A 12-year-old female patient referred to the Department of Pediatrics on April 9, 2013, with complaints of swelling, pain, reddening, and movement difficulty in her right hand. Her attacks repeated 3 times. Cervical stellate ganglion blockage relieved symptoms immediately at the last attack.

Keywords: Complex regional pain syndrome, stellate ganglion, treatment

Introduction

Complex regional pain syndrome (CRPS) is a clinical condition that features a group of typical symptoms, including spontaneous pain, edema, tenderness, swelling of an extremity associated with varying degrees of sweating, warmth and/or coolness, flushing, discoloration, and shiny skin. The condition is generally observed in the distal of the extremities, but may spread to the proximal parts. A gold standard order in the diagnosis of CRPS has not been found yet, diagnostics are depended on the patient’s medical history and correlating clinical findings. The International Association for the Study of Pain compiled and later on revised operational diagnostic criteria resulting in a satisfactory sensitivity and specificity for both research and clinical needs. Additionally, diagnostic examinations can support the clinical suspicion (1-3). The definite prevalence of CRPS is unknown. The reported prevalence rates of CRPS in predisposing medical conditions are as follows: 5-20% in coronary artery disease, 12-21% in hemiplegia, 0.2-35% in colles fracture, 3% in peripheral nerve injury, 1-2% in bone fractures, and 0.5% in other traumatic injuries (4-8). The therapy of CRPS is quite difficult, where the first step should be to take measures to decrease the risk in medical conditions triggering CRPS (4-6). The two aims of the therapy are to eliminate the pain and to restore function.

Case

A 12-year-old female patient referred to the Department of Pediatrics on April 9, 2013, with complaints of swelling, pain, reddening, and movement difficulty in her right hand. Formerly, she had been hospitalized with the diagnosis of familial mediterranean fever (FMF) upon attacks of abdominal pain occurring in every two weeks for two years. She had had no fever, rash or arthritis during the attacks. She had undergone surgery for abdominal hernia at the age of 6, and now and then she had secretions coming from the operational wound. During her hospitalization she had been examined for the
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presence of FMF and cellulitis, but no definite diagnosis could be reached. She had received deltacortril 5 mg 3x2 daily, and her complaints had started to regress. Following consultation, the patient was transferred to our clinic of physical therapy and rehabilitation. Her physical examination showed edema, hyperthermia, sweating, extreme tenderness, and severe pain in her right hand (Figures 1, 2). The patient could not move her right hand because of the pain. The patient was diagnosed as CRPS, and physical therapy was initiated, and steroid was continued at a dose of 5 mg. With therapy, her complaints disappeared completely, and steroid was stopped.

In her laboratory examinations, the values of hemogram and routine biochemical tests were within normal limits. The hepatitis markers, anti-nuclear antibodies, and anti-DsDNA were negative; and peripheral blood smear, complement values, and acute phase reactants were normal. There was no pathology in the X-ray examination of her right hand. Magnetic resonance showed soft tissue edema in her hand. The 3-phase bone scan of the hand was also normal. No peripheral nerve damage was detected in the electromyographic examination.

The patient re-visited our outpatient clinic for physical therapy with her former complaints on October 7, 2013. She was hospitalized again with the diagnosis of CRPS and received physical therapy and non-steroidal anti-inflammatory drugs (NSAID). Contrast bath, transcutaneous electrical nerve stimulation, ultrasound in water, massage therapy were administered 10 sessions as physical therapy programme. Her complaints regressed in time, and she was discharged. The second 3-phase bone scan of her right hand was again normal. The arterial and venous Doppler ultrasound of the right arm was also normal. Her laboratory test results were within normal ranges. She suffered surgery to clarify the secretion from the umbilicus. The results of abdominal and superficial peri-umbilical soft tissue ultrasound imaging detected no pathology.

The patient revisited our outpatient clinic for the third time with the same former complaints on November 25, 2013. As therapy, she again received NSAID and underwent stellate ganglion blockade. Her complaints regressed in time. As a result, the symptoms of the patient and positive response to applied treatments supported CRPS diagnosis.

Discussion

CRPS can strike at any age, but is relatively rare in childhood and adolescence. Its prevalence is about 10% in childhood. In childhood, the lower extremities are affected more frequently, and this situation is observed more often in girls (9). Reflex sympathetic dystrophy syndrome generally occurs following a trauma, surgery, strain or fracture. If it occurs following a nerve injury, a local or regional motor and sensory disorder may develop (7). In children, a minor trauma or psychological stress may trigger CRPS. The clinical findings may vary with the stage of the syndrome. The specificity and sensitivity of the bone scan used in the diagnosis may also alter. Bone scan is useful in the diagnosis of early cases with different clinical findings, but its sensitivity decreases in late cases. In the absence of CRPS, bone scintigraphy may give positive results in other clinical disorders with biological processes similar to those of CRPS. Consequently, bone scan is not essential for the diagnosis, and CRPS should be diagnosed clinically (10).

In CRPS, the three main pathophysiological changes are aberrant inflammation, vasomotor dysfunction, and incompatible neuroplasticity. Which of these pathophysiological changes mediates the bone changes is still a matter of dispute (11). The increased absorption of the tracer in bone scan may point to neurogenic inflammation, decreased sympathetic activity or bone demineralization caused by active bone metabolism (8,12). Bone scan may be used in determining patients who would benefit from biphosphonate therapy. In contrast to adults, bone scans in children and adolescents display diffuse decreased tracer absorption in all three stages of the disorder, but bone scans with increased absorption have also been reported (10-15). A study reported that among 103 children with CRPS, the bone scan of the affected area showed increased accumulation of the radiotracer in 31, decreased accumulation in 11, and normal absorption in the rest (14).
No factor is found in the etiology of about 35% of CRPS cases. Recurrent or migratory CRPS is rare (16, 17). The recurrence of CRPS may be decreased by physical therapy and cognitive behavioral therapy. In childhood CRPS, an intensive exercise therapy is crucial. Epidural or ganglion blockade, intravenous anesthetics, and repeated peripheral nerve blockade may also be added to the therapy (18). Sherry et al. (12) reported the psychological factors triggering the recurrence of CRPS in children with history of former suicide attempt, anorexia nervosa or bulimia, and less complaint of pain. They also found that intensive exercise therapy decreased the risk of recurrence of CRPS (14).

It is concluded that CRPS may be triggered by several factors, may show recurrence, and may yield normal three-phase bone scan which is thought like crucial for the diagnosis of CRPS. It should be performed very carefully in the differential diagnosis of children considered as CRPS and the examinations cannot get ahead of clinical findings for diagnosis.

Ethics
Informed Consent: It was taken.
Peer-review: Internal peer-reviewed

Authorship Contributions
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References