Complex regional pain syndrome: diagnosis and treatment at the very onset as the key to success?  
A case report with implications for first contact doctors

Cecylia Zych-Litwin¹, Jan A. Litwin²

¹Division of Rheumatology, MEDDIM Specialized Medical Center for Children and Adolescents, Cracow, Poland
²Department of Histology, Jagiellonian University Medical College, Cracow, Poland

Abstract
The case report describes a 67-year-old man who suffered from a minor left ankle injury. Physical examination on day 12 revealed swelling of the foot, erythema on its dorsal surface as well as elevated temperature, hyperesthesia, hyperalgesia and allodynia of that area. The treatment included local application of dexamethasone and oral administration of meloxicam. Within a week the symptoms disappeared and one-year follow-up did not show their recurrence. The presented symptoms allowed diagnosis of the earliest stage of complex regional pain syndrome (CRPS), which may be a disabling and difficult to treat adverse event. This report suggests that immediately introduced simple anti-inflammatory therapy may bring a quick and permanent recovery. Hence, first contact physicians should advise the patient to report such symptoms as burning pain of the injured area lasting for a few days and, if CRPS suspicion is justified by the results of physical examination, they should apply an anti-inflammatory treatment immediately.

Key words: complex regional pain syndrome, onset diagnosis, steroids, non-steroidal anti-inflammatory drugs.

Introduction
Complex regional pain syndrome (CRPS), previously known as reflex sympathetic dystrophy (RSD), reflex neurovascular dystrophy, or Sudeck's atrophy, is a disabling condition characterized by chronic pain, vasomotor and sudomotor changes, motor disturbances and ultimately trophic changes. CRPS mostly concerns extremities and occurs after even a minor injury, especially if followed by limb immobilization. Its pathomechanism is not clear and seems to include classic and neurogenic inflammation, dysfunction of the autonomic nervous system and central sensitization resulting from maladaptive brain plasticity [1–3]. Depending on the absence or presence of identifiable peripheral nerve damage, CRPS is classified, respectively, into type 1 and type 2, but the type of CRPS has no significant relevance to treatment [3].

Because of CRPS’s multifactorial pathomechanism, successful treatment remains a challenge. It includes anti-inflammatory drugs, alpha-adrenergic antagonists, calcitonin and bisphosphonates, free radical scavengers, antidepressants, physiotherapy, as well as invasive methods such as intravenous anesthesia, sympathetic blocks and spinal cord stimulation [3–5]. However, there is still no consensus to determine the optimal treatment for CRPS.

Some clinical studies published so far have emphasized the benefits of early diagnosis and treatment of CRPS [6–9], but they concerned the fully developed syndrome after at least a few weeks duration. The described case proves how important it is to start treatment quickly after the first symptoms of CRPS have occurred, leading to quick and permanent recovery.
Case report

A 67-year-old Caucasian male patient, with no history of diabetes, neurological or dermatological disorders, after a fall suffered from left ankle sprain and incomplete avulsion fracture of the left distal fibular metaphysis diagnosed by X-ray and was referred for further diagnosis. There were no symptoms of nerve injury. The orthopedist recommended ankle orthosis and limited walking with the help of crutches.

On the seventh day after the injury, the patient experienced an intermittent burning sensation/pain on the dorsal surface of the injured foot. According to the patient, this pain “felt like a first degree burn”. The pain occurred several times during the day, lasting 1–4 hours, and was waking the patient up at night. The intensity of the pain and its duration slightly increased in the subsequent days. The patient also reported moderately frequent, mild dystonic spasms of the left calf muscles.

Physical examination on the twelfth day after the injury revealed swelling of the foot, erythema on its dorsal surface, as well as elevated skin temperature, hyperesthesia, hyperalgesia and allodynia of that area.

The onset of CRPS type 1 was diagnosed and the treatment included local application of dexamethasone spray, 0.28 mg/g and oral administration of meloxicam, 15 mg, once a day. The local and oral treatment was continued for 10 and 20 days, respectively. Within a week after beginning of the treatment, all symptoms disappeared except edema, which resolved after the next four weeks. One-year follow-up did not show any symptoms of CRPS.

Discussion

Complex regional pain syndrome does not seem to be widely known, although it is not an uncommon complication of injuries. The diagnosis of CRPS is currently based on a set of the Budapest criteria, established during an international consensus workshop held in Budapest in 2003 and presented in Table I [10, 11].

These criteria modified the previous International Association for the Study of Pain criteria. However, the criteria of CRPS concern only the fully developed disease. It seems that when diagnosing the onset of CRPS, they should be applied with some flexibility. Initially, the pain may be intermittent, to become continuous later in the course of the disease. Since the first CRPS symptoms appear shortly – within a month – after trauma [12], edema and motor dysfunction (decreased range of motion, dystonia) may be attributed to the injury, although dystonia was found to be a symptom distinguishing CRPS type 1 from type 2 in which it does not occur [13]. Sweating asymmetry and trophic changes develop later, in the advanced phase of the disease. The core symptoms of CRPS include burning pain, erythema, elevated temperature, hyperesthesia, hyperalgesia and/or allodynia of the involved area. These symptoms suggest predominance of an inflammatory response and a simple anti-inflammatory therapy including local application of steroids combined with oral administration of non-steroidal anti-inflammatory drugs can bring a fast and favorable effect.

CRPS often affects the joint regions, and the patients are referred to rheumatologist with preliminary diagnosis of rheumatoid arthritis. The rheumatologists should therefore consider the possibility of CRPS, especially if the classical laboratory indicators of rheumatoid arthritis are negative, and the symptoms occurred after injury or overloading of the joint or a specific area.

Conclusions

The presented case should motivate first contact physicians to pay attention to the association of symp-

| Table I. Budapest clinical diagnostic criteria for complex regional pain syndrome [11] |
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| 1. Continuing pain, which is disproportionate to any inciting event |
| 2. Must report at least one symptom in three of the four following categories: |
| Sensory: reports of hyperesthesia and/or allodynia |
| Vasomotor: reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry |
| Sudomotor/edema: reports of edema and/or sweating changes and/or sweating asymmetry |
| Motor/trophic: reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin) |
| 3. Must display at least one sign at time of evaluation in two or more of the following categories: |
| Sensory: evidence of hyperalgesia (to pinprick) and/or allosthenia (to light touch and/or deep somatic pressure and/or joint movement) |
| Vasomotor: evidence of temperature asymmetry and/or skin color changes and/or asymmetry |
| Sudomotor/edema: evidence of edema and/or sweating changes and/or sweating asymmetry |
| Motor/trophic: evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin) |
| 4. There is no other diagnosis that better explains the signs and symptoms |
Onset diagnosis and treatment of complex regional pain syndrome

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References

1. Urits I, Shen AH, Jones MR, et al. Complex regional pain syndrome, Current concepts and treatment options. Curr Pain Headache Rep 2018; 22: 10.
2. Bussa M, Guttilla D, Lucia M, et al. Complex regional pain syndrome type I: a comprehensive review. Acta Anaesthesiol Scand 2015; 59: 685-697.
3. Goebel A, Barker CH, Turner-Stokes L, et al. Complex regional pain syndrome in adults (2nd edition). UK guidelines for diagnosis, referral and management in primary and secondary care. London: Royal College of Physicians 2018. https://www.rcplondon.ac.uk/guidelines-policy/complex-regional-pain-syndrome-adults
4. Żyluk A, Puchalski P. Effectiveness of complex regional pain syndrome treatment: A systematic review. Neurol Neurochir Pol 2018; 52: 326-333.
5. De Mos M, Huygen Fl, Van der Hoeven-Borgman M, et al. Referral and treatment patterns for complex regional pain syndrome in the Netherlands. Acta Anaesthesiol Scand 2009; 53: 816-825.
6. Gay AM, Béréri N, Legré R. Type I complex regional pain syndrome. Chir Main 2013; 32: 269-280.
7. Shah A, Kirchner JS. Complex regional pain syndrome. Foot Ankle Clin 2011; 16: 351-366.
8. Lee J, Nandi P. Early aggressive treatment improves prognosis in complex regional pain syndrome. Practitioner 2011; 255: 23-26.
9. Schürmann M, Gradl G, Rommel O. Early diagnosis in post-traumatic complex regional pain syndrome. Orthopedics 2007; 30: 450-456.
10. Harden RN, Bruehl S, Stanton-Hicks M, et al. Proposed new diagnostic criteria for complex regional pain syndrome. Pain Med 2007; 8: 326-331.
11. Harden RN, Oaklander AL, Burton AW, et al. Complex regional pain syndrome: practical diagnostic and treatment guidelines, 4th edition. Pain Med 2013; 14: 180-229.
12. McBride A, Atkins B. Complex regional pain syndrome. Curr Orthop 2005; 19: 155-165.
13. Verdugo RJ, Ochoa JL. Abnormal movements in complex regional pain syndrome: assessment of their nature. Muscle Nerve 2000; 23: 198-205.