Supplementary Information

Hornerin deposits in neuronal intranuclear inclusion disease: Direct identification of proteins with compositionally biased regions in inclusions

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Separate excel files

Supplementary dataset 1
Supplementary dataset 2
Supplementary dataset 3
Supplementary Table

Table S1. The identified hornerin peptides by LC-MS/MS and LC-TIMS-MS/MS.

The peptides at 1,584 m/z were highlighted. See figure S2 for a map.

| Analysis | #   | Samples   | Sequence                                                                 | Number of repeats |
|----------|-----|-----------|--------------------------------------------------------------------------|-------------------|
| LC-MS/MS | 1   | FA fraction | GPYESGSGHSSGLGHR                                                        | 3                 |
|          | 2   |            | GPYESGSGHSSGLGHRQESR                                                   | 1                 |
|          | 3   |            | GPYESGSGHSSGLGHR                                                        | 3                 |
|          | 4   | P4 245 kDa | GSGQSPSSQHQHTGFGR                                                       | 1                 |
|          | 5   |            | HGSGHSSSYGQHGSGGWSSSGR                                                 | 3                 |
|          | 6   |            | HGSGQSSSYGPR                                                            | 1                 |
|          | 7   |            | HGSGQSSSYSPYGSYGSGGWSSSR                                               | 2                 |
|          | 8   |            | QSLGHQHGSGSGQSPSPSR                                                    | 4                 |
|          | 9   | P4 100 kDa | QSLGHQHGSGGWSSPSRPSPSPSR                                                | 4                 |
|          | 10  |            | GSGQSPSYGR                                                             | 1                 |
|          | 11  |            | SGSGWSSSR                                                              | 4                 |
|          | 12  | Gray matter 1 | QGSGSGQSPGHQ                                                            | 4                 |
|          | 13  | Gray matter 2 | YQQQGSGSQPSR                                                           | 3                 |
|          | 14  |             | SSSSSSYGQHGSGS                                                          | 5                 |
|          | 15  |             | QSSSYGPHYGSGR                                                           | 1                 |
|          | 16  | White matter 1 | GPYESGSGHSSGLGQESR                                                    | 1                 |
|          | 17  |             | YQQQGSGSQPSR                                                           | 3                 |
|          | 18  |             | GPYESGSGHSSGLGHR                                                       | 3                 |
|          | 19  |             | SSSSSSYGQHGSGS                                                          | 5                 |
|          | 20  |             | QSSSYGQHESASR                                                           | 1                 |
|          | 21  |             | GSGQSPSSQHQHTGFGR                                                       | 1                 |
|          | 22  |             | HGSGQPSQPSR                                                             | 5                 |
|          | 23  |             | QSSSYGPHYGSGR                                                           | 1                 |
|          | 24  |             | GPYESGSGHSSGLGQESR                                                    | 1                 |
|          | 25  |             | YQQQGSGSQPSR                                                           | 3                 |
|          | 26  | White matter 2 | SGSQSSYSGQHGSSGHSSGSR                                                  | 1                 |
|          | 27  |             | SEQHSSSSSYGQHGSGSR                                                     | 1                 |
|          | 28  |             | GPYESGSGHSSGLGHR                                                       | 3                 |
|          | 29  |             | SSSRGPYESR                                                             | 4                 |
|          | 30  |             | HGSGQSSSYSPYGSYGSGGWSSSR                                               | 2                 |
|          | 31  |             | GEQHSSSSSYGQHGSGSR                                                     | 1                 |
|          | 32  |             | HGSGQSSSYGPYGSYGSGGWSSSR                                               | 1                 |
|          | 33  |             | HGAGSQSLSHGR                                                            | 1                 |
Supplementary Figures

**Fig. S1** The calculated percentage of amino acids in the sequence of nHTT-EGFP

The calculated percentage of amino acid numbers in the sequences of nHTT-EGFP with expanded polyglutamine (190Q) and normal repeat size (21Q) expressed in HD190QG mice.
Fig. S2 The horserin peptides identified by LC-MS/MS and LC-TIMS-MS/MS

Every repeating peptide sequence is indicated with a particular color. The peptide numbers are listed in table S1.