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Clinical Note

Complex regional pain syndrome in children and adolescents

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Abstract

Complex regional pain syndrome (CRPS) is a disorder characterised by pain, sensory and motor disturbances and represents a significant medical entity. This report discusses two cases of CRPS in children and adolescents, highlighting several critical issues for clinicians in the diagnosis and management of CRPS in these populations. Early diagnosis, referral and appropriate intervention are essential in decreasing pain, suffering and resorting function for children and adolescents with CRPS.

Keywords: CRPS; RSD; Children; Adolescents; Pain

1. Introduction

Complex regional pain syndrome (CRPS) is a disorder characterised by pain, sensory and motor disturbances and represents a significant medical entity (Merskey and Bogduk, 1994; Stanton-Hicks et al., 1998). CRPS is a relatively new diagnosis designed to encompass previous pain conditions with vasomotor and sudomotor disturbances, including causalgia, Sudeck's atrophy, neuroalgodystrophy, shoulder-hand syndrome and reflex sympathetic dystrophy (RSD), the latter more commonly used (Harden et al., 1999).

The pathophysiology of CRPS is not fully understood, however, it is believed to be a systemic disease involving both the central and peripheral nervous systems (Jänig and Baron, 2003). There is limited data on the incidence of CRPS, particularly in the paediatric and adolescent populations, however, given recent reports, it would seem that the incidence is both significant and worthy of investigation in this population (Lee et al., 2002; Sherry, 1999). In recent years there has been an increase in the reporting of CRPS in children and adolescents, with the term "Reflex Neurovascular Dystrophy" commonly used for CRPS in these populations (Lee et al., 2002; Sherry, 1999; Sherry, 2000).

In this report, two cases of CRPS are discussed in the paediatric and adolescent populations, illustrating the importance of early diagnosis, appropriate referral and early intervention.

2. Case reports

2.1. Case 1

An active 14 year old female was referred to a tertiary pain centre with a 23 week history of right elbow pain following minor trauma. The patient initially presented

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to her general practitioner and subsequent radiological examination was normal. Symptoms failed to resolve with conservative management resulting in referral to a sports physician. Bone scan displayed mild to moderate hyperaemia of the affected joint, and delayed images revealed generalised uptake. These results were interpreted as bony bruising and post traumatic synovitis. MRI displayed evidence of minor capsular oedema. A provisional diagnosis of CRPS was made and a treatment regimen initiated consisting of nortriptyline 25 mg/12 mg alternating daily and passive physiotherapy. This therapeutic regimen was discontinued after 16 weeks due to lack of progress.

Initial multidisciplinary assessment revealed a rigid arm, locked in extension. Pain was localised to the elbow and was described as throbbing and sharp, particularly worse with movement and localised touch. Active and passive range of flexion was less than 5°, with evidence of oedema and colour change localised to the elbow. Hyperalgesia was noted. A reduction in fine motor skill was noted in the right hand but power was normal. Examination under anaesthesia revealed severe limitation in elbow flexion (90°) suggestive soft tissue restriction. Significant psychological and environmental factors were also noted (such as fear of pain), through questionnaires such as the TSK (Tampa Scale of Kinesiophobia) and clinical assessment. A therapeutic regimen of gabapentin (1200 mg daily) and active physiotherapy, involving a series of motor retraining and range of movement exercises was commenced. The patient then attended a 3 week group multidisciplinary pain management programme, including over 3 h per day of specific exercises. The exercise programme was embedded within a broader cognitive behavioural programme that addressed psychological and environmental factors, and followed the structure detailed in a previous randomised controlled trial (Williams et al., 1996). After the programme the patient was able to achieve 90° active flexion and had discontinued gabapentin (7 weeks after initial assessment). Subsequent follow up at 11 and 24 weeks reveals ongoing improvement in pain (numerical rating scale) and function (elbow flexion and return to pre-morbid activities). Elbow flexion at 24 weeks was measured at 105 degrees. The localised hyperalgesia, oedema and colour change were not evident on assessment at 11 and 24 weeks. The patient reported one exacerbation of her pain due to a second trauma. Signs and symptoms of CRPS returned for 1 week, however, were not present at 11 and 24 weeks.

2.2. Case 2

A 10 year old male was referred for multidisciplinary assessment with a 7 week history of pain, colour and temperature changes and swelling in his left foot. The pain was of insidious onset and no significant traumatic precipitant was able to be identified. The pain was localised to the plantar surface of the foot, and was described as sharp, made worse with light touch and weight bearing. Since the onset of pain he was unable to weight bear despite intensive hydrotherapy. Initial radiographic investigations including bone scan were unremarkable. Psychological and environmental modulating factors were also evident (e.g. beliefs about the cause of the pain).

On examination, localised oedema and associated colour and temperature changes were noted. Hyperalgesia was demonstrated on the plantar surface of the foot, and a significant reduction in both muscle bulk and power was evident. Gait was marked by adaptive strategies to reduce weight bearing through the left leg. Subsequent radiographic investigation suggested minor osteoporotic changes. Nerve conduction studies were unremarkable. An ill-defined family history of Charcot-Marie-Tooth was noted, and specialist consultation was sought, ruling this disease out.

A treatment regimen consisting of medications (gabapentin 200 mg and amitriptylline 10 mg daily) and physiotherapy was initiated. The exercise regimen was combined with basic cognitive and behavioural strategies, primarily delivered by a physiotherapist. The cognitive-behavioural strategies included education on the syndrome and behavioural modifications during stance and gait. The exercise regimen included physical desensitisation, fine motor skill retraining and lower limb weight bearing. The patient completed and upgraded this exercise regimen over 2 weeks (two one hour sessions of physiotherapy each a week apart), and on review at 7 weeks post assessment was pain free and able to walk without the use of crutches. The patient reported resolution of sweating, colour and temperature changes in the foot. At 7 weeks, he had resumed all premorbid activities and was reducing medication (gabapentin 100 mg and amitriptylline 10 mg daily). Medication ceased at 11 weeks.

3. Discussion

CRPS is a relatively new diagnosis, first published by the International Association for the Study of Pain (IASP) in 1994 (Table 1), however, the pain syndrome

Table 1
IASP diagnostic criteria for complex regional pain syndrome

1	The presence of an initiating noxious event, or cause of immobilization
2	Continuing pain, allodynia, or hyperalgesia with which the pain is disproportionate to any inciting event
3	Evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of pain
4	This diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction

has been identified in the literature for many years, including in the paediatric and adolescent populations (Berde and Lebel, 2005; Bernstein et al., 1978; Dietz et al., 1990; Merskey and Bogduk, 1994; Wilder et al., 1992). A recent paper highlights the importance in recognising the associated signs and symptoms of CRPS, although these are currently not used for diagnosis (Table 2) (Harden et al., 1999). Consideration may also be given to proposals to modify the IASP diagnostic criteria to increase diagnostic sensitivity (Harden et al., 1999; Harden and Bruehl, 2005). Utilisation of IASP diagnostic criteria and awareness of the associated signs and symptoms is of paramount importance in early diagnosis and referral of CRPS patients, particularly children and adolescents given the impact on long-term management.

Although the clinical data on CRPS in children and adolescents is limited, the benefit of early mobilisation and intense physiotherapy is evident. Sherry and colleagues followed 103 children through an intensive exercise program (over 4 h per day for a mean of 14 days) delivered at a specialist centre, reporting initial resolution of symptoms in 92% of children with 88% symptom free at 2 years (Sherry, 1999). Lee and colleagues performed a smaller study using an exercise and cognitive-behavioural therapy approach, reporting pain reduction and functional restoration at short and long-term follow-up (Lee et al., 2002).

The two cases presented in this report highlight the importance of early diagnosis, mobilisation and referral to a specialist pain service. The severity of disability as a result of delayed diagnosis and referral is dramatically represented by the findings of the examination under anaesthesia performed on the female patient. Unfortunately in this case, an early diagnosis was not made, and management was heavily focused on passive therapy rather than active mobilisation and pain management under specialist supervision. Subsequent severe maladaptive musculoskeletal changes ensued, with significant joint restrictions and longer-term motor dysfunction. These maladaptive changes resulted in increased disability, suffering and posed significant barriers to management.

Case 2, however, is an example of early diagnosis, early mobilisation and referral to a specialist pain service. Underlying neurological conditions were quickly excluded by specialist input. Maladaptive musculoskeletal changes although present, were minimal at this stage, however, this patient was fortunate given the possible consequences of long-term non-weight bearing and lack of joint movement. A structured exercise regimen and pain management was introduced, resulting in alleviation of symptoms and restoration of function.

A recent epidemiological study of CRPS in the adult population found that patients had seen on average 4.8 different physicians before referral to a pain centre or clinic, and 47% had a history of physician-imposed immobilisation, despite recognition that immobilisation may strengthen and maintain the CNS abnormalities in CRPS (Allen et al., 1999; Jänig and Baron, 2004). Whilst there is a lack of epidemiological data in the paediatric and adolescent populations, these two cases highlight the importance of early mobilisation once exclusion of other pathology is ruled out by appropriate consultations and investigations.

Early diagnosis using the IASP criteria and awareness of the associated signs and symptoms, mobilisation and referral to a pain centre will greatly assist in reducing musculoskeletal changes and the subsequent impact on management, hopefully decreasing pain, suffering and restoring function in children and adolescents with CRPS.

Table 2
Additional signs and symptoms and listed in the IASP taxonomy however not used for diagnosis

1	Atrophy of the hair, nails, and other soft tissues
2	Alterations in hair growth
3	Loss of joint mobility
4	Impairment of motor function, including weakness, tremor, and dystonia
5	Sympathetically-maintained pain may be present

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