Reflex Sympathetic Dystrophy in Children

Çocuklarda Refleks Sempatetik Distrofi

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Özet

Refleks sempatetik distrofi (kronik bölgesel ağrı sendromu) çocukluk çağında ve pediatri pratiğinde sık karşılaşılan bir durum değildir (1). Refleks sempatetik distrofi (RSD) lokalize yaygın ağrı, sıklıkla şişlik, renk değişikliği, trofik değişiklikler ve vasomotor bozukluklar gibi otonomik anormalliklerle karakterize bir hastalıktır. Etiyolojisi tam olarak aydınlatılamamıştır. Hastalık kısmi olarak hasara uğrayan sinir tarafından innerve edilen bir alanda gelişir. Bu yazıda, çocukluk çağında nadir görülen refleks sempatetik distrofili iki kız hasta klinik ve laboratuar bulgularıyla literatür eşliğinde tartışılmıştır.

Anahtar Kelimeler

Refleks Sempetatik Distrofi; Bölgesel Ağrı Sendromu

Abstract

Reflex sympathetic dystrophy (chronic regional pain syndrome) isn't frequently encountered in practical pediatrics and childhood. Reflex sympathetic dystrophy syndrome (RSD) is a disorder characterized by widespread localized pain, often along with swelling, discoloration, trophic changes and autonomic abnormalities such as vasomotor disorders. Its etio-pathogenesis hasn't been completely determined. The disease can form in an area innerved by a partially damaged nerve and usually follows minor injury or trauma. In this paper, two girl patients with reflex sympathetic dystrophy are discussed along with the laboratory and clinic finding by accompaniment the literature as it is rarely seen in childhood.

Keywords

Reflex Sympathetic Dystrophy; Chronic Regional Pain Syndrome

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Intraduction

Reflex sympathetic dystrophy (chronic regional pain syndrome) isn't frequently encountered in practical pediatrics and childhood [1].

Reflex sympathetic dystrophy syndrome is a disorder characterized by widespread localized pain, often along with swelling, discoloration, trophic changes and autonomic abnormalities such as vasomotor disorders. Its etio-pathogenesis hasn't been completely determined. The disease can form in an area innerved by a partially damaged nerve and usually follows minor injury or trauma [2, 3]. Diagnostic criteria are the presence of regional pain and other sensory changes following a noxious event. The pain is associated with changes in skin color, skin temperature, abnormal sweating, edema, and sometimes motor abnormalities. The clinical course is commonly divided into three stages: first (acute or hyperemic), second (dystrophic or ischemic), and third (atrophic) stages [4]. Reflex sympathetic dystrophy can occur without any previous history of trauma, and may be recurrent and migratory [5]. The diagnosis is primarily clinical but laboratory tests can help to confirm the diagnosis. Medical approach, physiotherapy and sympathetic blockage can be administrated for therapy.

In this paper, two girl patients with reflex sympathetic dystrophy are discussed along with the laboratory and clinic finding by accompaniment the literature as it is rarely seen in childhood.

Case 1

15 year old female patient complained of pain in both her hands and left knee and was accepted into our pediatrics clinic. It was learned from the patient's history that the complaint started with increasing temperature, slight hyperemia, swelling and greater levels of pain approximately about six month ago but there was no history of trauma. She was constrained to move the joints particularly in the mornings and her pain was increasing with movement. Cold was beneficial to the swelling and pain in her hands. Also she was suffering from a stomachache which improved spontaneously and was not related to eating, particularly in the mornings. Juvenile Rheumatoid Arthritis was diagnosed with these findings and she was treated gradually with anti-inflammatory drug therapy such as ibuprofen, methotrexate and steroid but with no improvement. Although swelling and pain decreased in her left knee after the therapy, swelling and blueness showed an increase in her right hand wrist and back. The pain had started to be localized on the right side of her neck, right shoulder, right hand and wrist and she couldn't use her right hand. Drooping of her right upper eyelid had gradually started over the last two weeks. It was mild to moderate in intensity and non-variable during the day.

On physical examination; her right upper eyelid was ptosis but bilateral pupils' reflex, eye movements and fundus examination were normal (figure 1). Wrist, metacarpophalangial, proximal and distal interphalangial joints of her right hand was edematous and sweaty but didn't increase in temperature and hyperemia. Touching and movement were hypersensitive and painful. Her right hand was colder and of a greater violet red color than the other hand (figure 2). Range of motion of both her shoulder and elbows weren't limited but right shoulder's movements were painful. Perimeters of both knee joints were measured as 35 cm and didn't show pain, swelling and hyperemia. Erythrocyte sedimentation rate was 11 mm/h, C-reactive protein was <1 mg/dl, hemoglobin 13.1 g/dl, and white blood cell count 6820/mm3. Anti streptolysin-O titer, rheumatoid factor, and serum biochemistry were normal. Direct radiography revealed normal appearance in her right hand. Right upper extremity venous Doppler ultrasound and bone scan were normal.



Figure 1. The right upper eyelid of the patient was ptosis



Figure 2. The right hand of patient was colder and of a greater violet red color than the other hand

Case 2

17-year-old female patient presented with complaints of numbness in her right leg after having a 6 month history of IM injection. Complaints of leg weakness and difficulty walking began a week after the injection.

On examination; there was a 5x4 cm size reddish-purple skin color changes on the gastrocnemius muscle area and increasing regional sweating in the left leg. The area was hypersensitive and painful to touch and movement. The muscle power weakness of the left leg had a rate of 4/5 compared to the right leg. Central nervous system examination revealed normal tone, reflexes in all limbs. The examination findings of other systems of her body were normal. Erythrocyte sedimentation rate was 14 mm/h, C-reactive protein was <1 mg/dl, hemoglobin 14.3 g/dl, and white blood cell count 7400/mm3. Serum biochemistry was normal. Direct radiography revealed normal appearance in her left leg and left lower extremity venous Doppler ultrasound was

normal. Electromyography (EMG) findings didn't support the injury of sciatic nerve by IM injection. The patient was not given any medication and only received physical therapy.

Discussion

The incidence of reflex sympathetic dystrophy in childhood isn't known, but the disease is probably under diagnosed [1, 5]. The clinical diagnosis is suggested by the presence of at least two symptoms and signs from neuropathic pain (burning, dysaesthesia, paraesthesia, mechanical allodynia, hyperalgesia to cold) and autonomic dysfunction (cyanosis, skin mottling, hyper hidrosis, edema, temperature difference between extremities of >3°C) [1, 6]. As there isn't a current decisive test for diagnosis, although several tests are performed in a child with joint pain, the diagnosis may be delayed. Case 1 didn't recover from the arthritis in her hand and her symptoms didn't respond to anti-inflammatory drugs in the beginning.

Although previously published cases stated that minor injury or trauma may have been responsible for the onset of symptoms, some patients didn't have a history of trauma. [1, 3, 5]. Some authors also said that psychological problems frequently play a role in this disorder. Case 1 didn't have a history of trauma and psychological stress but case 2 had a history of IM injection.

Partial ptosis occurred due to the involvement of sympathetic fibers of the face and was a localized autonomic disorder [7]. The drooping of her right upper eyelid for two weeks was thought by us to be a localized autonomic disorder because her pupils were isochoric and her face was not anhidrotic. Tong and Nelson [5], stated that reflex sympathetic dystrophy may be recurrent and migratory, and an 11-year-old girl presented to their pediatric rehabilitation clinic three times with recurrent RSD in her bilateral arms over the next 5 years. We considered the migratory characteristic of the disease in the patient because of the regression of her left knee complaint pain occurring with movement in right side of neck and right shoulder and then the occurrence of the right eyelid drooping.

Early diagnosis has significantly increased the response to treatment and prognosis. While analgesic and anti-inflammatory drugs haven't created to decreasing pain, it has been noticed that amitriptilin, nifedipin, gabapentin, calsitonin, baklofen and biphosphanats are useful in the disease [1, 2, 6, 8]. Alongside a graduated program of physical therapy, transcutaneous electrical nerve stimulation and efferent sympathetic nerve activity, which can be interrupted surgically or chemically (intravenous regional guanethidin blockage, central sympathetic blockage, satellite ganglion blockage), have been fundamental principles in treatment [1-3, 5, 8]. The study of Ashwal et al. [3] reported that in contrast to adults, treatment with a graduated program of physical therapy and transcutaneous electrical nerve stimulation is beneficial in almost all patients and the prognosis of childhood reflex sympathetic dystrophy syndrome is favorable. In another case study four girl patients between the ages 9-14 with reflex sympathetic dystrophy were treated with intravenous regional guanethidin blockage and physiotherapy and all of them almost completely recovered [1].

Case 1 was started on 50 mg/day of pregabarin and physical therapy then the dose of pregabarin was increased. After two weeks of drug therapy, the pain in her shoulder and neck decreased and her hand's color minimally recovered. We thought that the patient completely recovered by sympathetic blockage. Case 2 gradually improved with physiotherapy and rest without drug treatment within 6 months.

We have discussed here two patients with reflex sympathetic dystrophy which is rarely seen in childhood and we wished to share our experiences.

Competing interests

The authors declare that they have no competing interests.

References

1. Lloyd-Thomas AR, Lauder G. Lesson of the week: Reflex sympathetic dystrophy in children. BMJ 1995;310(6995):1648-9.
2. Koc F, Zorludemir S, Yerdelen D, Bozdemir H. Atipik refleks sempatik distrofi sendromlu bir olgu. Cerrahpaşa J Med 2005;36(3):167-72.
3. Ashwal S, Tomasi L, Neumann M, Schneider S. Reflex sympathetic dystrophy syndrome in children. Pediatr Neurol 1988;4(1):38-42.
4. Kurvers HA. Reflex sympathetic distrophy: fact and hypotheses. Vasc Med 1998;3(3):207-14.
5. Tong HC, Nelson VS. Recurrent and migratory reflex sympathetic dystrophy in children. Pediatr Rehabil 2000;4(2):87-9.
6. Cimaz R, Matucci-Cerinic M, Zulian F, Falcini F. Reflex sympathetic distrophy in children. J Child Neurol 1999;14(6):363-7.
7. Mathias CJ. Disorders of the autonomic nervous system: autonomic dysfunction in pediatric practice. In: Swaiman KF, Ashwal S, Ferriero DM, editors. Pediatric neurology principles and practice. Philadelphia: Mosby Elsevier; 2006.p.2132-52.
8. van de Vusse AC, Stomp-van den Berg SG, Kessels AH, Weber WE. Randomized controlled trial of gabapentin in complex regional pain syndrome type I. BMC Neurol 2004;4:13.