



Reflex sympathetic dystrophy

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Abstract

Objective: To describe eight patients with reflex sympathetic dystrophy in terms of clinical and laboratory characteristics and treatment.

Description: Eight children (four girls) with reflex sympathetic dystrophy were retrospectively analyzed. The diagnosis of reflex sympathetic dystrophy was based on the presence of pain in the distal extremities, local edema, vasomotor instability and impairment of sensibility. Two patients had associated systemic lupus erythematosus, one had juvenile idiopathic arthritis and one had Glanzmann's thrombasthenia. Mean age was 11.5 years. Most of the patients had lower extremity involvement (7/8). The most important clinical signs were pain, edema and vasomotor instability in the affected extremity (8/8), functional impairment (7/8), and impaired sensibility (3/7). The erythrocyte sedimentation rate was abnormal in three patients and the bone scans in five. All patients received non-steroidal anti-inflammatory drugs and physical therapy with improvement of the symptoms in seven patients, until six months of treatment. Three patients were submitted to acupuncture with good response. One patient had a severe disease and received tricyclic antidepressants, with improvement more than one year after.

Comments: Reflex sympathetic dystrophy should be included as part of the differential diagnosis of limb pains of childhood, so that physicians can make an earlier diagnosis and prevent functional impairment.

J Pediatr (Rio J). 2004;80(2):159-62: Reflex sympathetic dystrophy, pain, edema.

Introduction

Reflex sympathetic dystrophy (RSD) was first described by Mitchell in 1864, during the American civil war, as a syndrome in which, after gunshot wounding, an extremity developed edema accompanied by vasomotor and trophic abnormalities.¹ Since then, this disease has been termed in a number of different ways, such as :algodystrophy, causalgia, Sudeck's atrophy, hand-shoulder syndrome, neuroalgodystrophy, post-traumatic sympathetic dystrophy or complex regional pain syndrome type 1.²

Clinically, RSD most often presents as persistent pain of great intensity in one extremity, generally out of proportion to the triggering event. Pain is associated with neuropathic pain descriptors (burning, dysesthesia, paresthesia, allodynia and hyperalgesia to cold) and clinical signs of autonomic dysfunction (cyanosis, edema, cold and localized transpiration and hair growth abnormalities).³

There is currently great controversy over the pathogenesis of RSD. Some authors believe that the disease is the result of a post-traumatic reflexive neuronal mechanism which leads to abnormal pain perception and exacerbated efferent sympathetic activity.

Innumerable conditions are associated with the development of RSD. In more than 60% of cases described in adults there is a history of trauma. Reflex sympathetic dystrophy in childhood is both rare and under-diagnosed, while the history of trauma is less common and, when present, usually of lesser intensity.³ There are no studies of incidence and prevalence among the pediatric age group.

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Reflex sympathetic dystrophy also occurs more often among adults who are emotionally unstable, manic, depressive and insecure. Children present a peculiar profile being generally perfectionist and hard working, and the syndrome can be preceded or aggravated by stress factors such as family conflict or the death of family members, starting school, etc.⁴ The RSD symptoms are very often associated with other pathologies involving autonomic dysfunction: migraine, syncope and abdominal pains.⁵

The objective of this study is to describe the clinical, laboratory and treatment characteristics of eight children with RSD in order to throw light on the profile of the disease in childhood, since late treatment can lead to significant functional impairment.

Case reports

Between 1992 and 2002 eight patients with RSD were treated at the Instituto da Criança Pediatric Rheumatology Unit at the Hospital das Clínicas of the Medical Faculty of the Universidade de São Paulo.

The age of onset of the disease varied between 8 and 13 years (mean 11.5 years and median 12 years) and affected the two sexes equally. Four patients exhibited associated conditions: two had systemic lupus erythematosus (SLE), one had juvenile idiopathic arthritis (JIA) and one was suffering from Glanzmann's thrombasthenia.

The diagnosis of RSD was based on the presence of prolonged, intense pain in the distal segment of a limb, frequently associated with a diffuse edema of the area, changes in color and temperature, abnormal sensitivity and functional incapacity.

The patients' clinical and laboratory characteristics are listed in Table 1.

It is important to point out that a majority of the patients had already sought help from other professionals before the diagnosis of RSD was made. This diagnosis was established an average of 8.8 months after onset of symptoms (varying from 2 months to 2 years). Six patients had had the affected limb immobilized with plaster and splints, more than once, without improving the condition. One patient had had corticosteroids injected into the affected area, but the symptoms persisted (case 8).

The lower limb involvement occurred in seven patients and was associated with significant functional incapacitation and difficulty walking (two of these patients required wheelchairs in order to move around). They presented high intensity pain that was alleviated by rest and associated with diffuse edema of the foot and ankle. Three patients mentioned prior traumas: two sprains and one fracture of the ankle. Only one patient exhibited no sign of vasomotor instability, which was manifest as increased temperature in two affected limbs, reduced temperature in one, fixed erythema in two, cyanosis and diaphoresis in one. Three patients also presented numbness and paresthesia of the affected area.

Table 1 - Clinical, laboratory and therapeutic characteristics of eight patients with reflex sympathetic dystrophy

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8
Age of onset	10 y	13 y	13 y	11 y	13 y	13 y	11 y	8 y
Pain	+	+	+	+	+	+	+	+
Edema	+	+	+	+	+	+	+	+
Abnormal sensitivity	+	+	-	+	-	-	-	-
Vasomotor instability	+	+	+	+	+	+	+	-
Trophic abnormalities	-	-	-	+	-	+	+	+
Location	III	III	ulr/l	llr	llr	llr	llr	llr
Functional impairment	+	+	-	+	+	+	+	+
Associated disease	SLE	SLE	-	JIA	-	GT	-	-
Elevated BSR	+	+	-	+	-	-	-	-
Abnormal BS	-	+	+	+	+	-	-	+
Treatment	NHAI PT AD	NHAI PT	NHAI PT A	NHAI PT P	NHAI PT O	NHAI PT A	NHAI PT A O	NHAI PT

+ = present; - = absent; ll= lower limb; ul = upper limb; r = right; l = left; SLE = systemic lupus erythematosus; JIA = juvenile idiopathic arthritis; GT = Glanzmann thrombasthenia; BSR = blood sedimentation rate; BS = bone scintigraphy; NHAI = nonhormonal anti-inflammatory agent; PT = physical therapy; A = acupuncture; AD = antidepressive; O = opiates; P = psychotherapy.

Trophic alterations, such as atrophy of adjacent musculature and reduction in hair and nails were discovered in four of the seven patients with lower limb involvement. One of these presented a difference of 2.5 cm between one lower limb and the other (case 6).

The only patient who had an affected upper limb (case 3) began with imprecisely limited edema, heat and pain in the right hand, forearm and elbow not associated with a local trauma, improving after five months of treatment. This patient then suffered two relapses: the first was a similar condition affecting the contralateral limb and the second, six years later, returned to the upper, right-side limb after having carried weight.

Reflex sympathetic dystrophy diagnosis was eminently clinical, but laboratory tests were also performed for all patients. Erythrocyte sedimentation rates were elevated in three patients, all suffering from underlying diseases (two had LES and one JIA). Musculoskeletal scintigraphy using technetium revealed abnormalities in five cases, there was hyperdeposition in three cases and hyperabsorption in two. Standard radiography found abnormalities in just one patient (case 1) who had chronic arthritis prior to RSD, revealing subchondral sclerosis and rarefied bone which were also observed with computerized tomography.

One patient had exhibited such significant pain at the start of the condition that, at another service, spinal tap and electromyography were performed – both with normal results.

As soon as diagnosis was confirmed, all patients received non-hormonal anti-inflammatory drugs (NHA1) (acetylsalicylic acid, indomethacin, ibuprofen or naproxen) associated with physiotherapy sessions. Three patients received acupuncture in conjunction with the treatments described above, with obvious improvements after four sessions.

The patient whose clinical course was most chronic and who was most incapacitated (case 8) required tricyclic antidepressants (amitriptyline), improving after a year of treatment when walking was achieved without the aid of crutches. A further four patients exhibited improvement in symptoms in an average of five months after treatment was started. One patient attended psychotherapy sessions for associated depression.

Discussion

Reflex sympathetic dystrophy is both rare and under diagnosed during childhood, particularly in our country, rarely being recognized by pediatrician. The majority of studies to date have described adult population. Veldman *et al.*⁶ studied 829 patients with the disease among which there was just one patient less than 9 years old. In our sample, the average age of patients was 11.5 years, which is compatible with published data,⁷⁻¹⁰ but the predominance of the female sex which occurs during early childhood was not observed.^{7,8,11,12}

The majority of patients involved in this study had received inappropriate diagnoses and treatment before seeking our service. Murray *et al.*³ assessed 46 children with RSD observing that the average number of professionals consulted before correct diagnosis was 2.3 (varying from one to five) and that the time between onset of symptoms and diagnosis was almost six months.

Reflex sympathetic dystrophy should be suspected when a child presents continuous pain and burning sensations and refuses to move a limb, associated with imprecisely limited edema, and varying degrees of pallor, hyperthermia, hypothermia or hyperesthesia.² Bernstein *et al.*⁸ studied 23 children with RSD, whose most common signs and symptoms were: pain in the affected limb in 100% of cases, sensitivity abnormalities in 91%, edema of the extremity in 82% and temperature changes in 78%. Trophic abnormalities were not found.

In our study only one patient had upper members compromised. Reflex sympathetic dystrophy generally affects lower limbs and is rarely bilateral.^{3,6,8-10} Wilder *et al.*⁷ observed 70 children with RSD and found that 87% of episodes occurred in lower limbs and that 31% of the children exhibited events in more than one location.

Within the pediatric age group it is less likely that there is a precipitating event, such as a trauma^{8,11-13} and the incidence of trophic abnormalities is also rarer than in adults.^{8,11,13,14} This rarity during infancy may be due to the shorter duration of the disease or immobility.⁸

Routine x-rays of the affected limbs of children are generally normal,¹¹⁻¹³ in contrast with the adult population in which 50% of cases exhibit osteoporosis,² and cortical erosion and reactive bony neoformation can occur. In our sample just one patient presented cortical erosion associated with JIA, manifesting prior to the appearance of RSD.

In children, musculoskeletal mapping is a useful auxiliary examination for RSD diagnosis, presenting better sensitivity than radiography (72% versus 36%). In a study of 11 children, mapping revealed hyperdeposition in four cases, hyperabsorption in four and was normal for three, suggesting, as in our sample, that the first two conditions occur with similar frequency.¹¹ Computerized tomography or nuclear magnetic resonance do not help in RSD diagnosis, frequently returning normal results or finding non-specific soft body abnormalities.⁵

Reflex sympathetic dystrophy is not accompanied by blood test abnormalities or acute phase tests.^{6,12} In this study, the three patients who exhibited increased ESR also presented associated inflammatory diseases.

In common with published data,^{3,10} our study found that a significant number of patients received inappropriate therapies before an RSD diagnosis was confirmed. Six of our patients had had limbs immobilized, which intensified their pain.

Reflex sympathetic dystrophy treatment is primarily based on physiotherapy and on the alleviation of

pain.^{7-9,12} Two studies^{7,15} evaluated children with RSD, employing physiotherapy, TENS (transcutaneous electrical nerve stimulation) and psychotherapy, almost always associated with NHA1. This treatment was effective for around 70% of the patients, with significant functional improvements. In the most resistant cases⁷ tricyclic antidepressants and sympathetic blockades were employed. These last were performed for 53% of the patients, with a temporary reduction in pain in just 46% of the cases in which they were used. Due to the risks and potential side effects of steroids we suggest that they not be used for DSR.^{7,14}

In our study, with the exception of one patient, substantial improvement was achieved in six months of treatment based on physiotherapy and NHA1, with no need for sympathetic blockades or TENS. Acupuncture has been employed as an excellent supporting therapy. In published literature, the majority of cases described show substantial improvement in six to eight weeks.^{3,10}

The psychological approach is very important to RSD treatment. Murray *et al.*³ observed that psychological factors may have contributed to the disease in 25% of cases.

Families should be informed that while prognosis is generally favorable, between 25% and 33% may suffer recurrences in the same location or in another area.^{3,5,9}

In conclusion, it should be emphasized that RSD is a chronic, painful disease associated with significant morbidity in children and adolescents and may cause temporary or permanent functional incapacitation. Pediatricians should be alert, since diagnosis is eminently clinical and, when made early, it can prevent undesirable treatment and investigations, which, without doubt, will exacerbate and prolong the condition.

References

- Mitchell SW, Morehouse GR, Keen WW. Gunshot wounds and other injuries of nerves. Philadelphia: Lippincott; 1864.
- Fermaglich D. Reflex sympathetic dystrophy in children. *Pediatrics*. 1977;60:881-3.
- Murray CS, Cohen A, Perkins T, Davidson JE, Stills JA. Morbidity in reflex sympathetic dystrophy. *Arch Dis Child*. 2000;82:231-3.
- Sherry DD, Weisman R. Psychological aspects of childhood reflex neurovascular dystrophy. *Pediatrics*. 1988;81:572-8.
- Small E. Chronic musculoskeletal pain in young athletes. *Pediatr Clin N Am*. 2002;49:655-62.
- Veldman PHJM, Reynen HM, Arntz IE, Goris RJA. Signs and symptoms of reflex sympathetic dystrophy: prospective study of 829 patients. *Lancet*. 1993;342:1012-5.
- Wilder RT, Berde CB, Wolohan M, Vieyra MA, Masek BJ, Micheli LJ. Reflex sympathetic dystrophy in children. *J Bone J Surg*. 1992;74:910-9.
- Bernstein BH, Singesen BH, Kent JT, Kornevitch H, King K, Hicks R, *et al.* Reflex neurovascular dystrophy in childhood. *J Pediatr*. 1978;93:211-5.
- Sherry DD, Wallace CA, Kelley C, Kidder M, Sapp L. Short and long term outcomes in children with complex regional pain syndrome type 1 treated with exercise therapy. *Clin J Pain*. 1999;15:218-23.
- Lee BH, Scarff L, Sethna NF, McCarthy CF, Scott-Sutherland J, Shea LM, *et al.* Physical therapy and cognitive-behavioral treatment for complex regional pain syndromes. *J Pediatr*. 2002;141:135-40.
- Laxer RM, Allen RC, Malleon PN, Morrison RT, Petty RE. Technetium 99m-methylene diphosphonate bone scans in children with reflex neurovascular dystrophy. *J Pediatr*. 1985;106(3):437-40.
- Kesler RW, Saulsbury LM, Rowlingson JC. Reflex sympathetic dystrophy in children: treatment with transcutaneous electric nerve stimulation. *Pediatrics*. 1988;82:728-32.
- Kavanagh R, Crisp AJ, Hazleman BL. Reflex sympathetic dystrophy in children: dystrophic changes are less likely. *BMJ*. 1995;310:1648-9.
- Ruggeri SB, Athreya BH, Daugherty R, Gregg JR, Das MM. Reflex sympathetic dystrophy in children. *Clin Orthop Rel Res*. 1982;163:225-30.
- Stanton RP, Malcom JR, Wesdock KA, Singesen BH. Reflex sympathetic dystrophy in children: an orthopedic perspective. *Orthopedic*. 1993;16:773-80.

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