

Supplementary Table S3. Statistical analysis to assess any differences among the patients who underwent nerve biopsy based on the clinical phenotype.

| | Atypical (n=13) | | | Typical (n=30) | | | FET | CS | MWUT |
|--|-----------------|---------------|----|----------------|---------------|----|-----|----|------|
| | count (%) | mean (SD) | n | count (%) | mean (SD) | n | | | |
| Age at the time of nerve biopsy (years) | | 51.46 (17.92) | 13 | | 51.97 (21.20) | 30 | | | NS |
| Time from disease onset to nerve biopsy (months) | | 74.31 (91.69) | 13 | | 66.47 (87.02) | 30 | | | NS |
| Final histopathological findings | | | 13 | | | 30 | | NS | |
| Demyelinating | 0 (0%) | | | 2 (6.7%) | | | | | |
| Mixed | 10 (76.9%) | | | 19 (63.3%) | | | | | |
| Axonal | 2 (15.4%) | | | 7 (23.3%) | | | | | |
| Normal | 1 (7.7%) | | | 2 (6.7%) | | | | | |
| Evidence of demyelination by semithin sections and/or teased fibre analysis | | | 13 | | | 30 | | NS | |
| Absent | 2 (15.4%) | | | 9 (30.0%) | | | | | |
| Only by teased fibre analysis | 2 (15.4%) | | | 4 (13.3%) | | | | | |
| Only by semithin sections analysis | 4 (30.8%) | | | 3 (10.0%) | | | | | |
| Both by semithin sections and teased fibre analysis | 5 (38.5%) | | | 11 (36.6%) | | | | | |
| Not evaluable | 0 (0%) | | | 3 (10.0%) | | | | | |
| Macrophage-mediated demyelination (TEM) | 1 (7.7%) | | 13 | 0 (0%) | | 30 | NS | | |
| Evidence of remyelination by teased fibre analysis | 3 (23.1%) | | 13 | 6 (20.0%) | | 30 | NS | | |
| Onion bulbs | 1 (7.7%) | | 13 | 7 (23.3%) | | 30 | NS | | |
| Loss of fibres | | | 13 | | | 30 | | NS | |
| Absent | 1 (7.7%) | | | 6 (20.0%) | | | | | |
| Mild | 3 (23.1%) | | | 7 (23.3%) | | | | | |
| Moderate | 7 (53.8%) | | | 4 (13.3%) | | | | | |
| Severe | 2 (15.4%) | | | 13 (43.3%) | | | | | |
| Axonal degeneration | 8 (61.5%) | | 13 | 14 (46.7%) | | 30 | NS | | |
| Regeneration clusters | 8 (61.5%) | | 13 | 18 (60.0%) | | 30 | NS | | |
| Inflammatory infiltrates | 1 (7.7%) | | 13 | 1 (3.3%) | | 30 | NS | | |
| Vasa nervorum abnormalities | 0 (0%) | | 13 | 0 (0%) | | 30 | NP | | |
| Intramielinic edema | 1 (7.7%) | | 13 | 2 (6.7%) | | 30 | NS | | |
| Endoneural or subepineural edema | 2 (15.4%) | | 13 | 3 (10%) | | 30 | NS | | |

Legend to the table: Abbreviations: SD, standard deviation. FET, Fishers' exact test; CS, Chi-squared test; MWUT, Mann-Whitney U Test; TEM, Transmission Electron Microscope.