Coexistence of Complex Regional Pain Syndrome Type 1 and Type 2 in a Patient: A Case Report

Kompleks Bölgesel Ağrı Sendromu Tip 1 ve 2'nin Aynı Hastada Birlikteliği: Olgu Sunumu

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Summary

Complex regional pain syndrome (CRPS) is a disorder of the extremities characterized by pain and sensory changes, accompanied by observable findings such as swelling and autonomic, motor and/or trophic abnormalities, and it is classified into type 1 (no nerve lesion) and type 2 (with a nerve lesion). Bilateral or multiple presentation of complex regional pain syndrome type 1 (CRPS 1) is well known. To our knowledge, the coexistence of CRPS 1 and 2 in a patient has not been reported yet. In this case report, a 37-year-old female who developed CRPS 1 in the left upper limb and CRPS 2 in the right upper limb after bilateral fracture of the distal radius, is presented. *Turk J Phys Med Rehab 2010;56:34-6.*

Key Words: Complex regional pain syndrome 1, complex regional pain syndrome 2, bilateral presentation

Özet

Kompleks bölgesel ağrı sendromu (KBAS) ekstremitelerin ağrı ve duyu değişiklikleri ile karakterize, beraberinde şişlik, otonomik, motor ve/veya trofik anormalliklerinin eşlik ettiği bir bozukluktur ve tip 1 (sinir lezyonu yok) ve tip 2 (sinir lezyonu var) olarak sınıflandırılır. KBAS tip 1'in bilateral ve multipl prezentasyonları iyi bilinmektedir. Bilgilerimize göre KBAS tip 1 ve KBAS tip 2'nin aynı hastada birlikteliği daha önce bildirilmemiştir. Bu olgu sunumunda bilateral distal radius kırığından sonra sol üst ekstremitesinde KBAS tip 1, sağ üst ekstremitesinde KBAS tip 2 gelişen 37 yaşında bir kadın hasta sunulmuştur. *Türk Fiz Tıp Rehab Derg 2010;56:34-6.*

Anahtar Kelimeler: Kompleks bölgesel ağrı sendromu 1, kompleks bölgesel ağrı sendromu 2, bilateral

Case Report

A thirty-seven year-old housewife accidentally fell from the balcony of her house and developed bilateral distal radius fractures. She was transferred to a university hospital and was operated in the orthopedic surgery department on the same day. There was a tissue defect on the volar side of the right wrist extending dorsomedially. In exploration, the median nerve was edematous and was compressed by the surrounding soft tissues. Open reduction and internal fixation were performed in her right upper limb and the tissue defect was repaired. Closed reduction under scopy and percutaneous K-wire fixation were done to stabilize the styloid of her left upper limb. After one month, the K-wire was removed. Soon after this procedure, intense pain, swelling, hyperemia, temperature changes, increased hair growth over the dorsum of the hand, dystrophic changes in the nails, abnormal sweating and tremor began in both hands. Three-phase scintigraphy was performed and the 3-hour delayed static image indicated increased periarticular uptake in both hands, consistent with complex regional pain syndrome (CRPS) (Figure 1). Nonsteroidal antiinflammatory drugs were initiated. The signs and symptoms of CRPS improved in the left hand by time. After 3 months, the patient was re-operated for revision of the external fixator. After the procedure, the signs and symptoms in the right hand were increased, and the patient was referred to our department. History and physical examination revealed intense pain, allodynia, hyperpathia, hyperemia, contracture of the wrist, of the metacarpophalangeal

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joints and of the proximal interphalangeal joints, abnormal sweating, and increased hair growth over the dorsum of the hand, hypoesthesia, dystrophic nail changes and tremor in the right hand. Left hand was normal except for pain, slightly increased hair growth on the dorsum of the hand and slight tremor (Figure 2). Visual analogue scale (VAS) score for pain was 9 for the right hand and 2 for the left hand. Because of the hypoesthesia in all dermatomes of the right hand, bilateral nerve conduction velocities and needle EMG was performed. Median and ulnar nerve motor conduction velocities were reduced; sensory compound nerve action potential (CNAP) amplitudes in the radial nerve and motor CNAP amplitudes in the radial, median and ulnar nerves were reduced. Right median, ulnar and radial nerve innervating the distal muscles had neurogenic motor unit action potential (MUAP) changes with fewer MUAPs. These findings revealed partial axonal degeneration of right median, ulnar and radial nerves. EMG of the left upper extremity was normal. With the clinical and scintigraphic findings, CRPS type 1 in the left hand and CRPS type 2 in the right hand were diagnosed. Oral prednisone (30 mg per day, tapering one month regimen) was started. Peripheral steroid with Bier block (2 times), daily therapeutic ultrasound on the stellate ganglion (0.5 W/cm^2), daily range of motion exercises and daily stretching of the contractured joints were performed. The symptoms and signs, except for tremor in the left hand, decreased gradually within 1 month, but symptoms and signs in the right hand persisted. After 3 months, the patient was discharged from the hospital with sequel in her right hand.

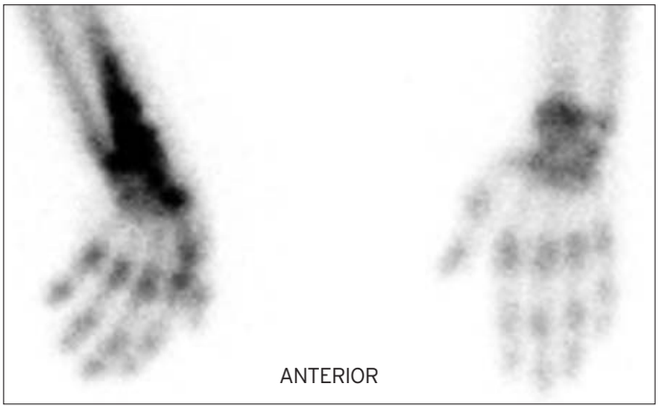

Figure 1. The delayed (third) phase image of the bone scintigraphy demonstrated increased periarticular uptake distally to the fracture zone in both hands.

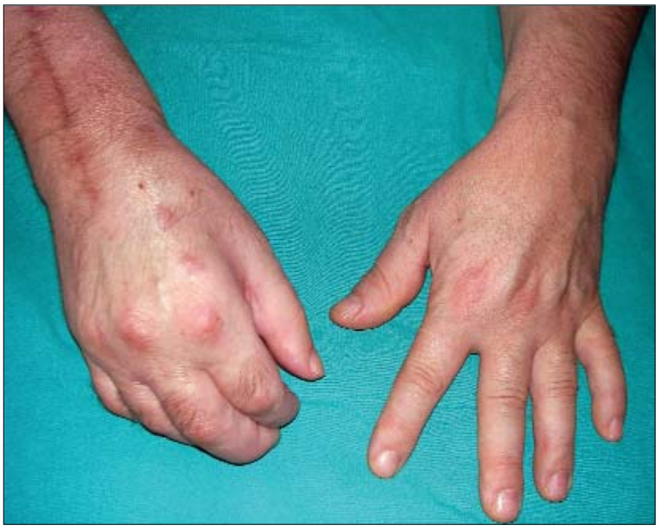

Figure 2. Swelling, increased hair growth and contractures in right hand and increased hair growth over the dorsum of left hand.

Discussion

CRPS is a disorder of the extremities characterized by pain and sensory changes, accompanied by observable findings such as swelling and autonomic, motor and/or trophic abnormalities (1). CRPS type 1 (formerly known as reflex sympathetic dystrophy syndrome) typically develops after a minor trauma such as fracture, sprains, bruises, skin lesions, or surgery with no nerve lesion. CRPS type 2 (formerly known as causalgia) develops after a nerve lesion (2). CRPS type 1 and CRPS type 2 are reported to be different syndromes. CRPS type 2 is by definition a neuropathic pain syndrome due to a detectable peripheral nerve lesion. Pain in CRPS type 1 is more commonly localized in the deep somatic tissues and the sympathetically dependent changes are more prominent in CRPS type 2 (3).

Only a small minority of people develop CRPS after a peripheral trauma. The estimated overall incidence rate of CRPS is 16.2 per 100.000 person-years (4). Of these patients, a very rare group develops CRPS type 1 in bilateral or multiple limbs (5-9). For example, Veldman and Goris (9) analyzed 1183 CRPS type 1 patients and only 10 patients developed CRPS bilaterally. In all those presented cases, trauma that caused CRPS was unilateral and the non-traumatic limb showed CRPS symptoms. CRPS in a limb, mainly upper limb, puts the other limb at a higher risk of developing CRPS symptoms without an injury (7). The opposite extremity is much more likely to respond to peripheral and central autonomic stimuli as the affected extremity, rather than as a normal control extremity (10).

The case presented here is unique because of the development of CRPS type 1 and type 2 in one and the same patient after trauma of bilateral limbs. We think that our patient may have a personal vulnerability to CRPS. It is not very well known why some people develop CRPS and why others do not. The pathophysiology of CRPS is still not very well known, although many peripheral and central theories including neuroimmune and psychological mechanisms have been presented (11,12). The connection between CRPS and HLA-Loci remains unclear, but some kind of CRPS genetic mechanism was found in patients with familial predisposition (13). CRPS is generally accepted to be a systemic disease affecting the central and peripheral nervous system (12). Our case and the previous cases reported in patients developing CRPS bilaterally or in multiple limbs underscore the important role of central nervous system in the pathophysiology of CRPS.

We suggest that, despite the fact that CRPS type 1 and CRPS type 2 may have different mechanisms, they may be the subgroups of a common mechanism, which is a personal vulnerability to a central and peripheral nervous system dysfunction. The reasons for the personal vulnerability to CRPS must be investigated, because the awareness of the predisposition for CRPS, especially in trauma patients, may allow us to take preventive measurements in those patients.

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