#### **Clinical case**

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# Clinical course of a complex regional pain syndrome type II in upper limb

Evolución clínica de un síndrome doloroso regional complejo tipo II en miembro superior

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ABSTRACT. Introduction: the complex regional pain syndrome type II, also called causalgia, is a rare clinical condition that appears after a traumatic or surgical event with evidence of nervous system involvement. Its clinical presentation is the consequence of a multifactorial pathogenic process that involves peripheral and central mechanisms and has variable clinical manifestations. We present the photographic record of a patient with complex regional syndrome type II. Clinical case: 43-year-old patient who consulted for neuropathic pain during the last four years, of severe intensity, associated with sensory, vasomotor and trophic changes in the right upper limb, as a consequence of neurectomy of the palmar digital nerves of the third finger. Conclusion: achieving the photographic record of the clinical phases of complex regional pain syndrome type II in its entirety is difficult, because not all patients present all clinical phases, a fact that gives relevance to this case.

**Keywords:** causalgia, neurectomy, complex regional pain syndrome.

**RESUMEN.** Introducción: el síndrome doloroso regional complejo (SDRC) tipo II, también llamado causalgia, es una condición clínica poco frecuente que aparece después de un evento traumático o quirúrgico con evidencia de afectación del sistema nervioso. Su presentación clínica es consecuencia de un proceso patogénico multifactorial que involucra mecanismos periféricos y centrales y tiene manifestaciones clínicas variables. Presentamos el registro fotográfico de un paciente con síndrome regional complejo tipo II. Caso clínico: paciente de 43 años que consultó por dolor neuropático durante los últimos cuatro años, de intensidad severa, asociado a cambios sensoriales, vasomotores y tróficos en miembro superior derecho, como consecuencia de neurectomía de los nervios digitales palmares propios del tercer dedo. Conclusión: lograr el registro fotográfico de las fases clínicas del SDRC tipo II en su totalidad resulta difícil, debido a que no todos los pacientes presentan todas las fases clínicas; hecho que otorga la relevancia a este caso.

**Palabras clave:** causalgia, neurectomía, síndrome doloroso regional complejo.

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## Introduction

The complex regional pain syndrome (CRPS) is a progressive entity that appears after a traumatic or surgical event in a limb. The International Association for the Study of Pain (IASP) classified the syndrome in two types: type I, previously called reflex sympathetic dystrophy or Südeck syndrome,<sup>1</sup> characterized by absence of nervous injury, and it represents 90% of the clinical cases. And type II, also called causalgia, is triggered after an injury that involves a peripheral nerve.<sup>2</sup>

Main clinical manifestations include sensory, vasomotor and trophic changes as disproportionate persistent pain, oedema and blood flow abnormalities.<sup>3</sup> It has been described three evolutionary phases: *acute or inflammatory*, which may last from weeks to months; pain is the main symptom and can be accompanied by oedema, hyperaemia, muscle spasm and hot dry skin. The *dystrophic phase*, usually developed from third to sixth month of the beginning of the syndrome, is characterized by persistent pain, hyperhidrosis, coldness, hair growth, cutaneous striae, hard oedema, cyanosis, muscle and bone atrophy. The *atrophic phase* may last many years after the beginning of the syndrome, in this phase pain and vegetative symptoms may lessen, prevailing cutaneous, muscle and bone atrophy, with muscle retraction, joint stiffness and ankylosis, as a result of fibrosis.<sup>4</sup>

Below, we describe, by photographic record, the evolutionary phases in the clinical course of a CRPS type II in the right upper limb that appeared after a neurectomy of the proper palmar digital nerves of the third finger.

#### **Clinical case**

A 43-year-old male patient, mechanic by trade, with no significant medical history, who suffered trauma to the third finger pulp of the right hand with local oedema, pain of moderate intensity, paresthesias and vascular changes that did not improve with anti-inflammatory, ice and rest treatment. Neurectomy of the proper palmar digital nerves of the third finger was performed according to the decision of the doctor treating the patient that triggered clinical presentation, increasing pain, lymphedema, skin retraction and functional loss of the right arm. This was the reason to consult our institution.



Figure 1: Inflammatory phase after neurectomy of the proper palmar digital nerves of the right third finger, first year.



Figure 2: Dystrophic phase in right upper limb, third year.





#### Figure 3:

Atrophic phase with lymphedema changes in right upper limb, fifth year.

## Clinical findings

Clinical history and photographs supplied by the patient, gave evidence of the three evolutionary phases previously described:

- **1.** Acute or inflammatory phase: after neurectomy of the proper palmar digital nerves, the patient developed a progressive presentation of vascular and trophic skin changes, characterized by local oedema, hyperaemia and hyperalgesia (*Figure 1*).
- **2.** Dystrophic phase: persistent pain with inflammatory signs, thin cyanotic skin, cracked friable nails, cutaneous retraction and hand joint fibrosis (*Figure 2*).

**3. Atrophic phase:** after four years of clinical manifestations, pain continued (hyperalgesia and allodynia) in the right hand and compromised the ipsilateral shoulder (shoulder-hand syndrome); contracture in the fingers got worse and lymphedema changes with complete loss function of the limb appeared (*Figure 3*).

Patient manifests a clinical presentation of a CRPS type II severe, which appeared after a neurectomy of the proper palmar digital nerves of the third finger.

## Therapeutic intervention

Patient was treated by a multidisciplinary team, including psychiatric evaluation for major depressive disorder associated with his chronic, painful condition. Patient required pharmacological management with antidepressants, multimodal analgesic management and interventional pain management with multiple peripheral blocks at shoulder level with partial improvement of symptoms, improvement of quality of life, but without recovery of limb functionality.

## Discussion

CRPS type II is a painful, chronic disabling process, secondary to trauma or surgery event in any limb that compromises a nervous structure. The annual incidence varies between 5.47 and 26.2 cases/100,000 people.<sup>3,5</sup> The average age of presentation is 45, with a predominance of feminine gender (60-80%).<sup>5,6</sup> Diagnosis is clinical, based on Budapest criteria, published by IASP in 2007, although they are not yet validated, they have sensitivities of 85% and specificities of 69%.<sup>7</sup> Based on these criteria for diagnosis, a persistent pain, disproportionate to the trigger event, and a symptom in three of the four categories below, must be found. The categories are: sensory (hyperesthesia/allodynia); vasomotor (temperature asymmetry, changes in skin color); sudomotor (oedema, sweating) and motor/atrophic (decreased range motion, weakness, skin changes);<sup>8,9</sup> findings consistent with the clinical picture of the patient in this article.

CRPS type II treatment varies from conservative management (not to perform any intervention), pharmacological management based on the combination of analgesics such as tricyclic antidepressants and anticonvulsants for the management of neuropathic pain, or interventional management with peripheral nerve blocks and psychological therapy,<sup>2,8</sup> but none proves to be superior to the other for pain relief and functional limb recovery. The most effective way is to combine the previously mentioned interventions with a multidisciplinary team of professionals to improve the patient's quality of life,<sup>9,10</sup> as it was done in the patient of this study.

## Conclusion

The photographic record of the clinical phases of a CRPS type II is usually difficult because some patients start their clinical presentation in the dystrophic phase without an acute/inflammatory phase, or because the initial phase is sub diagnosed. For this reason, this manuscript may allow faster and more successful diagnosis and treatments for this clinical entity.

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## Fe de Erratas

En el artículo titulado *Escoliosis de inicio temprano: fisiopatología, diagnóstico y tratamiento*, publicado en el volumen 37, número 2 de 2023 (pp. 99-105), el nombre del autor dice: Sauri-Barraza JC; debe decir: Dabaghi-Richerand A,\* Santiago-Balmaseda E<sup>‡</sup>

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