Case Report

A rare cause of chronic musculoskeletal pain in children: complex regional pain syndrome

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Introduction

Complex regional pain syndrome (CRPS) is a chronic condition characterized by intense, squeezing or burning pain of a distal extremity. CRPS-I, also known as reflex sympathetic dystrophy, is without any nerve injury whereas, CRPS type II follows injury to a named nerve. The first report of CRPS in a child surfaced in 1971 [1]. Accurate data on its incidence in children is lacking. CRPS manifest as a range of motor and autonomic symptoms of the distal part of the affected limb. It causes physical impairment and, most of all, psychological distress to both the child and the family. Generally, these children are brought for medical attention repeatedly with recurring pain. However, due to lack of awareness of healthcare providers and the heterogeneity of the condition, diagnosis is often delayed [2]. Treatment becomes less effective when delayed [2,3]. Thus, it is important to disperse knowledge on CRPS among healthcare providers caring for children. This is the first published case of CRPS type I in a child in Sri Lanka who presented with debilitating unilateral upper extremity pain.

Case report

A 9-year-old girl presented with right forearm and hand pain and swelling of 6 months duration. She recalled a minor injury to the right elbow before the onset of symptoms. There had been several visits to the general practitioner and she had been treated with diclofenac sodium on one occasion with partial resolution of symptoms. The pain was a sharp, throbbing pain that increased with touch and movement. She had been missing school due to unbearable symptoms. Although there was a history of hair loss, there were no other features to suggest an autoimmune disease. One of her second-degree relative was diagnosed with systemic lupus erythematosus (SLE). She had a stable family background.

On examination, the hand and forearm were warm, red and evenly swollen. There was intense pain, disproportionate to the pain stimulus (hyperalgesia) and marked tenderness on touch (allodynia). On presentation, pain was severe, with a score of 7-8 in the Wong-Baker facial grimace scale. There were restricted movements of the right hand and forearm as adaptations
to minimize pain. Neither trophic changes of the skin nor nail dystrophy was noted. Change in perspiration was not observed in the affected area. Rest of the examination, including the nervous system, was normal.

Laboratory evaluations including haematological parameters (complete blood count, blood picture and clotting profile), biochemical parameters (liver function tests, vitamin B₁₂, creatinine kinase and serum lactate) were normal. The antistreptolysin-O levels were within normal limits and the erythrocyte sedimentation rate was 15mm/1st hour. Antinuclear factor antibody, rheumatoid factor and anti-double-stranded DNA antibody were negative. X-rays of the right hand did not show osteopenia. Cervico-thoracic spine x-ray was normal. Ultrasound scan confirmed tissue swelling, and there were no synovial thickening or joint effusions. Doppler study of the forearm vessels was normal. The 2D echocardiogram was normal. Nerve conduction studies were normal.

Our patient fulfilled the International Association for the Study of Pain (IASP) criteria for CRPS (Tabel-1) [4]. The patient had continuous pain, often disproportionate to any inciting event, hyperaesthesia to pinprick, allodynia to light touch, vasomotor signs (asymmetry in temperature and skin colour), sudomotor signs, decreased range of motion and no other diagnosis that better explained her symptoms and signs. She was initially started on ibuprofen with transient improvement. Simultaneously, cognitive behavioural therapy, including education about the condition, physiotherapy and occupational therapy, were initiated. Subsequently, amitriptyline was added to the treatment regimen. Since the response was only partial, a course of oral steroids was commenced (1mg/kg/day) and continued for one week and tailed off over two weeks. With physical therapy, cognitive therapy and steroids she became completely symptom-free and regained the full function of the affected limb.

Table 1: New International Association for the Study of Pain (IASP) clinical diagnostic criteria for CRPS [4]

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<td>1.</td>
<td>Continuing pain, which is disproportionate to any inciting event.</td>
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<td>2.</td>
<td>Must report at least one symptom in three of the four of the following categories:</td>
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<td>• Sensory: reports of hyperaesthesia and/or allodynia</td>
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<td></td>
<td>• Vasomotor: reports of temperature asymmetry and/or skin colour changes and/or skin colour asymmetry</td>
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<td></td>
<td>• Sudomotor/oedema: reports of oedema and/or sweating changes and/or sweating asymmetry</td>
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<td></td>
<td>• Motor/trophic: reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)</td>
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<td>3.</td>
<td>Must display at least one sign at time of evaluation in two or more of the following categories:</td>
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<tr>
<td></td>
<td>• Sensory: evidence of hyperalgesia (to pinprick) and or allodynia (to light touch and/or deep somatic pressure and/or joint movement)</td>
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<td></td>
<td>• Vasomotor: evidence of temperature asymmetry and/or skin colour changes and/or asymmetry</td>
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<td>4.</td>
<td>There is no other diagnosis that better explains the signs and symptoms.</td>
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Discussion

Musculoskeletal pain is a common presentation in paediatrics. Amplified musculoskeletal pain syndrome (AMPS) is a term given for pain syndromes of unknown aetiology and includes CRPS and fibromyalgia [5]. In AMPS, there is amplification of pain stimuli; mildly painful stimuli or stimuli with no pain are perceived as intense pain. This leads to functional impairment of the affected extremity to overcome pain.

CRPS is a rare entity in children and involves the distal extremities. The new IASP clinical diagnostic criteria support the diagnosis [6]. Clinical features include hyperalgesia (mildly painful stimulus is felt as intense pain), allodynia (pain sensation in response to non-painful touch), autonomic disturbances (temperature differences, change of skin colour, swelling & hyperhidrosis), trophic changes and motor disturbances (muscle weakness, dystonia, tremors or spasms of the affected limb) [7]. It is more common among adolescent girls and the mean age of diagnosis in children is around 12 years [7]. Most childhood CRPS occur following minor trauma (sprain, twist or soft tissue injury) as in our patient [6]. Some cannot recall such an event. Also, psychological stresses related to family environment and school, inflammatory and immune abnormalities and genetics, including mitochondrial disease, may play a role [7,8]. Investigations did not suggest an autoimmune process although this girl had hair loss and a family history of SLE.

Diagnosis is based mainly on clinical features and by exclusion of other possible causes [2,7]. Typically, these patients have normal laboratory investigations and imaging. However, bone imaging may show osteoporotic changes with long-standing symptoms [6]. In the absence of diagnostic testing, awareness of the condition and utilization of IASP diagnostic criteria is vital for early detection. Early treatment will prevent secondary complications such as psychological disturbances, physical disabilities and deterioration of school performance [2].

Management is through a multidisciplinary approach. The recommendations from the 2012 UK NICE guidelines include four important management strategies; patient information and education, physical and vocational rehabilitation, psychological interventions and pain relief with medications and procedures [9]. Physical rehabilitation with physiotherapy and occupational therapy has improved the outcome by reducing pain and minimizing functional limitations [10]. Studies have shown a rapid improvement in pain, abnormal skin temperature and oedema with early physiotherapy. Therefore, active physiotherapy is indicated in treating CRPS [10]. In the presence of psychological stresses, inability to cope with the disease or chronic pain behaviour, cognitive interventions with relaxation therapy and biofeedback are warranted. However, studies on the efficacy of physical therapy and psychological interventions in children are limited [10]. There are no specific pharmacological treatment recommended for paediatric CRPS. Non-steroidal anti-inflammatory drugs, antidepressants (amitriptyline), anticonvulsants (phenytoin, gabapentin) and steroids are used in the management [11,12]. Invasive treatment modalities are reserved for refractory CRPS. Our patient responded to a multidisciplinary approach along with drug treatment. In general, pediatric CRPS patients have a better prognosis than adults [6]. However, relapses are common. Median time for relapses is two months, and about 80% of the relapses occur within six months of treatment completion [13].
References


