Rituximab for treatment of chronic inflammatory demyelinating polyradiculoneuropathy: a retrospective chart review

Matthew Varon, Mamatha Pasnoor, Omar Jawdat, Melanie Glenn, Jeffrey Statland, Duaa Jabari, Constantine Farmakidis, Richard J. Barohn, Mazen M. Dimachkie.

Neuromuscular Division – Department of Neurology
The University of Kansas Medical Center, Kansas City, Kansas, USA

BACKGROUND

- Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an autoimmune neuropathy of unknown precise etiology.
- First-line therapies include corticosteroids, immunoglobulin (IVIG), and plasma exchange (PE).
- Several case reports and small observational studies have demonstrated variable efficacy of rituximab for treatment of CIDP.

OBJECTIVE

- To evaluate the therapeutic response to rituximab in patients presenting with treatment-resistant CIDP.

METHODS

- Retrospective chart review was performed on the patients who presented to the neuromuscular clinics with CIDP and received rituximab for this diagnosis.
- Demographic and clinical information was obtained.
- The following were obtained or inferred from data available in the chart:
  - Baseline and post treatment MRC sum score.
  - Inflammatory neuropathy cause and treatment (INCAT) disability score.
  - Modified Rankin score (mRS).

RESULTS

- 5 patients (2 males and 3 females; age range 36 to 72) were identified.
- Rituximab was chosen based on prior use of multiple therapies without adequate clinical response.
- The median number of prior therapies was three.
- Prior treatments included corticosteroids, mycophenolate mofetil, IVIG, azathioprine, and PE.
- Three patients received rituximab for management of chronic symptoms of CIDP. Two patients received rituximab infusions while admitted for acute symptomatic exacerbation.

RESULTS

Subject (Sex, age)	Symptom duration prior to RTX (months)	Past therapies (* current therapy at time of RTX)	RTX cycles (1gm Q2 wks x2) (**500mg Q2 wks x2)
Male, 65	4	MMF*, IVIG*, prednisone*	2
Male, 72	10	MMF*, prednisone, IVIG*, PLEX	1
Female, 64	18	AZA*, prednisone*, IVIG*, PLEX*	1
Female, 36	168	AZA, prednisone, IVIG	1**
Female, 66	4	IVIG, PLEX, prednisone*	1

RESULTS

Subject	Pre-RTX			Post-RTX			
	MRC ss	INCAT	mRS	MRC SS	INCAT	mRS	Time (mos)
Male, 65	54	4(2+2)	2	55	4(1+3)	2	12 months
Male, 72	40	6(3+3)	3	48	6(3+3)	3	7 months
Female, 64	12	10(5+5)	5	54	4(2+2)	2	20 months
Female, 36	60	2(2+0)	1	60	0(0+0)	0	9 months
Female, 66	0	10(5+5)	5	60	2(2+0)	1	24 months

RESULTS

Subject	Change in MRC ss	INCAT improvement	mRS improvement
Male, 65	1	0	0
Male, 72	8	0	0
Female, 64	42	6	3
Female, 36	0	2	1
Female, 66	60	8	4

DISCUSSION

- 3/5 subjects showed improvement in response to rituximab. This included improvement at 9 to 24 months in:
  - Average MRC sum score increase: 22 (range 0-60)
  - Average decrease in INCAT: 3.2 (2-6)
  - Median Modified Rankin Scale (mRS) decreased from 3 to 2
- Limitations include lack of antibody testing, single site, abstraction of INCAT, concomitant therapies, and missing data.

Literature Review

Literature Review	No of subjects	No improved	Follow up period
Knecht, 2004	1	1	7 months
Munch, 2007	1	1	4 weeks
Gorson, 2007	2	1	4 weeks
Rose, 2010	1	1	4 weeks
Sadnicka, 2011	1	1	4 weeks
Benedetti, 2011	13	6	4 weeks
Velardo 2017	2	2	4 weeks
Querol 2017	4	3	18 months
Roux 2018	28	21	0.75-9 years

CONCLUSIONS

- Rituximab may be a treatment option for some CIDP patients whose symptoms are refractory to more conventional treatments.

- Further investigation with a randomized controlled trial of B-cell depletion for treatment of CIDP may help to determine the full effect.