**Supplementary Table 1.** Comparison of discriminatory power of CSF biomarkers to distinguish atypical AD from typical CJD.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Biomarker** | **Area under the curve****(AUC)** | **Cut-off value for CJD diagnosis** | **Patients diagnosed as having CJD** | **Sensitivity****%** | **Specificity****%** |
|  | **Typical CJD** | **a/rp AD** |  |
| **t-tau** | 0.980±0.011 | >1200 pg/ml | 54/54 | 22/44 | 100 | 50.0 |
| **14-3-3** | - | - | 54/54 | 7/44 | 100 | 84.1 |
| **Aβ42 /p-tau** | 0.921±0.027 | >4.68 | 45/54 | 7/44 | 83.3 | 84.1 |
| **Aβ42 /(p-tau × PrP)** | 0.941±0.023 | >0.020 | 46/54 | 6/44 | 85.2 | 86.4 |
| **t-tau/p-tau** | 0.996±0.004 | >25.7 | 53/54 | 1/44 | 98.1 | 97.7 |
| **t-tau/t-PrP** | 0.968±0.014 | >7.79 | 47/54 | 4/44 | 88.9 | 90.9 |
| **Aβ42 × t-tau/p-tau** | 0.999±0.001 | >9522 | 54/54 | 1/44 | 100 | 97.7 |
| **t-tau/(p-tau × t-PrP)** | 0.989±0.007 | >0.107 | 52/54 | 2/44 | 96.3 | 95.5 |
| **(t-tau × Aβ42)/(p-tau × t-PrP)** | 0.997±0.002 | >37.5 | 53/54 | 1/44 | 98.1 | 97.7 |

Statistically significant differences: t-tau (p<0.001), Aβ42 (p<0.001), t-tau/p-tau ratio (p<0.001), t-tau/t-PrP (p<0.001), Aβ42/p-tau (p<0.001), Aβ42/(p-tau × t-PrP) (p<0.001), Aβ42 × t-tau /p-tau (p<0.001), CJ factor (p<0.001), (Aβ42 × t-tau)/(p-tau × t-PrP) (p<0.001).

**Supplementary Table 2.** Comparison of discriminatory power of CSF biomarkers to distinguish typical AD from atypical CJD.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Biomarker** | **Area under the curve****(AUC)** | **Cut-off value for CJD diagnosis** | **Patients diagnosed as having CJD** | **Sensitivity****%** | **Specificity****%** |
|  | **Atypical CJD** | **Typical AD** |  |
| **t-tau** | 0.865 ±0.036 | >1200\* | 33/54 | 0/45 | 61.1 | 100 |
| **14-3-3** | - | - | 14/54 | 0/45 | 25.9 | 100 |
| **Aβ42/p-tau** | 0.927±0.030 | >6.35 | 47/53 | 5/44 | 88.7 | 88.6 |
| **Aβ42/(p-tau × PrP)** | 0.976±0.013 | >0.024 | 49/53 | 2/44 | 92.5 | 95.5 |
| **t-tau/p-tau** | 0.985 ±0.015 | >10.9 | 51/53 | 1/44 | 96.2 | 97.7 |
| **t-tau/t-PrP** | 0.959 ±0.022 | >3.83 | 51/54 | 3/45 | 92.6 | 93.3 |
| **Aβ42 × t-tau/p-tau** | 0.979±0.016 | >4682 | 52/54 | 3/44 | 96.3 | 93.2 |
| **t-tau/(p-tau × t-PrP)** | 0.991±0.007 | >0.050 | 52/53 | 1/44 | 98.1 | 97.7 |
| **(t-tau × Aβ42) /(p-tau × t-PrP)** | 0.995±0.005 | >18.65 | 52/53 | 0/44 | 98.1 | 100 |

\*pg/ml. Statistically significant difference: t-tau (p<0.001), Aβ42 (p=0.001), t-tau/p-tau ratio (p<0.001), t-tau/t-PrP (p<0.001), Aβ42/p-tau (p<0.001), Aβ42/(p-tau × t-PrP) (p<0.001), Aβ42 × t-tau/p-tau (p<0.001), CJ factor (p<0.001), (Aβ42 × t-tau)/(p-tau × t-PrP) (p<0.001).