CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP) IN A 3 YEAR OLD BOY: A CASE REPORT

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PIN PERDOSNI
ACEH 2025

INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic autoimmune neuropathy characterized by demyelination of the nerve roots.

This demyelinating polyneuropathy has a higher prevalence in men than in women, namely a sex ratio of 1.5 to 4.7.

Clinical presentation includes episodes of difficulty walking that are progressive or relapsing slowly, symmetrical weakness of proximal and distal muscles, hyporeflexia or areflexia, and sometimes paresthesias. The pathogenesis mechanism of CIDP in children is the same as in adults, so that the treatment of CIDP in adults is applied to cases of CIDP in children.

Symptoms of CIDP, by definition, develop gradually over more than 8 weeks in most cases, although especially in children, the onset of the disease is most often acute, which develops in less than 4 weeks, or subacute in 4-8 weeks.

INSIDEN CIDP

- 1,0–1,9 per 100.000 populasi pada orang dewasa
- 0,48 per 100.000 populasi pada anak

Case Report

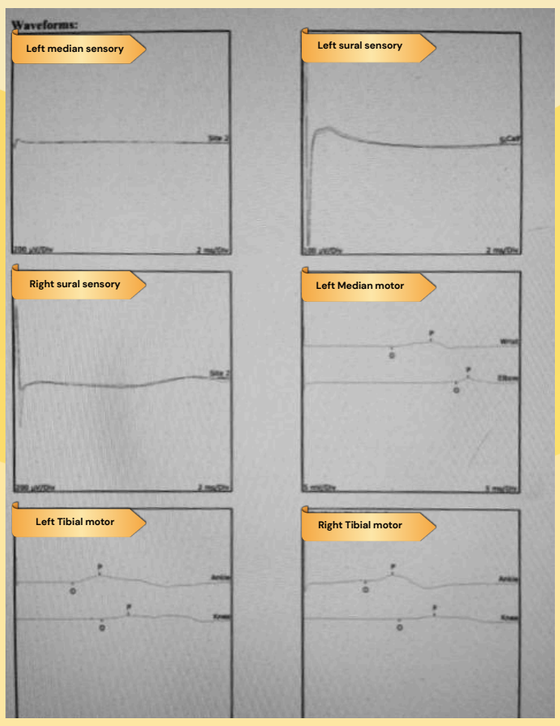
3 year old boy

- Complaints begin with fever
- 7 days later the patient complained of pain in both legs.
- 10 days later the patient complained of weakness in both legs.
- 3 weeks later, the complaints of weakness in both legs became more severe.
- Onset 1 month the patient was referred to PON Hospital, found weakness in both legs with Incat score 4
- Giving MP Pulse 1 therapy, there was still weakness in both legs with an Incat score of 4
- 5th MP Pulse Therapy, weakness of both legs began to improve with Incat score 3
- 5 days after the 5th MP Pulse therapy, the weakness of both legs improved with an Incat score of 2
- 1 month after MP Pulse therapy, the patient went for a check-up at the polyclinic, the weakness in both legs improved significantly, with an Incat score of 1.

2 months after MP Pulse therapy, the weakness of both legs improved significantly, with an Incat score of 0. However, there was a fluctuating period, the patient experienced mild weakness in the lower legs, especially triggered by heavy activities, such as playing ball or walking long distances, where both lower legs looked like the letter "o", but within a few days it slowly began to improve spontaneously.

In NCS, lower limb sensory action potentials are negative, motor conduction is prolonged, amplitude is low, conduction velocity is slow and F wave latency is prolonged, indicating sensorimotor polyneuropathy.

Neurophysiological examination results



DISCUSSION

- CIDP complaints involve symmetrical motoric, dominant in the lower extremities. Physiological reflexes are decreased / negative. 1/3 of pediatric patients experience sensory symptoms, characterized by paresthesia / dysesthesia.
- Differentiating CIDP from GBS is difficult because the initial symptoms are similar. In our patient with acute onset was initially diagnosed with GBS.
- CSF protein levels were increased in 86% of cases, and were found in our patient.
- Confirmatory diagnosis is NCS in sensory and motor nerves, which shows a pattern of demyelination characterized by (1) prolonged distal latency, (2) decreased motor conduction velocity, and (3) negative / prolonged F wave latency according to electrodiagnostic criteria.
- Although IVIG produces a good response of 50-80%, there is a risk of drug dependence in 80%. In contrast, steroid treatment was successfully stopped in about 83% of cases. In our patient after MP pulse therapy the patient experienced significant improvement.

CONCLUSION

CIDP is a rare disorder in children. This disease can cause permanent disability. However, if diagnosed and treated in time, it will generally improve the clinical outcome of the patient. In the course of the disease, the most common percentage is polyphasic - relapse - remission, so it requires close observation.

cerebrospinal fluid analysis

Makroskopik	Nilai
Kejernihan	Keruh
Bekuan	Positif

Kimia	Nilai
Nonne	Positif
Pandy	Positif
Glukosa serum	124
Glukosa cairan otak	98
Protein cairan otak	184
Klorida (Cl)	125

Mikroskopik	Nilai
Hitung sel	14
PMN	22
MN	78
Pewarnaan tinta india	Tidak ditemukan Cryptococcus

Video

Reference

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Keywords: Chronic inflammation, demyelination, polyneuropathy