CASE STUDY

Recurrent and migratory reflex sympathetic dystrophy in children

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Summary

Reflex sympathetic dystrophy is a syndrome characterized by superficial pain and tenderness associated with swelling, vasomotor instability, and dystrophic changes of the skin. In children, it is rarely reported and is felt to have a more benign and self-limited course. This case illustrates that, in children, reflex sympathetic dystrophy can occur without any previous history of trauma, and may be recurrent and migratory. A review of the literature is included. An 11-year-old girl, with no history of trauma, presented in 1992 with spontaneous onset of right leg pain. She was diagnosed with reflex sympathetic dystrophy, and she was treated unsuccessfully with oral medications. Her symptoms then resolved in 2 weeks after receiving epidural anaesthesia and aggressive physical therapy. Over the next 5 years, she presented to the paediatric rehabilitation clinic three times with recurrent RSD in her bilateral arms. The first two times were refractory to conservative management and resolved with four stellate ganglion blocks. The third recurrence persisted with three stellate ganglion blocks and resolved with gabapentin.

Introduction

Reflex sympathetic dystrophy (RSD) is a syndrome characterized by superficial pain and tenderness associated with swelling, vasomotor instability, and dystrophic changes of the skin. Although many cases of reflex sympathetic dystrophy have been reported in adults since the 1800s, there are few cases reported in children, and fewer cases of recurrent sympathetic dystrophy, reported in the literature. In children, it is felt to have a more benign and self-limited course. The following is a case that is not only recurrent, but slowly progressive in its severity.

Case study

An 11-year-old girl, with no history of trauma, presented to a children’s hospital in 1992 with spontaneous onset of right leg pain. She had a past medical history of sinus infections, acne, and allergies to cats and mould. She had sinus surgery in 1990 and a tonsillectomy in 1992. Medication taken at that time was only allergy shots every week. She had no known drug allergies. Her sister had a seizure disorder, but there was no family history of muscle or nerve disease. She was an elementary school student and did not drink alcohol or smoke.

Physical examination of her head, neck, heart, lungs, and abdomen was normal. Strength testing, passive range of motion, and tone were normal in all four limbs. Her right leg had mild oedema with rubor, warmth, and dysesthesia, but no atrophy.

A full work-up including x-rays and bone scan did not reveal a definitive diagnosis. After 2 months of unsuccessful treatments, a diagnosis of reflex sympathetic dystrophy (CRPS type I—complex regional pain syndrome type I) was made, and she was treated unsuccessfully with oral medications. She then underwent epidural anaesthesia with aggressive physical therapy, and her pain and swelling resolved within 2 weeks.

Three years later, she presented to the paediatric rehabilitation clinic after spontaneous onset of bilateral arm swelling, pain, and erythema, which had lasted 6 weeks. This persisted for another 6 weeks despite treatment with amitriptyline, compression gloves, desensitization, massage, and oedema mobilization. She then underwent four stellate ganglion blocks, which completely resolved her pain and swelling within 4 weeks.
Ten months later, she presented again with acute onset of pain, swelling, redness, and heat in both hands (R > L) for 24 hours duration. Given the difficulty of controlling her problem in the past, she was immediately referred for a right stellate ganglion block, which helped, but she continued to have pain in her arms (L > R). She then underwent a left stellate ganglion block 2 days later. Her pain completely resolved over the next 2 weeks.

One year later, she presented again with similar symptoms in both hands. She underwent a right stellate ganglion block. She had partial relief on the right side and was started on gabapentin 100 mg QD. The pain worsened over 1 week and she underwent two more stellate ganglion blocks in 1 week with less relief and the gabapentin was increased to 300 mg QHS. Two weeks later, she underwent another block with minimal relief, and the gabapentin was increased to 300 mg TID, and she resumed using the compression gloves. Her symptoms then slowly resolved. Since then, she has continued using the gabapentin for 1 year after the symptoms resolved, and has not had a recurrent episode.

Discussion

Since it was first reported in 1864 as causalgia [1], reflex sympathetic dystrophy (RSD), coined by Evans [2], has been called by many names. In 1993, it was renamed complex regional pain syndrome type I (CRPS type I) by the International Association for the Study of Pain in a consensus workshop [3]. Although there are many cases reported in adults, there are fewer cases reported in the paediatric literature. Before 1978, only a few cases of RSD had been reported in children. Bernstein et al. [4] reported 24 cases in 1978, and Ruggeri et al. [5] reported six cases in 1978. Most of the cases were self-limited or mild. However, there is little data on whether there is recurrence or if the symptoms can migrate. This case illustrates that CRPS type I can be recurrent and migratory in children. This phenomenology of presentation is supported by only one study that the authors could find in the current literature.

In 1988, Greipp et al. [6] reported on 27 children and young adults who were registered with the RSDSA (Reflex Sympathetic Dystrophy Syndrome Association). In this study, 55% had both extension and migration of their symptoms. Twenty-six of 27 subjects failed to receive ‘complete and permanent resolution’ of RSD. However, their patient population may not be representative of the natural course of paediatric RSD, since the subjects were selected from an association membership list, the study was retrospective, and it included subjects who were young adults (up to 19 years old).

In the literature search, there were no other studies which looked at the recurrence of RSD in children, but there was a similar case reported by Rush et al. [7] in 1985, where a child had several episodes of RSD affecting the arms and legs.

When patients present with symptoms of superficial pain and tenderness associated with swelling, vasomotor instability, and dystrophic changes of the skin, the diagnosis of RSD is made clinically. There is no definitive clinical diagnostic test for RSD. However, there are a minimum of three International Association for the Study of Pain diagnostic criteria for CRPS type I, all of which can be established from the clinical examination and history [8]: (1) The patient must have persistent pain disproportionate to injury, allodynia, or hyperalgesia; (2) At some point in the disease there must be a clinical sign of sympathetic abnormality. This sign or symptom can be an objective or subjective cold or warm limb, red or cyanotic skin, hyperhidrosis, or hypoalgesia, or oedema; and (3) There can not be any other diagnosis that would otherwise account for the symptoms. The case presented met the first two criteria by clinical observation, since she had presented with allodynia, swelling and warmth in the affected limb.

The third criteria stipulates that, for every patient who presents with a possible diagnosis of RSD, a reasonable differential diagnosis list be made and evaluated as a possible cause of the symptoms. For the case presented, the differential diagnosis on initial presentation included a fracture (gross or occult) in the leg, cellulitis, local arthritis, and local inflammatory disease/autoimmune disease. The x-ray and bone scan were adequate to evaluate for fractures (gross or occult), arthritis, and local inflammatory disease. Cellulitis was evaluated for clinically by the history, physical examination, and checking her temperature. She was clinically diagnosed with RSD after these other causes were ruled out. Her diagnosis was confirmed by her response to epidural anaesthesia in the first episode, and by her response to sympathetic ganglion blocks with the last three episodes.

The patient’s treatment regimen for the first two episodes follows what is generally recommended for treatment of RSD. Physical therapy (including desensitization, massage, and heat with stretching), compression garments, and oral medications (including amitriptyline or gabapentin) were tried. Even though these approaches are widely used, the physical modalities have not been adequately studied for their efficacy,
because the treatments are hard to blind. Amitriptyline and gabapentin have been shown in randomized blinded controlled studies to help with neuropathic pain [9–12]. However, they have not been specifically studied with respect to RSD pain. If needed, sympathetic ganglion blocks are then used.

For the third and fourth episode, since the patient needed to have an epidural or sympathetic ganglion block in the past two episodes, she was immediately referred to the pain clinic for stellate ganglion blocks. During the fourth episode, gabapentin was continued after the stellate ganglion block procedures, since she only achieved partial relief with the procedures. There is no data on the use of gabapentin in RSD after an episode has resolved, but, since the patient had three episodes within 3 years, the gabapentin was continued for 1 year before it was tapered off. Since then, the patient has not had a recurrent episode.

The use of gabapentin in this patient for neuropathic pain was a novel idea at that time. It has long been known that anti-epileptic drugs such as carbamazepine can be helpful as adjunctive drugs for treating chronic neuropathic pain. Because of this, gabapentin was tried on neuropathic pain associated with post-herpetic neuralgia and diabetic neuropathy. At the time this medication was used in this patient, the results of the first randomized controlled trials showing gabapentin to be helpful in post-herpetic neuralgia and painful diabetic neuropathy were just being reported [11]. Even though gabapentin is a structural analogue of gamma-aminobutyric acid (GABA), it does not bind to GABA receptors or to the N-methyl-d-aspartate (NMDA) receptors which play a role in the sensitization of spinal cord dorsal horn neurons in response to abnormal, repetitive peripheral nociceptive input [11]. Currently, the exact mechanism by which gabapentin decreases neuropathic pain is uncertain [11].

Conclusion

This case illustrates that reflex sympathetic dystrophy in children may be recurrent and migratory. Treatment for recurrent RSD should be started as soon as possible after diagnosis. The first line of therapy should include past successful treatments, but, as illustrated in this case, other modalities may also need to be used.

Even though there have been several case reports of reflex sympathetic dystrophy in children, very little is known about the natural history of recurrent RSD in this population. Even less is known about the efficacy of various treatment modalities in this subgroup of patients, and which ones if any may decrease the recurrence rate. Prospective studies are needed to better characterize the natural history of recurrent RSD as well as its response to different treatments.

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References