**Table e-1. Detailed clinical features of patients with antibodies to Nfasc155, Nfasc140/186, CNTN1, Caspr1**

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Patient** | **Case 1** | **Case 2** | **Case 3** | **Case 4** | **Case 5** | **Case 6** | **Case 7** | **Case 8** | **Case 9** | **Case 10** |
| **Ab** | Nfasc155 | Nfasc155 | Nfasc155 | Nfasc155 | Nfasc155 | Nfasc155 | Nfasc155 | Nfasc155 and Nfasc140/186 | Nfasc155 | Nfasc155 |
| **Positive test** | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA | ELISA, CBA |
| **IgG isotype** | IgG4 | IgG4 | IgG4 | IgG4 | IgG4 | IgG4 | IgG4 | Nfasc155: undetectable  Nfasc140/186: IgG4 | IgG3 | undetectable |
| **Other Abs** | None | None | IgM anti-GM1 (low titre) | None | None | anti-GA1 and GM2 | None | None | None | None |
| **Demographic** | 18 y, male | 63 y, female | 45 y, male | 36 y, female | 13 y, female | 19 y, male | 22 y, male | 2 y, male | 56 y, female | 74 y, male |
| **Comorbidities** | None | NHL, Basedow disease, diabetes | Diabetes | Spinocellular carcinoma of the skin | None | Asperger syndrome | None | None | Dyslipidemia | Hypertension |
| **Mode of onset** | Chronic | Acute | Subacute | Subacute | Subacute | Chronic | Subacute | Subacute | Chronic | Chronic |
| **Chief complain** | Distal numbness, walking difficulties,unsteadiness | Walking difficulties and unsteadiness | Ataxia, distal weakness and tremor | Lower limbs touch loss and distal weakness | Lower limbs distal weakness | Distal weakness | Distal lower limbs weakness and numbness, upper limbs tremor | Gait Instability | Distal paresthesia | Distal upper and lower limbs paresthesia |
| **Weakness**  **UL proximal** | Normal | + | Normal | Normal | + | + | Normal | Normal | + | + |
| **Weakness**  **UL distal** | ++ | ++ | + | + | + | ++ | Normal | Normal | ++ | + |
| **Weakness**  **LL proximal** | ++ | +++ | Normal | Normal | + | + | Normal | ++ | + | Normal |
| **Weakness**  **LL distal** | +++ | +++ | +++ | + | ++ | +++ | +++ | +++ | ++ | + |
| **Sensory ataxia** | + | + | + | + | + | + | - | - | + | + |
| **Tremor** | + | + | + | + | + | + | + | - | - | - |
| **Pain** | - | + | - | - | - | - | - | - | - | - |
| **Cranial nerve involvement** | - | - | - | - | - | - | + (Dysphagia) | - | - | - |
| **CNS involvement or additional features** | - | - | - | - | - | Bilateral optic neuritis | - | - | - | - |
| **ONLS upper limbs** | 2 | 3 | 3 | 4 | 2 | 3 | 2 | 0 | 3 | 2 |
| **ONLS lower limbs** | 3 | 6 | 3 | 3 | 2 | 5 | 1 | 4 | 2 | 2 |
| **ONLS total** | 5 | 9 | 6 | 4 | 4 | 8 | 3 | 4 | 5 | 4 |
| **ONLS total follow-up** | 5 | 2 | 2 | 4 | 2 | 8 | 3 | 1 | 3 | 2 |
| **EFNS/PNS criteria** | Definite CIDP | Definite CIDP | Definite  CIDP | Definite  CIDP | Probable  CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite  CIDP |
| **Response to IVIg** | Partial/transitory | Partial/transitory | Partial/transitory | Partial/transitory | Yes | No | Not performed | Yes | Yes | Yes |
| **Response to steroids** | Partial/transitory | Partial/transitory | Yes | Partial/transitory | Yes | Partial/transitory | Partial/transitory | No | Partial/transitory | Not performed |
| **Response to other treatment** | No (AZA) | Yes (Rituximab) | Not performed | Not performed | Not performed | Not performed | Not performed | Not performed | Not performed | Not performed |
| **Disease course** | Relapsing | Relapsing | Relapsing | Relapsing | Relapsing | Progressive | Progressive | Relapsing | Relapsing | Relapsing |

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Patient** | **Case 11** | **Case 12** | **Case 13** | **Case 14** | **Case 15** | **Case 16** | **Case 17** | **Case 18** | **Case 19** |
| **Ab** | CNTN1 | CNTN1 | CNTN1 | CASPR1 | CASPR1 | CASPR1 | CASPR1 | CASPR1 | CASPR1 |
| **Positive test** | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | ELISA, CBA, IHC (paranode) | CBA, IHC (paranode) | CBA, IHC (paranode) | CBA, IHC (paranode) | CBA, IHC (paranode) | CBA, IHC (paranode) |
| **IgG isotype** | IgG4 | IgG3/IgG4 | IgG4 | IgG4 | IgG4 | IgG4 | IgG1 | IgG1 | Undetectable |
| **Other Abs** | Anti-GD1A, GD1B, GQ1B (1:640) | None | None | None | None | None | None | Anti-MAG (low titre) | None |
| **Demographic** | 30 y, male | 58 y, male | 82 y, male | 24 y, male | 46 y, male | 57 y, female | 7 y, female | 67 y, male | 10 y, female |
| **Comorbidities** | None | Membranous glomerulonephritis, Basedow disease, cervical and lumbar stenosis | Hypertension, ischemic cardiopathy | None | None | Previous melanoma, multinodular dysthyroidism | Early pubarche | Hypertension, alcohol abuse, psoriasis | None |
| **Mode of onset** | Subacute | Subacute | Chronic | Chronic | Subacute | Subacute | Subacute | Chronic | Chronic |
| **Chief complain** | Lower limbs distal weakness and numbness | Paresthesia at four limbs, paraparesis, sensory ataxia | Proximal and distal weakness of four limbs, distal numbness | Weakness and paresthesia | Distal weakness | Lower limbs proximal weakness | Proximal and distal weakness, postural and intentional tremor, mild ataxia | Gait ataxia | Gait disturbances |
| **Weakness**  **UL proximal** | ++ | + | + | ++ | Normal | ++ | Normal | Normal | Normal |
| **Weakness**  **UL distal** | +++ | Normal | Normal | +++ | +++ | +++ | Normal | + | Normal |
| **Weakness**  **LL proximal** | ++ | +++ | +++ | ++ | +++ | ++ | ++ | Normal | ++ |
| **Weakness**  **LL distal** | +++ | +++ | +++ | ++ | +++ | +++ | +++ | + | +++ |
| **Sensory ataxia** | + | + | + | - | + | - | + | + | - |
| **Tremor** | + | - | - | - | - | - | + | - | + |
| **Pain** | - | + | + | - | - | - | - | - | - |
| **Cranial nerve involvement** | + | - | + | - | + (Dysphagia) | - | - | - | - |
| **CNS involvement or additional features** | - | - | - | - | Increased sweating and erectile dysfunction | - | - | - | - |
| **ONLS upper limbs** | 3 | 2 | 4 | 4 | 4 | 4 | 1 | 0 | 0 |
| **ONLS lower limbs** | 2 | 6 | 6 | 4 | 5 | 4 | 2 | 2 | 2 |
| **ONLS total** | 5 | 8 | 10 | 8 | 9 | 8 | 3 | 2 | 2 |
| **ONLS total follow-up** | 3 | 2 | 10 | 7 | 6 | 8 | 2 | 2 | 1 |
| **EFNS/PNS criteria** | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP | Definite CIDP |
| **Response to IVIg** | Partial/transitory | No | No | Partial/transitory | Partial/transitory | No | Yes | Not performed | Yes |
| **Response to steroids** | No | Partial/transitory | No | Partial/transitory | Yes | No | No | Not performed | Not performed |
| **Response to other treatment** | Yes (Cyclophosphamide) | Yes (PEX; Cyclophosphamide) | No (PEX) | No (PEX)  Yes (IFN-alfa) | Yes (Methotrexate) | Not performed | Not performed | Not performed | Not performed |
| **Disease course and outcome** | Progressive | Relapsing | Progressive | Relapsing | Relapsing | Not known | Relapsing | Progressive | Monophasic |

AZA azathioprine, CBA cell-based assay, CSF cerebro-spinal fluid, IFN-alfa interferon-alfa, IHC immunohistochemistry on rat teased nerve fibers, IVIg intravenous immunoglobulins, NCS nerve conduction studies, PEX plasma exchange, UL upper limbs, LL lower limbs, + present, - absent