**Supplementary Online Content**

Manuscript: “Clinical Spectrum and Diagnostic Pitfalls of Neurological Syndromes With Ri Antibodies”

**Supplementary Table1**. Clinical characteristics of the French Cohort

**Supplementary Table 2**: Case reports of anti-Ri-associated syndromes published in the literature

**Supplementary Table 3**: Case series of anti-Ri-associated syndromes published in the literature

**Supplementary Figure 1**: PRISMA Flow Diagram Describing Identification, Screening, Eligibility, and Inclusion of Studies in the Systematic Review

**Supplementary Table 1:** Clinical characteristics of 36 patients with anti-Ri antibodies identified at the French Reference Center for Paraneoplastic Neurological Syndromes (1999-2018)

|  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Patient | Age | Sex | First symptom | Second symptom | Third symptom | Mode of onset | Peculiar features | Initial diagnosis (other than PNS) | Cancer | Coexisting Abs |
| 1 | 78 | F | Opsoclonus-myoclonus | Spasticity | - | Acute | - | - | Breast | - |
| 2 | 54 | F | Tremor | Cerebellar syndrome | Oculomotor disturbances | Subacute | Jaw dystonia | Multiple sclerosis | Breast | - |
| 3 | 52 | F | Stiff-person syndrome | Myoclonus | -  | Progressive | - | - | -  | GAD |
| 4 | 52 | F | Cerebellar syndrome | Opsoclonus-myoclonus | Oculomotor disturbances | Subacute | - | - | Breast | - |
| 5 | 82 | F | Tremor | Cerebellar syndrome | Oculomotor disturbances | Subacute | - | - | Breast | - |
| 6 | 83 | F | Cerebellar syndrome | Oculomotor disturbances | -  | Subacute | - | - | - | - |
| 7 | 71 | M | Confusion |  - |  - | Acute | - | - | Neuroendocrine carcinoma probably of lung origin | - |
| 8 | 56 | F | Tremor | Stiff-person syndrome |  - | Progressive | Hyperekplexia | Genetic hyperekplexia | Breast | - |
| 9 | 79 | M | Tremor | Peripheral neuropathy |  - | Subacute | SIADH |  - | SCLC | - |
| 10 | 67 | F | Oculomotor disturbances | - | - | Acute | - | - | Breast | - |
| 11 | 61 | F | Parkinsonism | Oculomotor disturbances |  - | Subacute | - | Atypical parkinsonism | Breast | - |
| 12 | 77 | F | Cerebellar syndrome | Opsoclonus-myoclonus | -  | Progressive | - | - | Breast (biopsy not performed) | - |
| 13 | 70 | F | Oculomotor disturbances | Dystonia | Cerebellar syndrome | Progressive | Jaw dystonia | - | Breast | - |
| 14 | 76 | F | Oculomotor disturbances | - | - | Subacute | Excessive Daytime Sleepiness | Bickerstaff's encephalitis | Breast | - |
| 15 | 61 | F | Tremor | Cerebellar syndrome | Opsoclonus-myoclonus | Progressive | - | Functional disorder | Breast | Hu |
| 16 | 78 | M | Cerebellar syndrome | Oculomotor disturbances |  - | Subacute | - |   | Urothelial  | - |
| 17 | 87 | M | Cerebellar syndrome | Oculomotor disturbances | - | Subacute | - | - | Cancer (biopsy not performed) | - |
| 18 | 72 | F | Cerebellar syndrome | Oculomotor disturbances | - | Subacute | SIADH, fluctuant diplopia | - | Angiosarcoma | - |
| 19 | 77 | M | Confusion | - | - | Acute | SIADH | - | Cancer (biopsy not performed) | - |
| 20 | 75 | F | Tremor | Cerebellar syndrome | Oculomotor disturbances | Subacute | Ondine's syndrome | - | Breast | - |
| 21 | 54 | F | Oculomotor disturbances | Cerebellar syndrome |  - | Subacute | Dramatic improvement with CTx | - | Breast | - |
| 22 | 64 | F | Oculomotor disturbances | Parkinsonism | Dystonia | Progressive | Freezing of gait | PSP | Breast | - |
| 23 | 64 | F | Cerebellar syndrome | Oculomotor disturbances | Dystonia | Progressive | Laryngospasm | - | Breast  | - |
| 24 | 66 | F | Tremor | Cerebellar syndrome | Spasticity | Subacute | - | - | Breast | - |
| 25 | 78 | M | Cerebellar syndrome | -  | -  | Subacute | - | Multiple sclerosis | SCLC | GAD and SOX |
| 26 | 59 | F | Cerebellar syndrome | Opsoclonus |  - | Progressive | - | - | Breast | - |
| 27 | 47 | F | Cerebellar syndrome | Oculomotor disturbances | Spasticity | Subacute | - | Vestibular neuritis | Breast | - |
| 28 | 50 | M | Limbic encephalitis |  - |  - | Progressive |  | - | Mediastinal seminoma | - |
| 29 | 70 | F | Cerebellar syndrome | -  | -  | Subacute |  - | - | Oesophageal | - |
| 30 | 67 | F | Cerebellar syndrome | Oculomotor disturbances | Dystonia | Progressive | Ondine's syndrome | - | SCLC | - |
| 31 | 56 | M | Tremor | Cerebellar syndrome | Oculomotor disturbances | Subacute | - | - | Neuroendocrine carcinoma of the urinary bladder | - |
| 32 | 63 | F | Tremor | Cerebellar syndrome | Parkinsonism | Subacute | - | - | Breast | - |
| 33 | 62 | F | Opsoclonus-myoclonus | Spasticity |  - | Subacute | - | - | Breast | - |
| 34 | 71 | F | Cerebellar syndrome | Opsoclonus | Spasticity | Subacute | - | - | Breast | - |
| 35 | 59 | F | Oculomotor | Cerebellar syndrome | Tremor | Subacute | - | - | - | - |
| 36 | 50 | F | Cerebellar syndrome |  - | -  | Subacute | - | - | Breast | - |

Abbreviations: CTx, chemotherapy; F, female; M, male; PNS, paraneoplastic syndrome; PSP, progressive supranuclear palsy; SIADH, syndrome of inappropriate antidiuretic hormone secretion; SCLC, small cell lung cancer

**Supplementary Table 2**: Case reports of anti-Ri-associated syndromes published in the literature

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| 7 | Sex | Age at symptoms’ onset(years) | Confirmation with Western blot or Immunodot | Cancer | Reference |
| 1 | F | 58 | Yes | + |  (Budde-Steffen et al., 1988) |
| 2 | F | 45 | Yes | - | (Dropcho et al., 1993) |
| 3 | F | 59 | Yes | + | (Escudero et al., 1993) |
| 4 | F | 57 | Yes | - | (Casado et al., 1994) |
| 5 | F | 73 | Yes | - |  (Hormigo et al., 1994) |
| 6 | F | 58 | Yes | + | (Jongen et al., 1998) |
| 7 | F | 60 | Not specified | + | (Ohmer et al., 1999) |
| 8 | M | 57 | Not specified | + | (Prestigiacomo et al., 2001) |
| 9 | F | 65 | Non specified | + | (Bataller et al., 2001) |
| 10 | M | 59 | Yes | + | (Kastrup et al., 2001) |
| 11 | M | 65 | Yes | + | (Wirtz et al., 2002) |
| 12 | F | 68 | Yes | + | (Sutton et al., 2002) |
| 13 | F | 43 | Yes | + | (McCabe et al., 2004) |
| 14 | F | 71 | Yes | + | (Rajabally et al., 2004) |
| 15 | F | 79 | Not specified | + | (Weizman and Leong, 2004) |
| 16 | F | 33 | No | + | (Fadare and Hart, 2004) |
| 17 | M | 75 | Yes | - | (Romorini et al., 2004) |
| 18 | F | 68 | Yes | + | (Harloff et al., 2005) |
| 19 | F | 65 | Yes | - | (Leypoldt, 2006) |
| 20 | F | 54 | Not specified | + | (Martinaud et al., 2005) |
| 21 | M | 71 | Yes | - | (Fumal et al., 2006) |
| 22 | F | 66 | Yes | + | (Stich, 2006) |
| 23 | F | 71 | Not specified | + | (Brieva-Ruíz et al., 2008) |
| 24 | M | 49 | Yes | + | (Launay et al., 2008) |
| 25 | M | 68 | Yes | + | (Vigliani et al., 2009) |
| 26 | F | 68 | Not specified | + | (Tesseki et al., 2010) |
| 27 | M | 52 | No | + |  (Kim et al., 2009) |
| 28 | F | 85 | Not specified | + | (White and Beringer, 2010) |
| 29 | F | 63 | Yes | + | (Thümen and Moser, 2010) |
| 30 | F | 54 | Yes | + | (Ducray et al., 2010) |
| 31 | F | 80 | No | + | (Butt et al., 2013) |
| 32 | F | 55 | Not specified | + | (Bekircan-Kurt et al., 2013) |
| 33 | F | 67 | Not specified | + | (Tay et al., 2012) |
| 34 | F | 54 | Not specified | + | (Kim et al., 2013) |
| 35 | F | 49 | Not specified | + | (Younger et al., 2013) |
| 36 | F | 75 | Yes | + | (Mitchell et al., 2015) |
| 37 | F | 80 | Not specified | + | (Diard-Detoeuf et al., 2014) |
| 38 | F | 40 | Yes | + | (Boch et al., 2014) |
| 39 | F | 69 | Not specified | + | (O’Leary et al., 2017) |
| 40 | F | 76 | Not specified | + | (Goyal et al., 2016) |
| 41 | F | 66 | Yes | + | (Díaz et al., 2016) |

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| Number of cases | Sex | Median Age at symptoms’ onset (years) | Confirmed by Western blot | Reference |
| 8 | 8 F | 60,5 | Yes | (Luque et al., 1991) |
| 2 | Not specified | 54 | Yes | (Stourac et al., 2001) |
| 31 | 22 F9 M | 67.5 | Yes | (Pittock et al., 2003) |
| 3 | 3 F | 65 | Not specified | (Jarius et al., 2008) |
| 2 | 2 F | 56 | Yes | (Michalak et al., 2009) |
| 2 | 2 F | 61.5 | Yes | (Pittock et al., 2010) |

 **Supplementary Table3:** Case series of anti-Ri-associated syndromes published in the literature

**Supplementary Figure 1**: PRISMA Flow Diagram Describing Identification, Screening, Eligibility,

and Inclusion of Studies in the Systematic Review

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