CASE PRESENTATION – UNILATERAL DYSKINETIC CEREBRAL PALSY: TIME FOR A CHANGE IN CEREBRAL PALSY CLASSIFICATION?

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Introduction. *Unilateral dyskinesia is a rare neurological disorder characterized by involuntary, abnormal movements on one side of the body. In children, understanding its clinical presentation and age-appropriate rehabilitation strategies is essential for the best outcome.*

Material and methods. *In this case-presentation, we conducted an in-depth examination of a 10-year-old boy with unilateral dyskinesia, previously diagnosed with bilateral spastic cerebral palsy. Extensive pediatric neurological assessments, including neuroimaging and child-friendly clinical evaluations, were performed to comprehensively document the patient's condition.*

Results. *Birth data: 39 weeks gestation, singleton, weight – 3950 g, length – 56 cm, head circumference – 36 cm, Apgar score at 5 minutes – 7, without birth-related complications. Paraclinical investigations revealed MRI-identified basal ganglia lesions in the putamen. Functional classifications indicated GMFCS II, MACS II, epilepsy, normal vision, normal hearing, EDACS level I, Viking speech scale III, and moderate intellectual disability. The pediatric patient exhibited pronounced dyskinesia limited to the right side of the body, which has progressively worsened over the past six years.*

Conclusions. *The results are in line with specific variables of dyskinetic CP. This study emphasizes the challenges in diagnosing pediatric patients with unilateral dyskinesia during the early stages. Clinical features are evident; therefore, rehabilitation strategies should be selected accordingly.*

Cuvinte-cheie: *dischinezie, unila-teral, studiu de caz, reabilitare.*

STUDIU DE CAZ. PARALIZIA CEREBRALĂ (PC) DISCHINETICĂ UNILATERALĂ IMPUNE O SCHIMBARE A CLASIFICĂRII PC?

Introducere. *Dischinezia unilaterală este o afecțiune neurologică rară, caracterizată prin mișcări involuntare, anormale pe o parte a corpului. La copii, înțelegerea prezentării sale clinice și a strategiilor de reabilitare adecvate vârstei este esențială pentru obținerea rezultatului scontat.*

Material și metode. *În prezentarea de caz, a fost efectuată o examinare aprofundată a unui băiat de 10 ani cu dischinezie unilaterală, diagnosticat anterior cu paralizie cerebrală spastică bilaterală. Pentru a documenta în mod cuprinzător starea pacientului, au fost efectuate evaluări neurologice, pediatrice extensive, inclusiv neuroimagistice, și evaluări clinice prietenoase pentru copii.*

Rezultate. *Date la naștere: 39 de săptămâni de gestație, greutate – 3950 g, talie – 56 cm, perimetrul cranian – 36 cm, scor Apgar la 5 min. – 7, fără complicații legate de naștere. Cu ajutorul RMN s-au evidențiat leziuni ale ganglionilor bazali în putamen. Clasificările funcționale au indicat GMFCS II, MACS II, epilepsie, vedere normală, auz normal, EDACS I, scara de vorbire Viking III și dizabilitate intelectuală moderată. Pacientul pediatric a prezentat dischinezie pronunțată, limitată la partea dreaptă a corpului care s-a agravat progresiv în ultimii șase ani.*

Concluzii. *Rezultatele sunt în concordanță cu specificul PC dischinetice. Acest studiu subliniază provocările în diagnosticarea pacienților pediatrici cu dischinezie unilaterală în stadiile incipiente. Caracteristicile clinice fiind evidente, strategiile de recuperare ar trebui selectate conform acestora.*

INTRODUCTION

Dyskinetic cerebral palsy (CP), a subtype of cerebral palsy marked by uncontrolled, irregular movements, arises from damage to the extrapyramidal regions of the brain responsible for movement coordination. This particular form of CP is relatively uncommon, constituting between 6% and 17% of all cerebral palsy cases (1, 2). Unilateral dyskinesia, an unusual manifestation of this disorder, is characterized by involuntary movements on one side of the body and presents unique management challenges. Children with dyskinetic CP generally experience more severe motor impairments and are more likely to have associated disabilities compared to those with spastic CP (3, 4). Recognized risk factors for dyskinetic CP include sentinel events during childbirth, and characteristic lesions in the basal ganglia and thalamus are typically observed in neuroimaging (4, 5). Despite a general decline in the overall birth prevalence of CP in high-income countries (6) over the last two decades, data indicates an increase in the prevalence of dyskinetic CP in Europe from the 1970s to the 1990s (7 – 12). It's uncertain whether the observed decrease in dyskinetic CP prevalence from 2005 to 2010 represents a sustained trend or a temporary dip. The rise in dyskinetic CP's birth prevalence towards the late 20th century (7), concurrent with the stable prevalence of spastic quadriplegic CP despite significant advances in obstetric and perinatal care, suggests that these CP subtypes predominantly originate antenatally and are rarely preventable. This perspective was further supported in 1998 when an international consensus statement identified dyskinetic CP or spastic quadriplegic CP as key criteria for defining an acute intrapartum event (13). Understanding and preventing dyskinetic CP remains complex and challenging. Given its severity and resistance to treatment, a comprehensive, multi-disciplinary approach is essential for the effective management and support of individuals with this condition.

CASE PRESENTATION

In this study, we conducted an in-depth examination of a 10-year-old boy with unilateral dyskinesia, previously diagnosed with bilateral spastic CP. Extensive pediatric neurological assessments, including neuroimaging and child-friendly clinical evaluations, were performed to comprehensively document the patient's condition.

1. Birth data

Born in 2013 at a level II hospital, the pregnancy was the first, and it resulted in a single birth. The gestational age was 39 weeks, with a weight of 3980 g, length of 56 cm, head circumference of 36 cm, chest circumference of 34 cm, and abdomen circumference of 33 cm. The APGAR score was 4/7. The birth was through caesarean section due to hypoxia caused by a circular umbilical cord around the neck. The newborn was transferred to a tertiary level hospital on the same day and was hospitalized for 14 days in intensive care. The diagnosis at birth was moderate asphyxia and cerebral ischemia grade I-II, along with a convulsive syndrome. The child underwent repeated rehabilitation treatments and on 15.05.15, Botulinum Toxin Type A (Dysport) 200 IU was administered.

2. Clinical and paraclinical evaluations:

1. Cerebral Computed Tomography 2015 - cystiform dilatation of the cistern magna is determined. 2. Electroencephalography (EEG). During artifact-free periods, disorganized brain bioelectric activity is noted, without pathological interhemispheric asymmetry and epiphenomena. Functional probe reactivity attenuated. 3. M-Echo, 12.2023 structural averages are not deviated LS=LD=61mm Ventricle III=3 mm 4. Nuclear Magnetic Resonance (NMR), 2015 - Lesions of the basal ganglia in the putamen 5. Ophthalmologist consultation-07.2015 F.O. Moderate dilated veins, 03.2019 F.O. No changes.

3. Current functional assessments

3.1. Neurological status:

During the first year of life with a delay in motor development: the child was not sitting at 6-8 months, at 1.5 years he was not walking. At the age of 1 year, the diagnosis of Cerebral Palsy in the form of Spasticity was established. Since birth, he has had seizures, treated with Clonazepam. Last seizure 2 years ago. Last treatment in 12.2023. Hypertonic muscle tone, emphasis on the left. Bilateral muscle strength in upper limbs slightly diminished D=S=4p. In the bilateral lower limbs proximally 3 points; distal flexion 4 points; extension 2 points. ROT live superior D=S, inferior: patellar -exaggerated; Achilles-D=S; Babinski positive bilaterally, more pronounced on the left side. Unstable Romberg pose, the coordination tests performed with more pronounced inaccuracy on the right (when performing the index-

nose test, a tremor appears in the right hand).

3.2. Speech therapist evaluation
Dysarthria, global language disorders grade II. Viking speech scale III. EDACS (Eating and Drinking Ability Classification) level I.

3.3. Psycho-pedagogue evaluation
Purpose and objectives - training and development of attention. Conclusion - the cognitive level does not correspond to the chronological age, IQ=62.

3.4. Gross motor function evaluation
The patient is diagnosed with Cerebral Palsy, bilateral spastic form. After a more detailed analysis of the gross motor function and the biped movement pattern, the patient presents a mixed form - spatial form on the left side and dyskinetic - on the right. The patient experiences difficulties while moving, finding it hard to maintain balance with a swaying, inclined movement on the left side. While walking, the child stabilizes the right leg with the left upper limb. During movement with the legs, they go into more pronounced hyperextension on the right, and the patient has an asymmetrical posture with an inclination on the left side. Movement control is better in the left hemisphere. Gross Motor Function Level II (GMFCS II).

3.5. Assessment of fine motor function
While performing simple abduction and adduction movements, the patient demonstrates more concrete, coordinated, and precise movement with the left hand than with the right. The left side, being spastic, is better controlled by the patient than the right side, which is dyskinetic. When reaching for a fitball, the right hand is placed ahead of the left. When holding the ball, the left hand is placed below, the right above, and the head looks to the right. When holding an object with the left hand, the right hand goes into abduction, supination, and dorsiflexion, which demonstrates that the extrapyramidal system is damaged, and the brain eliminates the presence of the right hand. However, if asked to hold his hand close to his body, he struggles, and movements with his left hand become clumsier. With his right hand, he can only grasp and hold objects the size of a ping-pong ball and then throw it. The interdigital grip and the terminal opposition grip cannot be performed with the right hand. With the left hand, he performs prehension through terminal opposition only with the ring finger and the little finger, and with the index and the medius only by sub-terminal opposition. When building a wall with blocks, he holds the pieces very tightly with his right hand due to the lack of co-contraction, while with his left hand he makes an effort to join the pieces. Fine movements of the hand and fingers are performed with difficulty and may be accompanied by intentional tremors. Manual Ability Classification (MACS) level II.

Lower limbs
Standing and walking are achieved with a large base of support. During the orthostatic position, support is performed only on the left leg, with the right leg initially lifted and then the heel lowered. The patient cannot maintain a standing position without movement for more than 3 seconds. Due to the lack of coordination of the activity of agonists, antagonists, and synergists, movements become uncontrolled and are carried out with high amplitude. This results in a “mowing gait” characterized by an abduction pattern along with dorsiflexion and eversion of the leg, and as compensation, hyperextension of the hip and knee. In activities of daily living (ADL), all actions are performed with the left hand, with the right hand not involved in the action. During dressing and undressing, the left hand engages and dresses the right hand, while the right hand is put aside, with the feet fixed on the floor. When putting on and taking off pants, socks, and shoes, coordinating the right leg is difficult. He manages, with difficulty, to fasten and unfasten buttons, and can easily unfasten and fasten zippers.

DISSCUSIONS

When conducting the rehabilitation process for dyskinetic cerebral palsy, whether it is unilateral or bilateral, it must be customized to fit each person's individual needs, abilities, and limitations. However, there are some general differences in the rehabilitation approach between the two types:

Unilateral Dyskinetic Cerebral Palsy: 1. Specific Focus: Rehabilitation will often be more focused on the part the affected body, which allows an intensive concentration on improving the function and coordination of that part. 2. Physical and Occupational Therapy: It will be aimed at improving strength, flexibility, and coordination of the affected body part. Exercises and activities are often designed to stimulate the use of the affected limb

and to encourage the development of fine and gross motor skills. 3. Use of Assistive Equipment: May be required on the affected side, such as orthotics or other support devices, to improve alignment and operation. 4. Compensatory Strategies: Patients can learn to use more effectively the unaffected side of the body to compensate and perform daily activities.

Bilateral Dyskinetic Cerebral Palsy: 1. Broad Approach: Rehabilitation requires a more global approach because both parts of the body are affected. This involves more coordination complex and careful planning to address the needs on both sides of the body.2. Physical and Occupational Therapy: This will aim to improve motor control, strength, and coordination on both sides of the body. Therapists may use a variety of techniques, including whole-body exercises, activities involving both hands and feet, and relaxation techniques to control involuntary movements. 3. Extensive Adaptations in Daily Life: Due to bilateral involvement, a wider range of assistive equipment and modifications of the home or school environment may be necessary to facilitate independence and safety. 4. Management of Involuntary Movements: Complex approaches may be required for managing uncontrolled movements, including medication, therapy with controlled movement, and in some cases, surgery.

Common Considerations: 1. Speech and Language Therapy: Both types can benefit from speech and language therapy, especially if facial muscle control is affected and coordination is needed for speech and eating. 2. Psychosocial Support: Emotional and psychological support is crucial in both cases, as cerebral palsy can have a significant impact on self-esteem, independence, and quality of life. 3. Multidisciplinary Approach: A multidisciplinary team involving doctors, therapists, educators, and other specialist services is essential to provide comprehensive care.

CONCLUSIONS

1. The results are in line with specific variables of dyskinetic CP. This study emphasizes the challenges in diagnosing and rehabilitating pediatric patients with unilateral dyskinesia during the early stages. In conclusion, although there are common basic principles in CP rehabilitation, the specific approaches and intensity of interventions may vary considerably between unilateral and bilateral cases.
2. An individualized assessment is essential for creating a personalized treatment plan to effectively address each person's needs and goals.

CONFLICT OF INTEREST

Authors have no conflict of interest to declare.

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