

Clinical Rehabilitation

<http://cre.sagepub.com/>

A 2½-year-old girl with reflex sympathetic dystrophy syndrome (CRPS type I): case report

F Güler-Uysal, S Basaran, J HB Geertzen and K Göncü

Clin Rehabil 2003 17: 224

DOI: 10.1191/0269215503cr589oa

The online version of this article can be found at:

<http://cre.sagepub.com/content/17/2/224>

Published by:



<http://www.sagepublications.com>

Additional services and information for *Clinical Rehabilitation* can be found at:

Email Alerts: <http://cre.sagepub.com/cgi/alerts>

Subscriptions: <http://cre.sagepub.com/subscriptions>

Reprints: <http://www.sagepub.com/journalsReprints.nav>

Permissions: <http://www.sagepub.com/journalsPermissions.nav>

Citations: <http://cre.sagepub.com/content/17/2/224.refs.html>

A 2½-year-old girl with reflex sympathetic dystrophy syndrome (CRPS type I): case report

F Güler-Uysal, S Başaran Çukurova University, School of Medicine, Department of Physical Medicine and Rehabilitation, Adana, Turkey, **JHB Geertzen** Groningen University Hospital, Department of Rehabilitation, The Netherlands and **K Göncü** Çukurova University, School of Medicine, Department of Physical Medicine and Rehabilitation, Adana, Turkey

Received 23rd August 2001; returned for revisions 28th August 2001; manuscript accepted 4th October 2001.

An unusual case of a 2½-year-old girl with reflex sympathetic dystrophy (RSD) of the left arm is described. She is the youngest RSD case ever presented in the literature. Upper extremity involvement is also rare in childhood RSD. She had both physical and psychological trauma in an earthquake preceding the disease. The association of RSD with a psychological disorder is stressed and awareness of the condition to the general paediatrician is recommended for early diagnosis and successful treatment.

Introduction

Reflex sympathetic dystrophy (RSD), also called complex regional pain syndrome (CRPS) type I,¹ is a syndrome characterized by pain in one or more extremities, usually associated with vasomotor and sudomotor changes.² The predominant and most disabling feature of RSD is chronic and burning pain exacerbated by movement and emotional stress.³ Many diseases, precipitating events or drugs have been associated with RSD, but trauma, especially fractures, has been shown to be the most common initiating factor in adults.⁴ Opinions have varied widely on the issue of a psychological aetiology.^{5,6} Stressful life events besides trauma or surgery have also been shown to be risk factors in RSD.⁷ There are, however, a number of cases for which no precipitating factor can be identified.

The occurrence of RSD in childhood has long been thought to be rare,^{2,8–10} but it may be more common in children than previously thought.^{2,9–13} RSD could still be underestimated and misdiagnosed in the young paediatric age group. Delays in diagnosis averaged 12 months in a large series.⁹ Unnecessary testing and referrals may create a vicious circle, perpetuating signs and symptoms in young children. It may also be confused with psychiatric conditions such as conversion reaction and malingering.¹²

Children with RSD are usually of school age or in the teenage years.^{3,9} There is a preponderance of girls and also a marked tendency for lower extremity involvement.^{4,9,13,14} We therefore present this rare case of a 2½-year-old girl with RSD of the upper extremity as being the youngest RSD case ever presented in the literature.

Address for correspondence: Füsün Güler-Uysal, Çukurova Üniversitesi Tıp Fakültesi, Fiziksel Tıp ve Rehabilitasyon Anabilim Dalı, 01330 Balcalı, Adana, Turkey. e-mail: fusunu@cu.edu.tr

Case

A 2½-year-old girl was presented to the Physical Medicine and Rehabilitation outpatient clinic by her relatives, declaring that she could not use her left hand and resisted even the touch of anyone who attempted to examine it for the last three months. The patient had experienced the Marmara earthquake in Turkey in August 1999 and had been rescued after 12 hours of being trapped under the ruins. Her parents had died during the same disaster and she was being followed by the Paediatric Psychiatry Clinic with diagnoses of non-organic failure to thrive and acute stress disorder. An orthopaedic consultation was assessed soon after she had been rescued and no fracture nor soft tissue damage of the musculoskeletal system could be detected. The left hand pain was thought to be due to a minor compression of the upper extremity exaggerated by her psychiatric situation until she was referred to our clinic for a second opinion three months later. On admission, her relatives declared that she could even be disturbed by the touch of the bed sheets to her hand during sleep. This resembled allodynia, which is a common symptom of RSD.⁹ Dystrophic nail changes were observed on inspection of the extremity. A slight atrophy of the hand and forearm with traces of hypertrichosis was observed. She had severe pain and slight limitation of passive flexion of the fingers. Control radiographs of both extremities revealed prominent osteopenia of the carpal and metacarpal bones of the involved hand.

The diagnosis of RSD was confirmed and anti-inflammatory medication (nimesulide 50 mg b.i.d.) along with intranasal calcitonin (50 IU q.o.d.) and vitamin D supplementation (400 IU g.d.) were started. She was also taking anxiolytic medication (tizanidine) for her psychiatric condition. The patient started a rehabilitation programme of passive range of motion exercises in the beginning, followed by occupational therapy to the left hand which diminished the pain and resulted in a dramatic recovery of left hand function within three months. At six months after trauma, she was able to use her left hand for eating and playing again. She had left the guarding posture and cooperated well with the examination of left

hand function, which revealed slight limitation on passive flexion of interphalangeal joints as well as dystrophic nail and skin changes. One year after the initiating event, she only had mild coldness and minimal hypertrichosis of the left hand but no pain or limitation in joint mobility and hand function.

Discussion

Children with RSD are usually of school age or in their adolescence and a marked tendency for lower extremities has been described.^{10,11} The very young age of our case as well as the involvement of the upper extremity is remarkably unusual. She presented severe psychological problems due to loss of both parents and experiencing hours under the ruins after the earthquake. She cried continuously during her first visits to our clinic. The diagnosis was reached after a detailed interview with her aunt, a thorough physical examination and radiographic investigation excluding fractures. The diagnosis of RSD is mainly based on clinical grounds. Radiography can be supplementary, showing osteopenia of the affected extremity, but bone scan results can be controversial.^{2,13,15}

Half of the children with RSD have minor trauma or illness identified as an initiating factor.^{6,11} Significant emotional dysfunction is also found in the majority.¹¹ The analysis of 21 families of children with RSD has revealed that RSD is frequently related to stress.⁶ Our case is a good demonstration of both factors playing a role in the aetiology of RSD.

The prognosis of RSD in children has been

Clinical messages

- Reflex sympathetic dystrophy could easily be misdiagnosed in the very young paediatric age group.
- Unnecessary testing and referrals may create a vicious circle in children and should be avoided.
- Early and correct diagnosis depends on the awareness of the condition by clinicians.

reported to be more benign than in adults,^{2,15,16} as in our case who demonstrated an excellent outcome. Yet cases with severe residual dysfunction have also been described.⁹ Optimal treatment is still under debate. Literature data suggest that once early diagnosis is established, conservative therapy (mainly mobilization and physical therapy along with psychotherapy when needed) can be sufficient.^{2,9,14} RSD in children can be a self-limiting condition which responds well to mild analgesics and physical therapy.¹⁶ Less conventional pharmacologic treatments, such as tricyclic antidepressants, calcitonin, bisphosphonates, alpha- and beta-adrenergic antagonists, anticonvulsants, calcium channel blockers and steroids as well as sympathetic blocks have also been used.^{9,11} Nonresponders are usually patients with a delayed diagnosis or those with severe psychological problems.²

It has recently been suggested that childhood RSD is completely different from 'true' RSD and should be referred to as pseudodystrophy or disuse-related dystrophy.^{17,18} This rather mild form of RSD may also be seen in adults. According to Herregods *et al.*¹⁷ and Driessens,¹⁸ bone scan is of major importance in differential diagnosis for it is always negative in pseudodystrophy. Yet there are reports which support the lack of utility of a bone scan in the diagnostic work-up of RSD.^{15,19}

In conclusion, we describe this rare case of a very young child with RSD of her left arm in order to remind clinicians of this entity even in toddler years. The diagnosis of RSD in this very young age group undoubtedly depends on the awareness of the condition by clinicians. It is suggested that the syndrome should be suspected whenever the severity of the pain does not fit the pattern of injury and the pain is associated with extreme sensitivity to light touch (allodynia), episodic mottling, cyanotic discoloration of the skin, or swelling, or a difference in the skin temperature compared with the contralateral extremity.

References

- Merksey H, Bogduk N. *Classification of chronic pain: descriptions of chronic pain syndromes and definitions of terms*. Seattle: IASP Press, 1994.
- Cimaz R, Matucci-Cerinic M, Zulian F, Falcini F. Reflex sympathetic dystrophy in children. *J Child Neurol* 1999; **14**: 363–67.
- Cassidy JT, Petty RE. Musculoskeletal pain syndromes of nonrheumatic origin. In: Cassidy JT, Petty RE eds. *Textbook of paediatric rheumatology*. New York: WB Saunders, 1995: 108–32.
- Kozin F. Painful shoulder and the reflex sympathetic dystrophy syndrome. In: Koopman WJ ed. *Arthritis and allied conditions*. Baltimore: Williams & Wilkins, 1997: 1887–922.
- Lynch ME. Psychological aspects of reflex sympathetic dystrophy: a review of the adult and paediatric literature. *Pain* 1992; **49**: 337–47.
- Sherry DD, Weisman R. Psychological aspects of childhood reflex neurovascular dystrophy. *Pediatrics* 1998; **81**: 572–78.
- Geertzen JH, de Bruijn-Kofman AT, de Bruijn H, van de Wiel HB, Dijkstra PU. Stressful life events and psychological dysfunction in Complex Regional Pain Syndrome type I. *Clin J Pain* 1998; **14**: 143–47.
- Fermaglich DR. Reflex sympathetic dystrophy in children. *Pediatrics* 1977; **60**: 881–83.
- Wilder RT, Berde CB, Wolohan M, Vieyra MA, Masek BJ, Micheli LJ. Reflex sympathetic dystrophy in children. *J Bone Joint Surg* 1992; **74 A**: 910–19.
- Lemahieu RA, Van Laere C, Verbruggen LA. Reflex sympathetic dystrophy: an underreported syndrome in children? *Eur J Pediatr* 1988; **147**: 47–50.
- Barbier O, Allington N, Romboutts JJ. Reflex sympathetic dystrophy in children: review of a clinical series and description of the particularities in children. *Acta Orthop Belg* 1999; **65**: 91–97.
- Silber TJ, Majd M. Reflex sympathetic dystrophy syndrome in children and adolescents. Report of 18 cases and review of the literature. *Am J Dis Child* 1988; **142**: 1325–30.
- Stanton RP, Malcolm JR, Wesdock KA, Singsen BH. Reflex sympathetic dystrophy in children: an orthopedic perspective. *Orthopedics* 1993; **16**: 773–79.
- Dietz FR, Mathews KD, Montgomery WJ. Reflex sympathetic dystrophy in children. *Clin Orthop* 1990; **258**: 225–31.
- Ashwal S, Tomasi L, Neumann M, Schneider S. Reflex sympathetic dystrophy syndrome in children. *Pediatr Neurol* 1988; **4**: 38–42.
- Ruggeri SB, Athreya BH, Doughty R, Gregg JR, Das MM. Reflex sympathetic dystrophy in children. *Clin Orthop* 1982; **163**: 225–30.
- Herregods P, Willems J, Chappel R. Pseudodystrophy at the lower limb in children. *Clin Rheumatol* 1997; **16**: 425–28.

- 18 Driessens M, Dijs H, Verheyen G, Blockx P. What is reflex sympathetic dystrophy? *Acta Orthop Belg* 1999; **65**: 202–17.
- 19 Allen G, Galer BS, Schwartz L. Epidemiology of complex regional pain syndrome: a retrospective chart review of 134 patients. *Pain* 1999; **80**: 539–44.