



# Complex regional pain syndrome type 1 in a pediatric patient: Case report

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## Abstract

Complex regional pain syndrome type 1 is one of the causes of morbidity of childhood which is also named reflex sympathetic dystrophía. The syndrome is characterized with regional pain and vasomotor, sudomotor and sensory changes in the distal parts of the extremities involved. Complex regional pain syndrome type 1 shows difference in children in terms of clinical picture and imaging methods compared to adults. The most important point is that the prognosis is generally better in children if early diagnosis and treatment is provided. On the other hand, causes including presence of psychological factors or less contribution of imaging methods in children lead to delayed diagnosis or erroneous diagnosis. In this article, a 10 year-old male patient who was diagnosed with complex regional pain syndrome type 1 was described. Thus, we aimed to remind clinicians that this syndrome should also be kept in mind in the differential diagnosis of pain in children. (Türk Ped Arş 2014; 49: 77-80)

**Key words:** Child, complex regional pain syndrome, reflex sympathetic dystrophía

## Introduction

Complex regional pain syndrome type 1 (CRPS) is a syndrome which affects both adults and children and which leads to morbidity. The first pediatric cases of CRPS were described in 1970s. It was thought that pediatric CRPS was a rare condition and its properties were elucidated better in parallel to increased number of cases (1, 2).

Two types of complex regional pain syndrome have been defined. Complex regional pain syndrome type 1 is also named reflex sympathetic dystrophía. It is characterized with regional pain, vasomotor, "sudomotor" and sensory changes in the distal part of the extremity involved. Complex regional pain syndrome type 2 in which nerve damage accompanies all these changes is named as causalgia (3). The most typical finding in complex regional pain syndrome is long-term and severe pain. Allodynia and hyperesthesia are observed frequently. The patient limits the movements of the extremity involved due to pain. Changes in skin color and temperature, trophic changes in hair and nails, sweating and edema accompany pain. If treatment is not initiated, muscle atrophy, mineral loss in bones and contractures in joints may develop (2-4). The triggering factor is usually traumas like fracture or surgical interventions (2). Psychological factors are mostly effective in children (5, 6).

The diagnosis of complex regional pain syndrome type I is mainly based on clinical findings. History, physical examination and clinical properties are generally sufficient for the diagnosis of CRPS. Imaging methods are helpful in the diagnosis (4). Complex regional pain syndrome is more easily diagnosed in adults, but pediatric CRPS type I shows different properties (7). Therefore, pediatric CRPS type I may be missed or diagnosed erroneously. This is an undesirable picture in terms of pediatric patients who might have good outcomes with early treatment (1, 2).

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The aim of this study was to draw attention to presence of CRPS type I which is difficult to diagnose, for which the diagnosis may be delayed or which may be diagnosed erroneously in the pediatric patient group and to remind clinicians about the importance of early diagnosis and treatment.

### Case

A 10-year old male patient presented to our clinic with complaints of movement limitation and severe pain in the left ankle and difficulty in walking. The patient had a history of trauma in the form of non-communitued fracture in the left tibial distal 1/3 part related with a non-vehicle traffic accident. Open reduction-internal fixation with screw plates operation had been performed in the patient. Six months after the operation he was referred to physical therapy and rehabilitation clinic with complaints of movement limitation, erythema and edema in the left ankle. He had a history of undescended testicle operation at the age of 1.5 years in his personal history. His familial history was normal.

On physical examination of the patient, swelling, warmth, erythema and increased hair was found in the left ankle. Ankle joint mobility (JM) was limited; passive ankle dorsoflexion was 10 degrees and plantar flexion was about 30 degrees. When the pain of the patient was assessed by visual analogue scale (VAS), it was found to be seven. In addition, allodynia was found on physical examination. There was difference between extremity circumference measurements taken from 10 cm proximally and distally to bilateral patellar midpoints (right-left quadriceps circumference measurement: 28.5-27 cm, right-left cruris circumference measurement: 24.5-23.5 cm). JM was normal in all extremities excluding the left ankle. The patient had no pain, warmth or swelling in any other joint in his body. Posterior tibial and dorsalis pedis pulses were normally palpable. Neurological examination of the left lower extremity was performed under suboptimal conditions because of pain.

Hemogram, biochemistry tests, acute phase reactants (erythrocyte sedimentation rate and C-reactive protein) and complete urinalysis were found to be normal. Hematological findings did not suggest another underlying systemic disease. Extensive osteopenia was found in the left foot compared to the right foot on direct graphy (Figure 1).

The diagnosis was made mainly with clinical signs and symptoms. Pain, allodynia, swelling and increased hair which occurred after trauma were compatible with CRPS type I. Presence of extensive osteopenia observed on direct graphy supported this diagnosis. Three-phase bone scintigraphy was planned to confirm the diagnosis, but could not be performed because of technical impossibility.

Primarily analgesics were given to the patient and it was aimed to relieve him in terms of pain. In addition, physical therapy applications including electrical stimulation to quadriceps muscles and transcutaneous electrical stimulation (TCES) in the region with pain were performed in addition to contrast bath, joint mobility, quadriceps strengthening and ankle straining exercises. In



**Figure 1. Marked extensive osteopenia in the left foot**

the follow-up, the patient's pain regressed (VAS:2), swelling disappeared and passive JM measured in the left ankle was complete. Additional treatment was not planned, since the patient's complaints disappeared to a large extent. No additional complaint was found in the follow-up after discharge.

### Discussion

In population-based studies, the prevalence of CRPS type I ranges between 5.5/100 000 and 26.5/100 000 in adults (3). In children, the prevalence of CRPS type I is not known and the syndrome is possibly missed (8). Absence of typical clinical findings especially in the early period, absence of laboratory changes which could help for the diagnosis, the fact that the syndrome shows difference in pediatric patients, absence of prominent radiological and scintigraphic changes which are observed in adults lead to delayed diagnosis and treatment (9, 10). Confusion of pediatric CRPS type 1 with other psychiatric diseases, malingering or conversion may also lead to delayed diagnosis (2, 11). Studies have shown that the period between the onset of the findings and the diagnosis ranges between four weeks and 1 year (2). Our patient was an example case who was diagnosed early. He was referred to our clinic with the onset of the symptoms for the aim of rehabilitation and no time was lost.

Although complex regional pain syndrome type 1 in adults may occur in both genders and at any age, it is observed more commonly in women between the ages of 60 and 70 years (12). Pediatric CRPS type 1 is generally observed between the ages of 5 and 17 years and more frequently in girls (2, 9). In the literature, the youngest patient is 2.5 years old (13, 14). The gender difference is thought to be related with estrogen-dependent pain

responses or psychological problems (9). Our 10-year old patient was compatible with this information, but his gender was male in contrast to the literature.

The most common triggering event for CRPS type I in adults is trauma especially fractures. It is more frequently observed in intra-joint distal radius fractures, fractures related with ulnar styloid traumas and fractures treated by closed reduction and plastering. (15). The presence of triggering events is not as prominent as in adult patients. Low et al. (16) reported that trauma was not reported in half of pediatric CRPS cases, but they observed trauma with a high rate in the cases in their own study. In the study of Tan et al. (9), trauma was found in 45% of the cases. In contrast to adults, even a milder trauma in children may initiate the event (2). Hence, both Low et al. (16) and Tan et al. (9) found minor trauma in their cases. Although the triggering factors leading to complex regional pain syndrome in children are not very clear except for trauma, psychogenic factors are frequently confronted. In the literature, anxiety disorder, adaptation problems, post-traumatic stress disorder and conversion have been emphasized among the etiological causes causing to this syndrome (11). In addition, Munchausen syndrome, administration of rubella vaccine and presence of migraine are among other factors (17,19). In our patient, there was a history of a clear and important trauma and psychogenic or non-psychogenic factors mentioned above could not be found.

The localization of complex regional pain syndrome type I is different in adults and children. In adults, the upper extremities are involved with a higher rate, while the lower extremities are involved with a higher rate in children (2). In the study of Tan et al. (9), it was reported that CRPS type I was observed with the highest rate after ankle trauma. The left extremity was involved with a higher rate in children in the same study (9). In our patient, CRPS was found in the left ankle region (lower extremity) compatible with this information.

The diagnosis of complex regional pain syndrome type I is made primarily with clinical findings. Methods including radiology, scintigraphy, magnetic resonance, thermography and electromyography are helpful in confirming the diagnosis (19). In adult patients, extensive osteoporosis accompanied by patchy demineralization is observed by direct imaging. However, this image occurs in the late stage of the disease and is not specific (20). In children, extensive demineralization and osteoporosis are observed in 50% of the cases (21). Especially bone scintigraphy is used as a diagnostic method in adult patients. Increased uptake is the rule on scintigraphy. In studies conducted with pediatric patients, it has been reported that scintigraphy results may be completely normal or may be compatible with decreased uptake in contrast to adults (8,16). Cimaz et al. (8) emphasized that bone scintigraphy was not regular, since its specificity and sensitivity are insufficient in the diagnosis of pediatric CRPS (8). In various case series, it has been reported that scintigraphic findings are not as useful in making the diagnosis of the disease as in adults (9, 10). Similarly, in our patient, the diagnosis was made based on the clinical findings. In the history, presence of trauma, normal laboratory findings and osteopenia in radiology were directive in

the diagnosis. Scintigraphy could not be performed because of technical impossibility.

The treatment of CRPS in children is still controversial. In children, treatment should basically consist of non-invasive methods including medical treatment, physical therapy and psychotherapy, if necessary especially if the diagnosis is made in the early stage (22-24). Medical treatment includes various options including analgesics, non-steroid anti-inflammatory drugs, steroids, calcitonin, biphosphonates, alpha and beta adrenergic blockers, anticonvulsants, tricyclic and other antidepressants and gabapentin. In physical therapy, warm or cold application, contrast bath therapy, electrical stimulation, TENS, massage, tactile desensitization, hydrotherapy methods and exercises are used. However, further more qualified studies are needed to determine the efficiency of physical therapy methods in children. In resistant cases, sympathetic and epidural blocks and spinal cord stimulation may be needed (22, 25). It has been reported that very disciplined team rehabilitation increase response to treatment in children (26). This syndrome has a better prognosis with early treatment in children compared to adults. Patients who are resistant to treatment are generally the ones with delayed diagnosis or severe psychological problems (8).

In this case, the complaints of the patient were controlled with medical treatment which consisted of analgesics and physical therapy methods which consisted of contrast bath, exercise, electrical stimulation and TENS. Additional treatment was not needed. Observation of complete recovery especially with early diagnosis and treatment in children in contrast to adults was also true for our case (27). No additional complaint developed in the follow-up of our patient. However, it should be kept in mind that patients who developed dysfunction have been reported in the literature (22).

## Conclusion

In our study, we aimed to remind clinicians about complex regional pain syndrome type I by describing this syndrome in a 10-year old child. In this group, the syndrome is diagnosed especially with clinical findings. Therefore, CRPS type I should be considered in the differential diagnosis in clinical conditions including severe pain, edema, changes in the color and temperature of the skin, trophic changes in nails and hair and it should be kept in mind that the prognosis is satisfactory with early diagnosis.

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