CASE REPORT

A rare case of postoperative pain in congenital analgesia: case report

Um raro caso de dor pós-operatória em analgesia congênita: relato de caso

André Lavor Alves[1]. Flávio Lobo Maia[1]. Washington Aspilicueta Pinto Filho[2]. João Lucas Ferreira da Silva[1]. Joaquim Trajano de Lima Filho[1].

1 Hospital Universitário Walter Cantídio (HUWC), Fortaleza, Ceará, Brasil. 2 Hospital Infantil Albert Sabin (HIAS), Fortaleza, Ceará, Brasil.

ABSTRACT

Objective: we describe a case of a patient with familial dysautonomia and postoperative pain. **Methodology:** clinical follow-up for 10 years in a tertiary pediatric hospital. **Results:** it is a 12 years old teenager, diagnosed with Riley-Day Syndrome, after undergoing various orthopedic surgical procedures, evolves with changes in pain perception, in systematic assessments. **Conclusion:** Family Disautonomy is one of the most common Hereditary Sensory and Autonomic Neuropathies (HSAN). Children gradually evolve with sensory changes, leading to a progressive decrease in pain perception. In addition, spinal deformities and recurrent orthopedic trauma can occur. The reported patient presents, contradictorily, an increase in the perception of pain and allodynia after undergoing repetitive orthopedic procedures.

Keywords: Dysautonomia, familial. Hereditary sensory and autonomic neuropathies. Pediatrics. Pain, postoperative.

RESUMO

Objetivo: descrevemos um caso de paciente com disautonomia familiar e com dor pós-operatória. **Metodologia:** seguimento clínico por 10 anos em hospital terciário pediátrico. **Resultados:** trata-se de uma adolescente de 12 anos, com diagnóstico de Síndrome de Riley-Day, após submeter-se à vários procedimentos cirúrgicos ortopédicos, evolui com mudanças na percepção da dor, em avaliações sistemáticas. **Conclusão:** a Disautonomia Familiar é a mais comum dentre as neuropatias hereditárias sensoriais e autonômicas (NHSA). As crianças evoluem, gradualmente, com alterações sensoriais, levando a diminuição progressiva da percepção da dor. Além disso, podem ocorrer deformidades em coluna e traumas ortopédicos de repetição. A paciente relatada apresenta, contraditoriamente, aumento na percepção da dor e alodinia após submeter-se a procedimentos ortopédicos de repetição.

Palavras-chave: Disautonomia familiar. Neuropatias hereditárias sensoriais e autônomas. Pediatria. Dor pós-operatória.

Corresponding author: André Lavor Alves, Rua Barão de Canindé, 256, Montese, Fortaleza, Ceará. CEP: 60425-542. E-mail: andrelavoralves@yahoo.com.br
Conflict of interests: The authors have no conflicts of interest to declare.
Received: 12 Abr 2022; Revised: 25 Jul 2022; Accepted: 30 Ago 2022.

INTRODUCTION

The perception of postoperative pain in pediatrics is common. The duration of pain and its consequences for children are lasting, leading to several months of discomfort.[1]

Hereditary Sensory and Autonomic Neuropathies (HSAN) are a group of genetic disorders of the autonomous nervous system that reflect the relationship of this system with the sensory function in which their phenotype allowed several changes in sensory nervous and autonomous nervous system. These changes usually occurr in fine efferent and afferent nerve fibers, triggering variations that can affect both the autonomic system and sensory perception. The signs and symptoms of autonomous damage could be anhidrosis, paroxystic tachycardia, hyperthermia, and diaphoresis. Different genes are involved, affecting the development of small fibers and expressing variable phenotypes. These phenotypes usually appear until the second decade of life. Characteristic to all HSANs is that intradermal injection of histamine phosphate fails to elicit a normal axon-flare response. The most promising classification was proposed by Dyck et al in five subtypes, according to clinical criteria and mode of transmission.[2]

Type 3 congenital analgesia is a rare pathology of the autonomic nervous system, part of the group of genetic diseases known as HSAN. Is almost exclusive to individuals of Eastern European Jewish extraction, with incidence of 1 per 3600 live births. As children have a lack of or reduced pain perception, repetitive bone fractures may be common, justifying multiple orthopedic surgical procedures.[2] In addition to fractures, they may also present with kyphoscoliosis. The case demonstrates a child with type 3 HSAN who developed allodynia after undergoing several surgical interventions during clinical follow-up.

CASE REPORT

This is a case of a very rare aspect of congenital analgesia's disease. A 12-year-old girl who had been followed-up since she was 2 years old. Her first signs of illness were self-harm and denial of pain during routine vaccinations, which occurred in their first few months of age. At 2 years old, her first surgery was a leg fracture correction after falling from her crib. Since then, she was observed by the orthopedic and neurological services. Bimonthly, the orthopedic service requested X-ray to look for fractures. Next, the senior neurology and genetics team was responsible for qualifying her peripheral neurological disease using the HSAN Dyck criteria, without the need for genetic evaluation.

She was diagnosed with HSAN type 3 (familial dysautonomia or Riley-Day syndrome). The patient had unexplained tachycardia, sweating and fever at the time of anxiety. The child's parents are not related to each other and there are no reports of the pathology in their families.

Due to bone trauma and recurrent infections, she underwent several orthopedic surgical procedures. The patient required a total of 12 interventions, most of them surgical debridement for osteomyelitis treatment.

In clinical follow-up, after systematic evaluation, it was observed that the patient evolved with changes in pain perception. Previously, the patient only recognized tactile stimuli, evolving with progression in pain perception and allodynia. This condition is incompatible with the clinical presentation of the disease. The patient was treated for three months with an association of weak opioid (codeine) and tricyclic antidepressant (amitriptyline). We believe that drug intervention is responsible for the total reversal of the process, with an improvement in the clinical condition.

This case was collected in a reference center in pediatrics, through clinical monitoring for 10 years in the service of Pain and Anesthesiology, together with pediatric neurology. This case report was published with the written consent of the patient (CAAE 30771320.3.0000.5042).

DISCUSSION

Family Disautonomy or Riley-Day Syndrome is the most common among HSAN, has a gradual evolution, and is caused by mutations in the ELP1 gene. The patients are homozygous in almost all cases, with a defect in the codification of protein associated with kinase Ik B (IKAP). This protein is involved in the transcription and truncation of other proteins involved in the migration and motility of crest neurons, affecting sensory and autonomic functions. Signs and symptoms vary among individuals and depend on age.[3]

Sensory changes are demonstrated, as well as decreased perception of pain and temperature. The decrease in the perception of pain evolves progressively, and temperature does not change over time.[2]

Children commonly present emotional lability, mainly associated with episodes of crisis. Besides this symptom, episodes of vomiting, hypertension, tachycardia, sweating and exanthema constitute the "dysautonomic crisis", which occurs in 65–80% of patients, and is a frequent cause of hospitalizations. This crisis occurs as a response to physical or emotional stress or to daily arousal and seems to be associated with central autonomic dysfunction.[4]

The deformity of the spine is the most common orthopedic manifestation, with a prevalence of 48% at 10 years. Of these, scoliosis and kyphoscoliosis are the most prevalent.[5] Patients manifest a higher incidence of fractures compared to the population on the same age, although less active, most in the period prior to bone maturation. It is believed that progressive denervation causes spinal deformity and reduced pain perception often justifies the fractures.[4,5]

We believe that the incompatibility of the patient's clinical evolution, for progressively developing changes in the perception of pain and allodynia, was generated by the excess of orthopedic procedures, leading to central hypersensitization. As with the central sensitization phenomenon, repetitive stimulation of temperature and proprioception pathways may

have generated this symptom of pain in the patient. Several endogenous mediators are involved in these processes, such as histamine and nerve growth factor, which appear to play an important role in orchestrating a variety of signaling cascades necessary for an inflammatory response associated with this process.

The excessive number of procedures in this case was due to the families' lack of guidance regarding the disease and the children's difficulty in understanding. There is a need for greater knowledge of this pathology on the part of health professionals in order to improve care for this rare pathology. That is our message in this case report. At the moment, there are few consistent works and similar reports in the literature. The pathophysiology of this set of diseases is poorly understood, as well as there are no tools for a definitive diagnosis.

The limitation of the study is the absence of similar cases in the literature for comparison purposes.

REFERÊNCIAS

1. Rosenbloom BN, Pagé MG, Isaac L, Campbell F, Stinson JN, Wright JG, et al. Pediatric Chronic Postsurgical Pain And Functional Disability: A Prospective Study Of Risk Factors Up To One Year After Major Surgery. J Pain Res. 2019;12:3079-98.

2. Axelrod FB, Gold-von Simson G. Hereditary sensory and autonomic neuropathies: types II, III, and IV. Orphanet J Rare Dis. 2007;2:39.

3. Schwartzlow C, Kazamel M. Hereditary Sensory and Autonomic Neuropathies: Adding More to the Classification. Curr Neurol Neurosci Rep. 2019;19(8):52.

4. Dietrich P, Dragatsis I. Familial Dysautonomia: Mechanisms and Models. Genet Mol Biol. 2016;39(4):497-514.

5. Laplaza FJ, Turajane T, Axelrod FB, Burke SW. Nonspinal orthopaedic problems in familial dysautonomia (Riley-Day syndrome). J Pediatr Orthop. 2001;21(2):229-32.

Como citar:
Alves AL, Maia FL, Pinto WA Filho, Silva JL, Lima JT Filho. A rare case of postoperative pain in congenital analgesia: case report. Rev Med UFC. 2023;63(1):1-3.