Introduction: Chronic inflammatory demyelinating polyradiculoneuropathy is an immune-mediated disease of the peripheral nervous system, causing demyelination and axonal degeneration. This entity is uncommon in childhood, and with little literature available.

Case Presentation: We describe a case of a 26-month-old boy, who presented imbalanced gait and muscle strength decrease, with two days of evolution. Neurological examination revealed decrease in distal muscle strength and absence of lower limbs reflexes. He had elevated CSF protein with no increase in cells. Electromyography evidenced severe demyelinating neuropathy. Immunoglobulin, carbamazepine and physiotherapy was started, with partial recovery. One year later, presented a similar episode and immunoglobulin was restarted, having performed on a monthly basis for 8 months. At 9-years-old, there was a new recurrence.

Conclusion: Authors pretend to emphasize the challenges in the diagnosis and management of pediatric CIDP. We also highlight the importance of corticosteroids in this pathology, contrary to its role in Guillain-Barré syndrome.

Keywords: Pediatric CIDP; Demyelinating Neuropathy; Albumino-Cytologic Dissociation; Treatable Neuropathy

Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune mediated treatable disorder of the peripheral nerves, with predominant motor involvement and an insidious onset, or recurrent episodes.

Children present slowly progressive or relapsing episodes of gait difficulty, distal symmetric weakness and sometimes paranesthesia. Reflexes are absent or depressed.

Laboratory findings include elevated cerebrospinal fluid (CSF) protein with no increase in cells. Electromyography shows demyelination.

This entity is uncommon in childhood, and information regarding the clinical presentation, disease course, and response to therapy is very limited in the literature.

Citation: Sara Teixeira Pinto, et al. "Chronic Inflammatory Demyelinating Polyradiculoneuropathy in a 26-Month-Old Boy". EC Paediatrics 9.9 (2020): 68-70.
Chronic Inflammatory Demyelinating Polyradiculoneuropathy in a 26-Month-Old Boy

Case Presentation

A previously healthy 26-month-old boy, brought to the emergency department due to imbalanced gait and muscle strength decrease, with two days of evolution. There was no epidemiological context of disease, recent vaccinations or medication ingestion. Familial history was unremarkable for neurologic or autoimmune diseases. Neurological examination revealed gait instability and decrease in distal muscle strength. Reflexes were absent in the lower limbs and decreased in the uppers. He also presented altered sensitivity.

Laboratory studies including blood cell count, serum electrolytes, glucose, reactive protein C, total creatinine-kinase, and myoglobin were normal. Brain computed tomography scan was normal and urine drug screening negative.

Serology excluded recent or active infection by cytomegalovirus, epstein-barr, herpes simplex type 1 and 2, mycoplasma pneumoniae, chlamydia pneumoniae, legionella pneumophila and parvovirus.

Cerebrospinal fluid (CSF) analyze revealed elevated protein (50.6 mg/dl) with no increase in cells (1/uL white blood cells (WBC)); CSF cultures were sterile and polymerase chain reaction was negative for herpes virus type 1 and 2, epstein-barr; cytomegalovirus, enterovirus, campylobacter and Mycoplasma pneumoniae.

Patient’s clinic and albumin-cytologic dissociation were suggestive of Guillain-Barre syndrome (GBS), so treatment with intravenous immunoglobulin (IVIG) and carbamazepine was started. Electromyography supported the diagnosis by evidencing severe demyelinating neuropathy. He was oriented to physiotherapy, with partial recovery of limbs strength.

One year later, presented a similar clinical episode, with overlapping study, namely the electromyography. IVIG was restarted, having performed on a monthly basis for 8 months.

At 9-years-old, had a new recurrence, in post-infectious context, with affection of all limbs. Electromyography revealed severe demyelinating neuropathy, without alterations suggestive of acute lesion.

Nowadays, at 12-years-old, he has autonomous gait, with a marked distal motor deficit at lower limbs and slight in upper limbs.

Discussion

CIDP is rare enough in childhood to present diagnosis and treatment challenges. Some children may demonstrate an initial acute “GBS-like” symptom onset before their relapse and diagnosis with CIDP [1-3].

Clinic criteria includes progression of muscle weakness in proximal and distal muscles of upper and lower extremities over at least 4 weeks, or alternatively when rapid progression (“GBS-like” presentation) is followed by relapsing or protracted course (more than 1 year), and areflexia or hyporeflexia [4].

Diagnosis is made conjugating clinical characteristics, electromyography and nerve conduction studies and elevated protein in CSF [3,5,6].

Demyelinating process, with or without axonal damage, is revealed by electromyography and nerve conduction studies, and in some instances by nerve biopsy. Investigation of CSF shows typical albuminocytologic dissociation: elevated protein (> 35 mg/dl) without leukocytosis (WBC count < 10/uL) [3,4,7].

Differential diagnoses include its acute counterpart, GBS, as well as hereditary and metabolic causes of polyneuropathy [5,8].

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IVIG and corticosteroids are both effective as first-line treatment, and response is usually favorable. Recommendations regarding the choice of second-line therapy can only be made on the basis of current practice described in case reports. Safety and efficacy data are insufficient [3,9].

Swift recognition and empiric start of treatment are important to avoid potentially irreversible axonal damage [6,10].

The natural history of CIDP is highly variable. Although children usually tend to recover, some may suffer long-term disability [3-6].

Conclusion

Authors pretend to emphasize the challenges in the diagnosis and management of pediatric CIDP. Despite being a rare diagnosis, clinical suspicion, early recognition and investigation with prompt management can result in better outcomes of this potentially treatable neuropathy. We also highlight the importance of corticosteroids in this pathology, contrary to its role in GBS.

Bibliography


Volume 9 Issue 9 September 2020
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