

| Table e-1: Summary of the clinical features, laboratory findings, neurophysiological results, response to treatment and cause of death | | | | | | | | | | | | |
|----------------------------------------------------------------------------------------------------------------------------------------|-------------------------|-------------------------|------------------|--------------|--------------|------------------------------------------------------|---------------------------------------------------|--------------------------|-------------------------------------|--------------|-------------------------------------------------------------|-------------------------------------------------|
| Case | Sex/age at onset, years | Disease duration, years | Sensory deficits | Weakness | Bulbar signs | Upper motor neuron signs | Laboratory findings | Blink reflex | EMG signs denervation/reinnervation | | (Response to) treatment | Cause of Death |
| 1 | ♂77 | 4 | F | UE | + | - | CK 176 U/L, ANA positive | Absent bilaterally | B, C, LS | B, C | No treatment | Unknown |
| 2 | ♂47 | 14 | F, UE, LE | UE | + | - | CK 351 U/L | Absent R1 left | - | B | No treatment | - |
| 3 | ♂47 | 3 | F | Neck, UE | + | - | Homocysteine 16.5 µmol/L, LDL 4.7 mmol/L | Variable bilaterally | B, C | B, C | PSL, AZP, PDS, riluzole. PDS subjective effect | - |
| 4 | ♀58 | 10 | F, UE | Neck | + | - | ACE 74 U/L, antids-DNA slightly elevated | Absent bilaterally | - | C, LS | No treatment | Unknown |
| 5 | ♂40 | 23 | F, UE, UT | SG, UE, LE | + | Brisk reflexes LE + clonus | CK 650 U/L | Absent bilaterally | B, C, Th, LS | B, C, Th, LS | No treatment | Respiratory failure due to pneumonia |
| 6 | ♀44 | 4 | F | Neck, UE | + | - | Normal/negative | Delayed R2 bilaterally | B, C | B | No treatment | - |
| 7 | ♂54 | 8 | F, UE | UE | + | - | Normal/negative | Delayed bilaterally | C | - | No treatment | - |
| 8 | ♂47 | 26 | F, UT, UE | UE | + | - | Normal/negative | Delayed bilaterally | C, LS | C, LS | No treatment | Pneumonia/sepsis |
| 9 | ♂70 | 3 | F | - | + | - | Normal/negative | Absent bilaterally | B | B | No treatment | Lung cancer |
| 10 | ♀46 | 16 | F | - | + | Spasticity LE, hyperreflexia LE, plantar indifferent | HIV positive (under treatment), HepA/HepB treated | Pathological bilaterally | B, C | - | Parotis radiation for salivation/dysphagia with some effect | - |
| 11 | ♂74 | 4 | F | SG, UE, LE | - | Hyperreflexia LE | Normal/negative | Pathological bilaterally | B, C | - | IVIg with no effect | Respiratory failure due to pulmonary aspiration |
| 12 | ♂44 | 10 | F | - | + | - | Normal/negative | Absent bilaterally | B, C | B, C | No treatment | Respiratory failure due to pulmonary aspiration |
| 13 | ♂64 | 14 | LE | No | + | - | Normal/negative | Absent bilaterally | B | B, C, LS | No treatment | Unknown |
| 14 | ♂63 | 13 | F | Neck | + | - | Normal/negative | Altered bilaterally | C, LS | B, C, LS | No treatment | - |
| 15 | ♂57 | 7 | F, UE | UE | + | - | Normal/negative | Delayed bilaterally | B, C, Th, LS | B, C Th, LS | PSL, R with no effect | - |
| 16 | ♀63 | 4 | F, UE | UE, LE | + | - | Normal/negative | Absent bilaterally | B, C, Th, LS | B, C Th, LS | IVIg with no effect | - |
| 17 | ♂52 | 5 | F, UE | UE, LE | + | Babinski | Normal/negative | Delayed bilaterally | B, C, Th, LS | B, C Th, LS | IVIg, R with no effect | - |
| 18 | ♀78 | 8 | F | Neck, SG, UE | + | - | Normal/negative | Absent left | B,C, Th | B, C, Th | No treatment | - |
| 19 | ♂61 | 2.5 | F | Neck, SG, UE | + | Hyperreflexia UE, positive | Normal/negative | Delayed bilaterally | B, C | B, C | No treatment | - |

| | | | | | | | | | | | | |
|----|-------|-----|-----------|----------|---|---------|-----------------|----------------------------------------|-------------|-------------|---------------------------------|-----------------------|
| 20 | ♀53 | 20 | F, UE | Neck, SG | + | HT - | Normal/negative | Absent R1 bilaterally, with delayed R2 | B, C, Th | B, C, Th | No treatment | - |
| 21 | ♂59 | 6 | F, UE, LE | UE, LE | + | - | Normal/negative | Normal CR decreased bilaterally | N.M. | N.M. | No treatment | - |
| 22 | ♂57 | 12 | F, UE | Neck, UE | + | + | Normal/negative | Absent bilaterally | B, C, Th | N.M. | IVIg with no effect | Respiratory failure |
| 23 | ♀41 | 12 | F, UE, LE | Neck, UE | + | + | Normal/negative | Absent bilaterally | B, C Th, LS | N.M. | No treatment | Traumatic head injury |
| 24 | ♀48 | 6 | F, UE | Neck, UE | + | + | Normal/negative | Low R1, absent R2 | B, C Th, LS | N.M. | IVIg, MM, CPA, R with no effect | Respiratory failure |
| 25 | ♀52.5 | 5.5 | F, UE, LE | Neck, UE | + | + | Normal/negative | Absent bilaterally | B, C Th, LS | N.M. | No treatment | Respiratory failure |
| 26 | ♂58 | 11 | F, UE | Neck, UE | + | + | Normal/negative | Lowered (normal range) R1, R2 delayed | B, C, Th | N.M. | No treatment | - |
| 27 | ♂54.5 | 7.5 | F, UE, LE | Neck, UE | + | + | Normal/negative | Absent bilaterally | B, C Th, LS | N.M. | No treatment | Respiratory failure |
| 28 | ♀48 | 18 | F, UE | Neck, UE | + | + | Normal/negative | Absent bilaterally | B, C Th, LS | N.M. | No treatment | Respiratory failure |
| 29 | ♂56 | 7 | F, LE | Neck, UE | + | - | Normal/negative | Delayed bilaterally | B, C Th, LS | B, C Th, LS | IVIg, AZP, PSL With no effect | - |

AZP= azathioprine; B= bulbar; C= cervical; CPA= cyclophosphamide; CR=corneal reflex; EMG= electromyography; F= face; HT=Hoffman-Trömner reflex, IVIg= intravenous immunoglobulin; LE= lower extremities; LS= lumbosacral; MM = mycophenolatmofetil; N.M.= not mentioned; PDS= pyridostigmine; PE= plasma exchange; PSL= prednisolone; R= rituximab; SG= shoulder girdle, Th= thoracic; UE= upper extremities; UT= upper trunk.