CRPS to remain refractory. Multiple nerve injuries may have increased the intensity of pain. Nerve injury may have led to immobilization, making treatment more difficult. Abnormal activation of the sympathetic nervous system is accepted as the basic pathogenic mechanism of CRPS. Hypersensitivity to catecholamines develops after nerve damage.<sup>[12]</sup> Due to the multiple mechanisms of CRPS disease, it has been suggested that CRPS patients suffer from both nociceptive, neuropathic, and nociplastic pain from chronic pain types over time. The pain experienced by CRPS patients typically begins with nociceptive pain following trauma due to peripheral tissue damage and persistent inflammation. It continues with neuropathic pain, which progresses through peripheral sensitization and neuroinflammation. As the process continues, central sensitization may develop.<sup>[13]</sup> The presence of both nerve damage and soft tissue injury from shrapnel may have increased nociceptor activation and contributed to the non-response to treatment.

The case presented was treated by amputation. Quality of life improved in 66% of patients who underwent amputation, and 37% were able to use a prosthesis. Phantom limb pain, CRPS recurrence, and stump pain complications have been reported as the main risks after amputation, and there is also a risk of reduced quality of life in patients.<sup>[4]</sup> As amputation is an irreversible treatment, it may be considered as a treatment option after the patient has been fully informed of the risks. The benefits and harms of amputation in CRPS have been reported. It remains unclear whether amputation should be used to treat refractory CRPS.<sup>[14]</sup> However, in our case, amputation reduced pain and improved quality of life.

In conclusion, early recognition and appropriate management of post-traumatic complications,

including psychological conditions, are important for reducing the risk of CRPS development and progression. Despite multidisciplinary treatment, some patients may remain refractory to conventional therapeutic approaches. To the best of our knowledge, this is the first reported case describing the association of DN with CRPS. In selected refractory cases, amputation may be considered as a salvage treatment after careful multidisciplinary evaluation.

#### Author Contributions

H.C., Y.D., K.A.: This study was designed; K.U., G.K.K., N.K., H.C.: The data were collected; H.C., K.U., G.K.K., N.K., Y.D., K.A.: The data were analyzed; H.C.: The results were critically examined by all authors; N.K., Y.D., K.A.: had a primary role in preparing the manuscript, which was edited. All authors have approved the final version of the manuscript and agree to be accountable for all aspects of the work.

#### Conflict of Interest

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

#### Funding

The authors received no financial support for the research and/or authorship of this article.

#### Data Sharing Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

#### AI Disclosure

The authors declare that artificial intelligence (AI) tools were not used, or were used solely for language editing, and had no role in data analysis, interpretation, or the formulation of conclusions. All scientific content, data interpretation, and conclusions are the sole responsibility of the authors. The authors further confirm that AI tools were not used to generate, fabricate, or 'hallucinate' references, and that all references have been carefully verified for accuracy.

## REFERENCES

- 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;23:522-33. doi: 10.1016/S1474-4422(24)00076-0.
- Harden RN, McCabe CS, Goebel A, Massey M, Suvar T, Grieve S, et al. Complex regional pain syndrome: Practical Diagnostic and Treatment Guidelines, 5th Edition. *Pain Med* 2022;23:51-53. doi: 10.1093/pm/pnac046.
- Abd-Elseyed 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* 2024;56:2334398. doi: 10.1080/07853890.2024.2334398.
- Ayyaswamy B, Saeed B, Anand A, Chan L, Shetty V. Quality of life after amputation in patients with advanced complex regional pain syndrome: A systematic review. *EFORT Open Rev* 2019;4:533-40. doi: 10.1302/2058-5241.4.190008.
- Palaniappan V, Sadhasivamohan A, Karthikeyan K. Dermatositis neglecta. *Clin Exp Dermatol* 2022;47:1265-74. doi: 10.1111/ced.15184.
- Harden NR, Bruehl S, Perez RSGM, Birklein F, Marinus J, Maihofner C, et al. Validation of proposed diagnostic criteria (the "Budapest Criteria") for Complex Regional Pain Syndrome. *Pain* 2010;150:268-74. doi: 10.1016/j.pain.2010.04.030.
- Galer BS, Jensen M. Neglect-like symptoms in complex regional pain syndrome: Results of a self-administered survey. *J Pain Symptom Manage* 1999;18:213-7. doi: 10.1016/s0885-3924(99)00076-7.
- Ten Brink AF, Bultitude JH. Predictors of self-reported neglect-like symptoms and involuntary movements in complex regional pain syndrome compared to other chronic limb pain conditions. *Pain Med* 2021;22:2337-49. doi: 10.1093/pm/pnab226.
- Sundaram S, Webster GF. Vascular diseases are the most common cutaneous manifestations of reflex sympathetic dystrophy. *J Am Acad Dermatol* 2001;44:1050-1. doi: 10.1067/mjd.2001.114299.
- Kabani R, Brassard A. Dermatological findings in early detection of complex regional pain syndrome. *JAMA Dermatol* 2014;150:640-2. doi: 10.1001/jamadermatol.2013.7459.
- Dey S, Guthmiller K, Varacallo M. Complex regional pain syndrome. Treasure Island (FL): StatPearls Publishing; 2023.
- Alebouyeh MR, Morsali SF, Zojaji F, Ebrahimi SA, Ahani A, Antar A. Refractory Complex Regional Pain Syndrome: A case report and review of literature. *Anesth Pain Med* 2023;13:e135286. doi: 10.5812/aapm-135286.
- Mangnus TJP, Dirckx M, Huygen FJPM. Different types of pain in complex regional pain syndrome require a personalized treatment strategy. *J Pain Res* 2023;16:4379-91. doi: 10.2147/JPR.S432209.
- Bodde MI, Dijkstra PU, den Dunnen WF, Geertzen JH. Therapy-resistant complex regional pain syndrome type I: To amputate or not? *J Bone Joint Surg Am* 2011;93:1799-805. doi: 10.2106/JBJS.J.01329.