EFFICACY OF CYCLOSPORINE AND PREDNISONE COMBINATION THERAPY IN A-CIDP ASSOCIATED WITH MINIMAL CHANGE NEPHROTIC SYNDROME

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Background

The simultaneous occurrence of GBS and nephrotic syndrome has previously been reported. However A-CIDP and nephrotic syndrome remain rarely reported (Tab. 1)

Tab. 1

Cases of inflammatory neuropathies associated with focal segmental glomerulosclerosis.

Authors	Age/Gender	Clinical diagnosis	Duration from neuropathy onset to nadir	Relapse(s) or progression/ Treatment received
Olbricht et al. [7]	28/M	Acute-onset CIDP	5 weeks	Relapse #1 at 2 years
Souayah et al. [8]	49/M	Acute-onset CIDP (Probable)	2 weeks	Relapse #1 at ~ Week 5/ IVIg Relapse #2 at ~ Week 7 Progressed despite 5 days' IVIG / subsequent improvement with plasma exchange and prednisone
Careless et al. [1]	73/F	Relapsing GBS	17 days	Relapsed 2 days after plasma exchange was stopped at ~ week 4-5/ Plasma exchange and prednisone
Girolami et al. [2]	40/M	CIDP	Exact duration unknown	#1 Relapse at 2 months #2 Relapse at 13 months #3 Relapse at 18 months
Henderson et al. [4]	58/M	CIDP	4 months	Progression over 4 months
Heckmann et al. [3]	46/M	GBS	14 days	None reported
Oh et al. [6]	56/M	GBS	3 weeks	None reported
Lim et al. [5]	22/M	GBS	9 days	None reported

Clinical History

A 78-year-old woman reported paresthesias at hands and feet and progressive weakness of both legs, with onset two weeks after an upper respiratory tract infection. He underwent to neurological examination, laboratory tests on serum and cerebrospinal fluid and electroneurography (ENG).

Laboratory tests

SERUM: negative or normal for routine laboratory tests, ANA, ENA, ANCA, FR, immunoelectrophoresis, cryoglobulins, anti-HCV antibodies, thyroid hormones, anti-MAG, anti-ganglioside, anti-Ach-R antibodies, neoplastic markers.

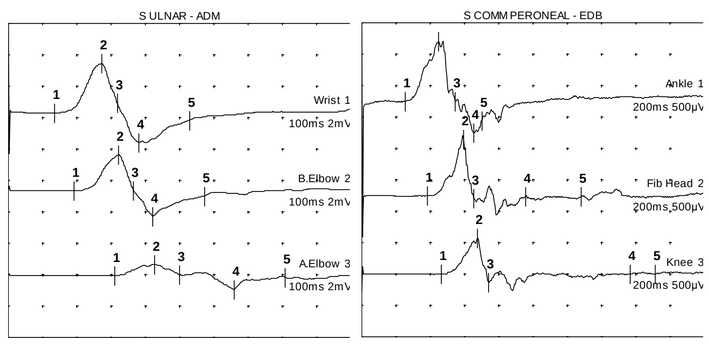
CSF: normal cell count, hyperproteinorrachia (3 g/L), and identical cerebrospinal fluid and serum oligoclonal bands.

Neurological Examination

Tetraparesis with MRC=2, Hughes scale= 4; decreased pinprick sensation and deep tendon areflexia at four limbs.

ENG

Nervo / Posizioni	Latency ms	Ampl mV	Distance cm	Velocity m/s	Area mVms	Dur. ms	Dur. %
S ULNAR - ADM							
Wrist	**13,70**	**3,2**	7		20,3	18,30	100
B.Elbow	19,25	**2,3**	24	**43,2**	17,4	17,40	95,1
A.Elbow	31,30	**0,7**	13	**10,8**	6,7	18,70	102
S COMM PERONEAL - EDB							
Ankle	**25,20**	**1,0**	8		12,5	29,40	100
Fib Head	38,40	**0,9**	33	**25,0**	9,0	27,20	92,5
Knee	46,60	**0,6**	10	**12,2**	7,7	27,40	93,2



Reduced motor conduction velocity, with conduction blocks in left peroneal and ulnar nerve. Absent sensory action potential of sural nerves.

Diagnosis and Therapies

GBS → INTRAVENOUS IMMUNOGLOBULINS (0.4 g/Kg/die for 5 days) → Neurological deficits slightly improved (MRC=3)

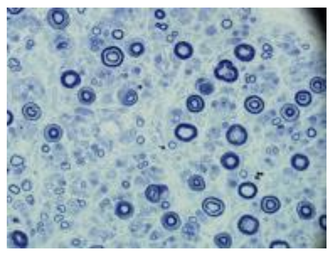
→ After three weeks clinical worsening with severe flaccid tetraparesis

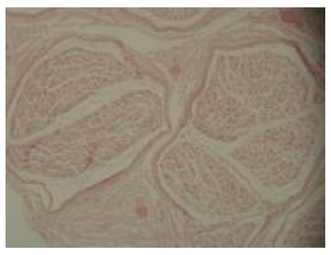
→ INTRAVENOUS IMMUNOGLOBULINS : INEFFECTIVE

→ PLASMA EXCHANGE: Not performed due to severe proteinuria (34 g/L) with hypoalbuminemia and hypoproteinemia: **NEPHROTIC SYNDROME**

→ RENAL BIOPSY: minimal change nephrotic syndrome (MCNS) (Data not shown)

NERVE BIOPSY: demyelinating and axonal damage





Final Diagnosis and Therapies

A-CIDP

Prednisone (up to 75 mg/day, then 50 mg/day)

Cyclosporine (from 50 to 75 mg/day)

IMPROVEMENT WITH FOLLOWING STABLE COURSE OF NEUROLOGICAL SYMPTOMS AND RENAL FAILURE

Conclusion

In this patient an initial diagnosis of GBS was performed because of the clinical worsening within 2 weeks from the onset. Although CIDP is defined by a clinical evolution over more than 2 months, 31% of patients with CIDP may present an acute-onset. The neurological picture was associated with a nephrotic syndrome. The association of inflammatory neuropathies and nephrotic syndrome has been previously reported with variable clinical course over time and severity. A-CIDP and MCNS responded to immunosuppressive therapy, suggesting a common pathogenesis. Moreover, in our case a favourable response to corticosteroid therapy corroborate the diagnosis of A-CIDP rather than GBS.

References

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