Normal brain imaging accompanies neuroimmunologically justified, autoimmune encephalomyelitis

**Supplemental data**

**Table e-1.** Additional neurological examination data of 11 patients with NINJA.

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| --- | --- | --- |
| **Case** | **Pyramidal weakness** | **Foveal flicker fusion threshold (up/down), Hz** |
| 1 | Muscle weakness and brisk DTR in four limbs with right extensor toe sign + | Right; red 13/12, green 15/14, yellow 16/15 Left; red 22/14, green 19/11, yellow 17/7 |
| 2 | Muscle weakness, spasticity, and brisk DTR in four limbs with right extensor toe sign + | Right; red 28/25, green 23/17, yellow 24/17 Left; red 22/17, green 23/18, yellow 24/19 |
| 3 | Muscle weakness, spasticity, and brisk DTR in four limbs with bilateral patellar clonus, bilateral ankle clonus, and right extensor toe sign + | ND |
| 4 | Right dominant muscle weakness and brisk DTR in four limbs with right ankle clonus | ND |
| 5 | Left dominant muscle weakness, spasticity, and brisk DTR in four limbs with bilateral extensor toe sign + | Right; red 32/33, green 34/33, yellow 35/33 Left; red 29/32, green 28/30, yellow 25/29 |
| 6 | Muscle weakness, spasticity, and brisk DTR in both lower limbs | Right; red 24/28, green 23/29, yellow 25/29 Left; red 27/26, green 24/30, yellow 24/31 |
| 7 | Muscle weakness and brisk DTR in four limbs | Right; red 21/19, green 23/20, yellow 26/21 Left; red 37/35, green 34/30, yellow 35/31 |
| 8 | Left dominant muscle weakness and brisk DTR in four limbs with left Hoffmann’s sign + | Right; red 3/3, green 4/3, yellow 5/4 Left; red 5/6, green 7/5, yellow 5/5 |
| 9 | Muscle weakness and brisk DTR in four limbs with left extensor toe sign + | ND |
| 10 | Left dominant muscle weakness and brisk DTR in four limbs | Right; red 18/15, green 21/19, yellow 20/18 Left; red 19/17, green 21/16, yellow 23/21 |
| 11 | Left dominant muscle weakness and brisk DTR in four limbs with left Hoffmann’s reflex + | Right; red 14/14, green 22/24, yellow 27/25 Left; red 22/23, green 29/28, yellow 27/26 |

Abbreviations: DTR = deep tendon reflex; ND = not done; NINJA = normal-appearing imaging-associated, neuroimmunologically justified, autoimmune encephalomyelitis.

**Table e-2.** Additional clinical data of 11 patients with NINJA.

|  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Case** | **Autoantibody** | **CSF cell counts** | **CSF protein, mg/dl** | **CSF IgG index** | **CSF MBP, pg/ml** | **CSF OB** | **SSEP** | **VEP** | **Efficacy of IFN-ß** | **Immunomodulating drugs at blood sampling** |
| 1 | Negative | 1 | 28 | 0.68 | 40 | Negative | Extension of CSCT | Extension of P100 | Non-responder | PSL 15.5 mg/day + BUC 200 mg/day |
| 2 | Negative | 1 | 33 | ND | 121 | Negative | ND | ND | Non-responder | TCM 3 mg/day |
| 3 | Negative | 1 | 21 | 0.43 | < 40 | Negative | ND | ND | ND | PSL 20 mg/day + TCM 3 mg/day |
| 4 | Anti-TPO antibody + | 1 | 27 | 0.53 | 40 | Negative | W.N.L | W.N.L | ND | PSL 4 mg/day + TCM 1.5 mg/day |
| 5 | Negative | 1 | 55 | 0.57 | < 40 | Negative | W.N.L | W.N.L | ND | GA 20 mg/day + PSL 10 mg/day + MTX 4 mg/week + BUC 300 mg/day |
| 6 | Negative | ND | ND | ND | ND | ND | ND | ND | ND | PSL 5 mg/day |
| 7 | Negative | 2 | 43 | 0.56 | < 40 | Negative | W.N.L | Extension of P100 | ND | PSL 12.5 mg/day |
| 8 | Negative | 0 | 22 | 0.63 | < 40 | Negative | W.N.L | W.N.L | ND | PSL 10 mg/day + MZR 75 mg/day |
| 9 | Negative | 1 | 35 | 0.53 | ND | ND | Extension of CSCT | W.N.L | Leukopenia | GA 20 mg/day |
| 10 | Negative | 1 | 28 | ND | < 40 | Negative | W.N.L | W.N.L | ND | PSL 11 mg/day + AZA75 mg/day + CyA 350 mg/day |
| 11 | Negative | 1 | 29 | ND | < 40 | Negative | W.N.L | W.N.L | ND | PSL 11 mg/day + AZA 25 mg/day + BUC 200 mg/day |

Abbreviations: AZA = azathioprine; BUC = bucillamine ; CSCT; central somatosensory conduction time; CSF = cerebral spinal fluid; CyA = cyclosporin A; GA = glatiramer acetate; IFN-ß = interferon-ß; IgG = Immunoglobulin G; MBP = myelin basic protein; MTX = methotrexate; MZR = mizoribine; ND = not done; NINJA = normal-appearing imaging-associated, neuroimmunologically justified, autoimmune encephalomyelitis; OB = oligoclonal bands; PSL = prednisolone; P100 = P100 latency; SSEP = somatosensory evoked potential; TCM = tacrolimus; TPO = thyroid peroxidase; VEP = visual evoked potential; W.N.L = within normal limit.

**Figure e-1.** Changes in EDSS within the past 5 years.

Among seven patients in whom we could evaluate the changes in expanded disability status scale (EDSS) within the past 5 years, (A) EDSS increased in four patients, (B) did not change in two patients, and (C) decreased in one patient over time.