Case Report

Complex Regional Pain Syndrome: Two Case Reports

Kompleks Bölgesel Ağrı Sendromu: İki Olgu Sunumu

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ABSTRACT

Complex regional pain syndrome (CRPS) is characterized by swelling, pain, skin changes and vasomotor instability of the affected extremities. The pathogenesis is not completely understood, multiple hypotheses have been suggested and several predisposing factors have been identified. Clinical diagnosis is usually based on symptoms and physical findings. We report two cases of CRPS which involve knee after total knee artroplasthy and index finger after needle stick injury. In this report, we aimed to reveal the importance of early recognition of CRPS, as delay in diagnosis and proper treatment may have detrimental effect on the functional outcome.

Keywords: *arthroplasty; knee; needlestick injury; neuropathic pain*

ÖZET

Kompleks bölgesel ağrı sendromu (KBAS) etkilenen ekstremitede ağrı, şişlik, ödem, cilt değişiklikleri ve vazomotor instabilite ile karakterizedir. Patogenez tam olarak bilinmemekle birlikte çok sayıda hipotez mevcuttur ve birçok predispozan faktör tanımlanmıştır. Genellikle semptomlar ve fizik muayene bulguları ile tanı konur. KBAS tanısında gecikme ciddi fonksiyonel kısıtlılığa yol açabilir. Burada KBAS'ın erken tanınmasının önemini vurgulamak amacıyla total diz artroplastisi ve işaret parmağına enjektör batmasını takiben gelişen iki KBAS olgusu sunuyoruz.

Anahtar Kelimeler: *artroplasti; diz; enjektör batması; nöropatik ağrı*

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INTRODUCTION

Complex regional pain syndrome (CRPS) is the name now given to group of conditions previously described as reflex sympathetic dystrophy (RSD), causalgia, algodystrophy, Sudeck's atrophy and a variety of other diagnosis (1). Two types of CRPS have been described. CRPS type I occurs without a definable nerve damage but with an initiating noxious stimulus, such as a crush or soft tissue injury; or by immobilization of the affected limb. Approximately %90 patients with CRPS have type I. CRPS type II follows a distinct nerve injury. The disorder is characterized by pain, swelling, limited range of motion, vasomotor instability (changes of temprature, colour and sweating), skin changes accompanied by severe functional impairment and patchy bone demineralization. The aetiology of CRPS is not fully understood but involves an exaggeration of physiological responses and is now believed to occur on multiple levels within the central nervous system (2). Peripheric, central, neurogenic and microvascular dysfunction are thougt to be contributing mechanisms in the pathophysiology of CRPS (3, 4). Detoriorated CNS processing with peripheral and central sensitization have association with disturbances within sympathetic nervous system which cause sympathic hyperactivity negatively affecting the damaged area. Studies suggest that amplified inflammatory response plus detoriorated healing are conductive to the unmanagable nature of malicious CRPS (5, 6).

An early study of patients with CRPS noted that the inciting events were soft tissue injury in 40 percent of cases, fractures in 25 percent, myocardial infarction in 12 percent, and cerebrovascular accidents in 3 percent (7). CRPS can occur in any part of the body, however it has been seen more common in the lower extremity than the upper extremity (8). CRPS in upper extremity most commonly occurs after trauma or surgery, but it can also occur after a stroke, heart disease or spontaneously (7).

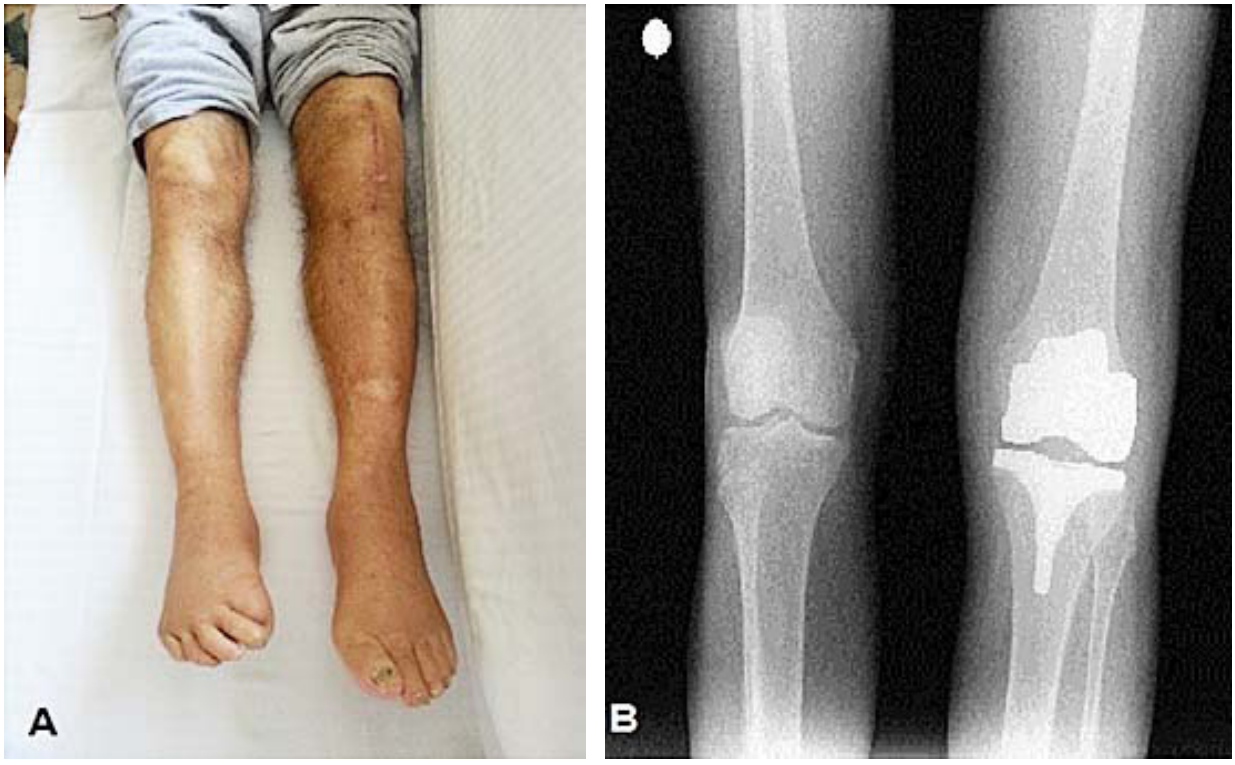

Figure 1-A: Asymmetry, change in skin colour, dystrophic changes in skin and nails. **B:** Anteoposterior radiograph of left knee showing total knee arthroplasty and soft tissue swelling.

In literature, there are no reports describing CRPS occuring after needlestick injury. We report two cases of complex regional pain syndrome which involves left knee after total knee artroplasty and left index finger after needlestick injury. The aim of this report is to increase the general awareness of the disorder and importance of early diagnosis and rehabilitation. Informed consent forms were obtained from the patients.

CASE - I

A 67 year old male who had undergone total knee arthroplasty five months ago was admitted to the outpatient clinic with complaints of persistent pain on his left knee. He complained of difficulty in walking, limitation in knee range of motion (ROM), pain with sensation of electric shock, burning, pins and needles and numbness. He had no history of chronic or metabolic disease. On physical examination he had edema, asymmetria in skin colour and temperature on his left leg (Figure 1A). Allodynia was detected on the left lower extremity with normal sensation and neurological examination otherwise. Functional ability of the patient was restricted to a larger extent, such that patient had difficulty in ambulation, managing stairs and most of household activities were compromised. Daily activities of the patient were restricted to indoor activities only. There were no abnormalities in motor examination. Goniometric measurements of the left knee demonstrated an available active range of 18°; passive flexion to 25° extension limitation was 10°. Total blood count, sedimentation rate, c-reactive protein, renal and liver function tests were within normal range. An anteroposterior roentgenogram of the knee was obtained (Figure 1B). The patient was diagnosed as CRPS type I and combination of pharmacological treatment and physical therapy was planned for the management of severe pain and limitation in ROM. Pregabaline (150 mg twice daily), tramadol (50 mg twice daily), Vitamin C (500 mg/day) and N-acetyl-cysteine (600 mg/day) were given initially. Daily pregabalin dose was increased to 600 mg and pain relief was achieved.

The rehabilitation programme including cold pack, contrast bath, transcutaneous electrical nerve stimulation (TENS) and pulsed ultrasound were applied together with knee ROM, streching and strengthening exercises. Substantial improvements in pain and edema was achieved but the gain in knee ROM was inadequate. Therefore knee manipulation was applied under anesthesia by the orthopedic surgeon.

CASE - II

A 36-year old female working as a nurse in intensive care unit was admitted to the outpatient clinic with the complaint of pain, swellling and limitation of ROM of left index finger. She had history of a sterile needle stick accident on the dorsum of left index finger 3 weeks ago while prepearing a drug for an injection. Her past medical history was otherwise unremarkable.

Figure 2: Edematous, shiny and indurated skin on the left index finger.

Physical examination revealed a red and shiny skin, edema, hyperalgesia, allodynia and limitation in left index finger ROM (Figure 2). Roentgenogram of left hand and laboratory investigation were within normal range excluding fracture and infection. Physical therapy and rehabilitation programme including TENS, contrast bath, massage, mobilization, ROM exercises together with kinesiotaping was applied with the diagnosis of CRPS. Substantial improvement was achieved in pain, edema and finger ROM at the end of 3 weeks. She is still on follow-up with kinesiotape application.

DISCUSSION

There is no specific test to diagnose CRPS, the current criteria used for the diagnosis are based mainly on detailed history, clinical and physical findings. Osseous changes are common in CRPS therefore diagnosis can be supported by radiography and three phase bone scintigraphy. A patchy demineralization in the affected part, which may be the result of disuse can be detected by radiography. Scintigraphy is more sensitive but is not a cost-effective method which is mostly referred to in challenging cases. Differential diagnosis is individualized for each patient, indicated diagnostic investigations should be conducted to exclude causes mimicking signs and symptoms of CRPS including neuropathies metabolic, systemic, vascular, and rheumatological disorders. The important issue in the treatment of CRPS is to provide functional improvement by collaboration of different disciplines. For functional improvement, various invasive or non-invasive modalities that are necessary for rehabilitation, usage and normalization of movements of the affected limb (4). Prompt diagnosis and early treatment is most effective in altering the course of the disease (8). Physical methods are mainly used for the treatment of CRPS.

Especially, application of physical therapy methods in earlier stages of disease helps to prevent edema, atrophy and contracture and reduce pain. TENS can be used as a pain relief method however some patients can not tolerate this modality due to allodynia and hyperalgesia Hence, physical methods applied should be selected on an individual basis (9). In addition, medical treatment and regional anesthetic blocks are other treatment options in CRPS. In medical treatment of CRPS, each treatment option should be tailored to patients' specific problems and symptoms.

In literature, simple analgesics like paracetamol, nonsteroidal anti-inflammatory drugs; corticosteroids in case of any inflammatory condition; opioids, kapsaisin, NMDA receptor antagonists and anticonvulsants (such as gabapentin and pregabalin) for neuropathic pain (allodynia and hyperalgasia); calcitonin, biphosphonates for severe osteoporosis; calcium channel blockers for obvious vasomotor instabilty; antidepressants and sedatives for anxiety, insomina and depression have been used (10, 4).

There are some which investigated the usage of free radical scavengers like dimetilsulfoksit and NAC, baklofen, botulinum toxin, in CRPS. A double-blind placebo-controlled study revealed that Vitamin C is an antioxidant that may reduce the prevalence of CRPS (11). Besides, the result of a study showed that DMSO %50 and NAC are effective in treatment of CRPS type 1 (12). According to our patients' issues we tailored and applied a treatment protocol chosen from physical therapy methods.

In addition pregabalin, opioids, NAC, Vitamin C and biphosphonates were used as a medical treatment. Patients' pain, allodynia and hyperalgesia symptoms were all reduced.

We obtained a functional improvement in second case whereas in first case we could not due to delayed initiation of rehabilitation. In some resistant cases, in addition to above mentioned treatments, psychosocial support, sympathetic and somatic blockade, spinal cord stimulation and spinal analgesia may be required.

Besides, physical and occupational therapy have an important role to have improvements in daily acitivities of the patients. 50-80% of patients who develop refractory CRPS type 1, have disability such as reduction in activities of daily living secondary to chronic pain, limited range of motion and contracture. In comparison of the long-term clinical and functional outcomes of the patient group with TKA knees complicated with CRPS versus the patient group with uncomplicated TKA, Burns AWR et al. found that prognosis and outcomes of TKA patients complicated with CRPS were worse than uncomplicated TKA patients due to having diffucult rehabilitation period due to increased pain, stiffness, anxiety, distress and low mood. Refractory cases seem to benefit from manipulation under anaesthesia as seen in our case. In conclusion, importance of early diagnosis and treatment is very clear and it appears to mitigate against poor results and unsuccessful outcomes.

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