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A Rare Cause of Swelling and Pain in Extremities in Children: Complex Regional Pain Syndrome

Complex regional pain syndrome is a condition of uncertain etiology characterized by spontaneous or stimulus-induced pain that is out of proportion to the inciting event. We report a 14-year-7-month-old girl with swelling of the left hand and wrist, was diagnosed as complex regional pain syndrome. The patient was treated successfully with physical therapy and non-steroidal anti-inflammatory drugs. This condition should be kept in mind in the differential diagnosis of musculoskeletal non-inflammatory and inflammatory pains.

Key words: *Diagnosis, Management, Recurrent pain*.

Musculoskeletal pain and swelling are common reasons of admission for pediatricians. The causes include a variety of inflammatory and non-inflammatory diseases such as arthritis, myositis, fibromyalgia, hypermotility, growing pains [1]. Complex regional pain syndrome (CRPS), previously known as reflex sympathetic dystrophy, is a chronic pain condition usually affecting distal extremities characterized by spontaneous or stimuli-induced pain [2]. Herein we report an adolescent patient diagnosed with CRPS and treated successfully.

A 14-year-7-month-old girl was admitted to our hospital with swelling of the left hand and wrist, pain and redness on the whole arm, and difficulty in moving the arm for 2 days. There was a feeling of numbness and tingling on the left hand and in the left upper extremity with an itching and burning sensation in her left arm described as hyperalgesia. The pain had flammable character and was exacerbated with movement or light touch suggesting allodynia. The decrease of the pain was reliable when the contact was stopped. The patient reported no preceding trauma prior to admission. No prior infection or fever was reported. Her past medical and family history was unremarkable.

On physical examination, the patient was afebrile. Hyperemia and swelling was prominent on the distal part of the left upper extremity and on the palms of the left hand, plantar skin and the fingers. There was edema of the entire left upper limb. She had difficulty in moving the left and the fingers. Hyperalgesia, hyperesthesia and allodynia were prominent in the left hand and upper extremity. Pain sensitivity was prominent. In the laboratory evaluation, all of the results were in normal ranges for the complete blood count, erythrocyte sedimentation rate, C- reactive protein, coagulation tests, vitamin B_{12}, antistreptolysin O (ASO), and routine blood chemistry. Serologic evaluation for infections and rheumatologic evaluation were negative. Superficial tissue and Doppler ultrasonography, Echocardiography, direct radiographs were all normal. The magnetic resonance imaging of spine was did not show any abnormality. These findings, in conjunction with the history, were suggestive of CRPS type I.

The patient received physiotherapy that was performed intermittently in the therapy pool; as well as using passive therapy as the massage of the left arm. Beside the physical therapy, non-steroid anti-inflammatory drug treatment with naproxen sodium was initiated with a dose of 10 mg/kg/day. On the third day of therapy, the pain of the patient improved. Medical treatment lasted for 15 days, but the course of the disease lasted for approximately one month. She has been followed out-patiently by physical therapy for has 3 months without any symptoms.

CRPS is a term to describe conditions predominantly characterized by spontaneous or stimulus-induced pain that is inconsistent with the provocative event [3]. CRPS diagnosis entirely depends on observable signs and reported symptoms, which have been put together into various diagnostic criteria sets for adults [4]. The diagnosis of CRPS in children may be delayed as long as four months because of the low incidence and considerably different clinical presentation compared with adults [5]. The clinical manifestations of CRPS may imitate rheumatologic diseases in children.

CRPS has been suggested to be a multifactorial condition that is related to an unusual host response to certain tissue damage. The disease often includes a wide diversity of autonomic and motor disturbances like hyperalgesia, allodynia, and sensory loss [4,5]. The patient can exhibit particularly painful, red, warm, and swollen extremities, mimicking trauma. Other probable accompanying features are changes in sweating, reduced hair and nail growth, allodynia and hyperalgesia, and also muscle weakness [2]. However, apparent trauma or initiating factor is absent in most of the pediatric patients. Exclusion of other possible causes is necessary and investigations are needed to exclude infections of skin, connective tissues, muscles, bone and joints. Further investigations may be needed if inflammatory diseases, fractures, neoplasms and deep venous thrombosis are suspected [6]. In our patient, the possible conditions were excluded by further clinical and laboratory evaluation.

The optimal management approach should consist a multidisciplinary treatment of noninvasive interventions including physiotherapy, occupational therapy, analgesics and psychotherapy [5,6]. Sensory rehabilitation is sometimes added in order to gradually improve the allodynia [6]. The standard medications include drugs non-steroidal anti-inflammatory drugs, antidepressants, anticonvulsants, topical analgesic patches. In contrast to adults, the response to treatment, particularly exercise therapy with behavioral management will achieve almost 97% remission in children [3], as in the reported child.

We report a pediatric CRPS Type I patient, treated successfully by conservative methods, in order to attract attention to this rare benign condition in children.

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Extra-nodal Kikuchi Disease and Kikuchi Encephalitis

Kikuchi disease is a rare but benign, self-limiting disease that typically presents with prolonged fever and cervical lymphadenopathy. We report neurological manifestations in an adolescent girl with recurrent Kikuchi disease.

Keywords: *Lymphadenopathy, Meningoencephalitis, Necrotizing lymphadenitis, Recurrence*.

Kikuchi-Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis has a recurrence rate of around 3-7% in reports from most centers but can range as high as 20% in predisposed East-Asian populations [1]. Its pathogenesis still remains controversial [2]. Neurological manifestations of Kikuchi disease are extremely rare and isolated, and include headache, peripheral neuropathy, aseptic meningitis to encephalitis [3,4].