Case Report
JCAM

A Rare Case in Pediatric Neurology: Complex Regional Pain Syndrome

Pediyatrik Nörolojide Nadir Bir Olgu: Kompleks Bölgesel Ağrı Sendromu

Complex Regional Pain Syndrome in Child

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Öz

Kompleks bölgesel ağrı sendromu (KBAS) duyusal, trofik ve motor anormallikler ile nitelenen çok etmenli bir ağrı sendromudur. Tip 1 KBAS'ta bir sinir hasarı saptanmazken, tip 2 KBAS'a belirlenebilen bir sinir hasarı eşlik eder. Çocukluk ve adölesan çağında nadir olduğu düşünülmesine karşın, son 20 yılda daha iyi tanımlanmaya başlamıştır. Ancak, erişkin ve çocukluk çağı KBAS klinik bulgularının farklılığı, özel laboratuar testlerin ve görüntüleme tekniklerinin bulunmayışı, tanıda bir yıla kadar gecikmeye neden olmaktadır. KBAS'ta erken tanı ve tedavi iyi prognoz ölçütü olduğundan, hastalığın klinisyenler tarafından farkındalığı önemlidir. Bu yazıda, sağ bacakta ağrı, renk değişikliği ve yürüme güçlüğü ile başvurup, KBAS tip 1 tanısı alan, rehabilitasyon ve pregabalin tedavisine çok iyi yanıt veren 11 yaşında bir erkek hasta sunulmuştur.

Anahtar Kelimeler

Tip 1 Kompleks Bölgesel Ağrı Sendromu; Çocuk; Livedo Retikülaris; Ekstremite Ağrısı

Abstract

Complex regional pain syndrome (CRPS) is characterized by multifactorial pain disorder in combination with sensory, autonomic, trophic and motor abnormalities. CRPS type 1 refers to cases in which no specific nerve injury is identified, while type 2 are cases accompanied by identifiable nerve damage. Once considered to be a rare disorder among children and adolescents, CRPS has become better recognized over the past two decades. The clinical differences between adult and pediatric CRPS and the lack of specific laboratory tests and imaging techniques cause a delay in diagnosis of up to one year. Awareness of the syndrome is very important for the early diagnosis and treatment and is a positive prognostic factor. Here we present an 11-year-old male patient with pain, color changes of the skin and motor deficit in the right leg diagnosed with CRPS type 1. The patient responded well to rehabilitation and pregabalin treatment.

Keywords

Complex Regional Pain Syndrome Type 1; Child; Livedo Reticularis; Extremity Pain

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Introduction

Complex regional pain syndrome (CRPS), formerly known as reflex sympathetic dystrophy, is a chronic neuropathic pain syndrome that is believed to be a result of a dysfunction in the central or peripheral nervous system. It is characterized by persistent burning pain and vasomotor changes. Generally only one extremity is involved; however, any part of the body may be affected [1]. CRPS type 1 occurs without a definable nerve lesion, whereas type 2 is associated with a definable nerve lesion usually secondary to a trauma [2]. Patients might describe allodynia (non-painful stimuli evoking pain), hyperalgesia (painful stimuli evoking more intense pain than usual) and hyperpathia (repeated painful stimuli causing exaggerated response) [2]. Here, we present an 11-year-old male patient diagnosed with CRPS type 1 who was referred to our pediatric neurology clinic with difficulty in walking, pain and color changes of the skin of his right leg.

Case Report

An 11-year-old male patient was admitted with pain, color changes and weakness in the right leg. The color change had first appeared 5-6 months previously. It had grown in size and was accompanied by an intense pain. Weakness and difficulty in walking were his more recent complaints. He stated that during a bath, the intensity of pain increased in contact with hot and cold water and the blue discoloration of the skin became darker. Sometimes he felt cold in the region of the color change. His mother stated that he is generally shy and he never wants to go to the school, and that this intense pain recently affected his school attendance negatively.

Results of a neurological examination were normal. Livedo reticularis was noticed in the region of the pain (Figure 1). Laboratory investigations including complete blood count, renal and hepatic function tests, electrolytes, C-reactive protein, and creatine kinase levels were all normal. Viral serology was negative. Screening for vasculitis revealed negative results. Doppler ultrasonography for deep venous thrombosis results were normal. Electroneuromyography did not show any pathological changes. All cranial, spinal and right femur magnetic resonance imaging (MRI) were reported to be normal.

The diagnosis of CRPS type 1 was established according to the modified Budapest International Association for the Study of Pain (IASP) criteria [1]. The patient was consulted with the Physical Medicine and Rehabilitation (PM&R) Department. Treatment with paracetamol and codeine (addition of peripheral and central analgesic effect), vitamin C, and 75 mg pregabaline twice daily were started. He was given stretching and muscle-strengthening exercises because of weakness, atrophy and shortness in the right quadriceps muscle. Muscle-strengthening exercises for the other muscle groups of the lower extremity were also started because of weakness related to immobilization. His complaints decreased markedly after two weeks of treatment. The intense pain of his right leg reoccurred just once, when his grandfather, who had come from his village for a visit, returned home. Fluoxetine was started and his complaints subsided gradually within a few weeks of treatment. Monthly control visits for 6 months revealed that school attendance had improved.

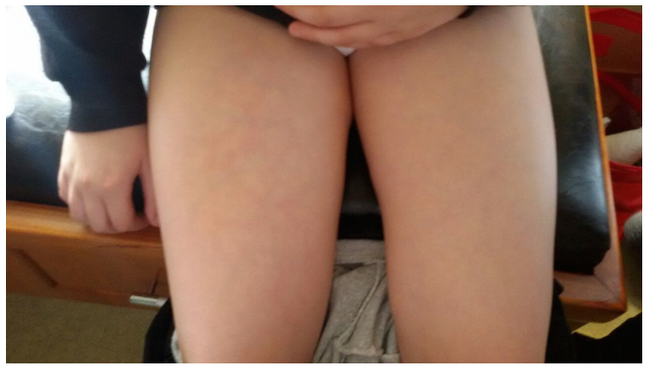

Figure 1. Livedo reticularis on the right thigh of the patient.

Discussion

In childhood, CRPS type 1 is a rare condition causing diagnostic challenges [3,4]. It is more common in school-aged children and mainly adolescent females, with a mean age at onset of 11-12 years [5]. Compared with adults, pediatric patients with CRPS type 1 have a higher incidence of lower extremity involvement and almost half of the patients have no history of trauma preceding the onset of symptoms [5]. Tan et al. [6], in a study of 78 children under age 16 with CRPS type 1 seen between 1980-2014, found that in children CRPS type 1 is less likely to be associated with edema and skin temperature is mostly cold, whereas edema and warm skin temperature are more commonly observed in adults with CRPS type 1. Psychogenic factors and anxiety are thought to play a major role in the development of CRPS, especially in childhood [7]. In contrast to the female preponderance described in the literature, our case was an 11-year-old male. Lower extremity involvement, lack of an initiating trauma, cooler feeling in the region of the color change, and lack of edema were features consistent with the literature.

The pathophysiology is not completely understood; however, current opinion is that activation of cutaneous nociceptors as a result of tissue damage stimulates unmyelinated C fibers and A delta afferents, leading to a neurogenic inflammation. Sympathetic pain has a major role in the early periods of the disorder. Peripheral sensitization occurs with the release of cytokines as a result of neurogenic inflammation and the release of algesic neuropeptides such as substance P. Central sensitization follows as a result of activation of N-methyl-D-aspartate (NMDA) receptors and altered functions at the level of the dorsal root ganglion [2].

The diagnosis of CRPS type 1 is clinical and a high level of vigilance is needed to make the diagnosis. Average delays in diagnosis of up to a year have been reported in the 1990's [8]. More recent studies have shown marked improvement with an average delay of about 3 months, which still is not ideal [5]. The differential diagnoses include inflammatory arthritis, cellulitis, osteomyelitis, deep venous thrombosis, malignancies and chronic vascular abnormalities [2].

The treatment of CRPS type 1 is multidisciplinary. Rehabilitation is the primary mode of treatment. The basic principles involve posture, range of motion, muscle stretching and strengthening exercises. The aim of the therapy is the prevention of immobilization caused by the persistent pain. Psychotherapy and psychiatric evaluation is a part of the treatment in child-

hood [5]. Nonsteroidal anti-inflammatory drugs, central analgesics, amitriptyline, pregabalin, gabapentin, corticosteroids and vitamin C can be used for pain management. In childhood, the response rate to conservative treatment is good, ranging from 70 to 90 %; however, recurrences have been reported in 30-50 % of cases [5,6]. Low et al. [5] reported that in children who were diagnosed early in the disease course (< 3 months), symptom resolution occurred much more rapidly than in those diagnosed later.

In conclusion, although rare, CRPS type 1 should be considered in the differential diagnosis of extremity pain. Being familiar with the symptoms and signs of the disorder makes early diagnosis and intervention possible and are positive prognostic factors.

Competing interests

The authors declare that they have no competing interests.

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