## Suppl. Fig. 1

### IL-6 ROC curve
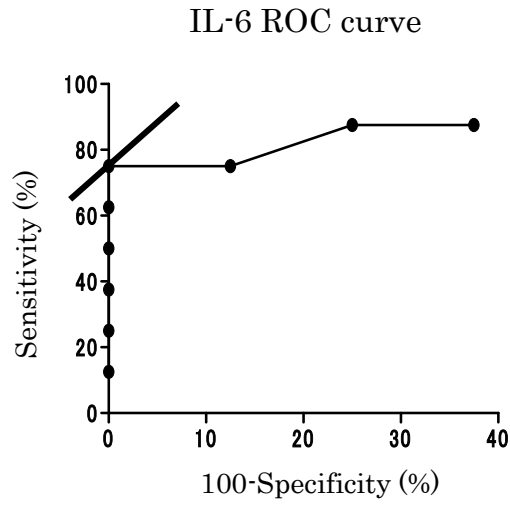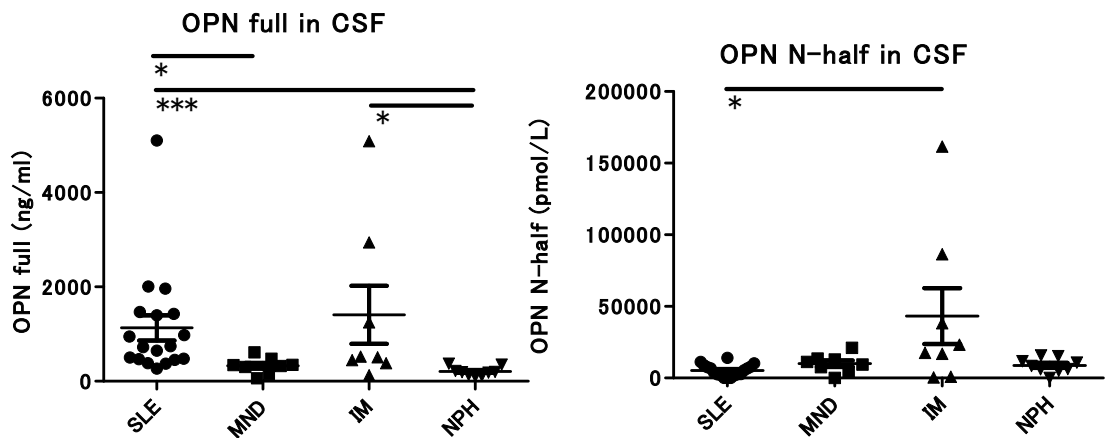


ROC curve with IL-6 in CSF to diagnose NPSLE. When the cut off value of OPN full in CSF was set as 74.9 pg/ml, sensitivity was 75.0% and specificity was 100% in the diagnosis of NPSLE. AUC: 0.89.

## Suppl. Fig. 2



Concentration of OPN full and OPN N-half in CSF of SLE and other neurological diseases. *p<0.05, ***p<0.001 by Dunn's multiple comparison test. IM: infectious meningoencephalitis, MND: motor neuron disease, NPH: normal pressure hydrocephalus.

## Suppl. Fig. 3
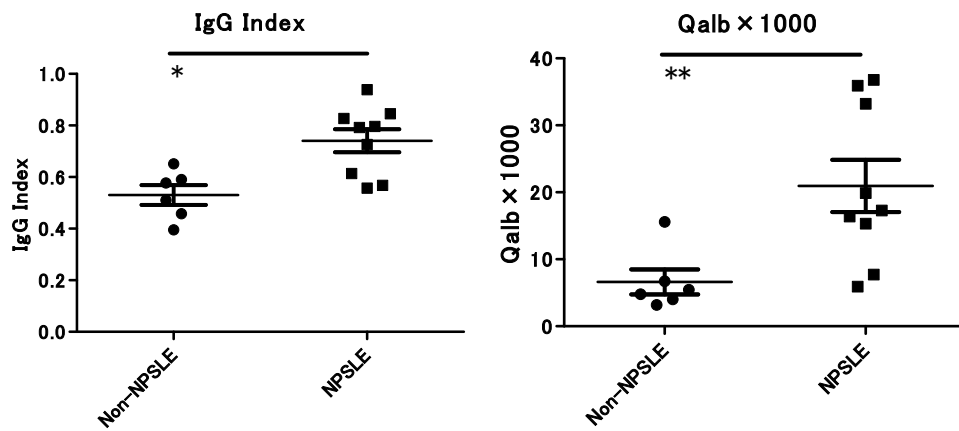


IgG index and Qalb in NPSLE. IgG index is calculated as ( [CSF IgG] / [CSF Alb] ) / ( [Serum IgG] / [Serum Alb] ). Qalb is calculated as ( [CSF Alb]/[Serum Alb] ). *p<0.05, **p<0.01 by Mann-Whitney U test.

## Suppl. Fig. 4
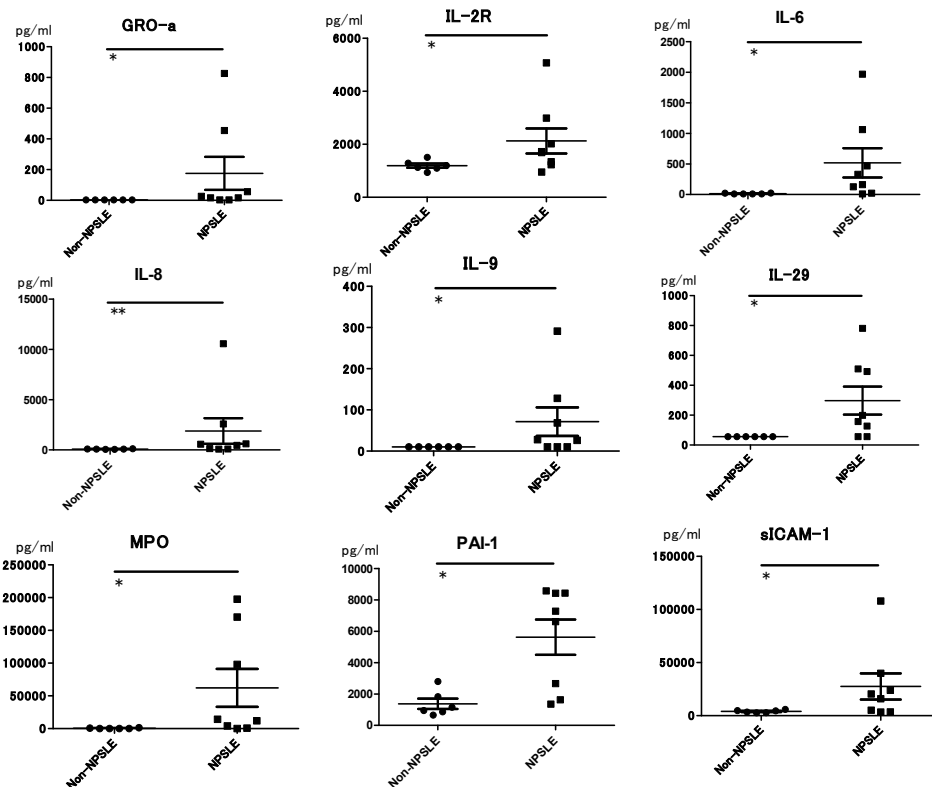


Cytokine and chemokine concentrations in CSF of non-NPSLE and NPSLE patients. In multiplex analysis, 41 of 67 molecules were measurable. Measurements of 9 molecules tended to be higher in NPSLE than in non-NPSLE patients. The p value for each Mann-Whitney U test (*p<0.05, **p<0.01) is shown. However, significance could not be determined because the null hypothesis could not be rejected when Bonferroni's correction was performed for repeated testing of 41 measurable molecules. GRO-a: growth related oncogene-alpha, IL: interleukin, MPO: myeloperoxidase, PAI-1: plasminogen activator inhibitor-1, sICAM-1: soluble intercellular adhesion molecule-1.

## Suppl. Fig. 5
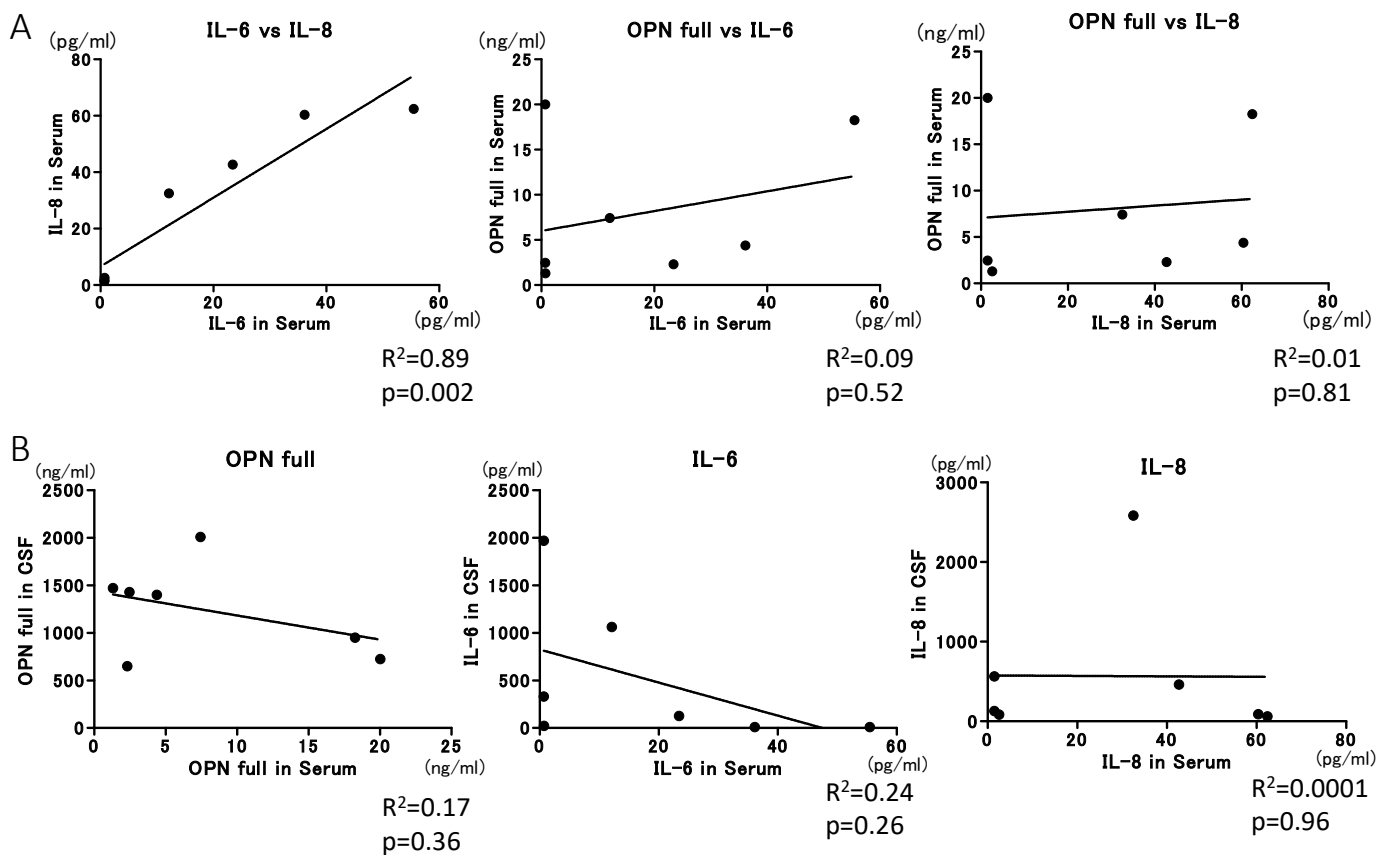


A: Correlation of IL-6, IL-8, and OPN full in sera of NPSLE and non-NPSLE by a linear progression analysis. There was a correlation between serum IL-6 and IL-8. However, serum OPN full did not correlate with serum IL-6 or IL-8.
B: Correlation of each cytokine level between serum and CSF. No significant correlations were found.

## Suppl. Fig. 6
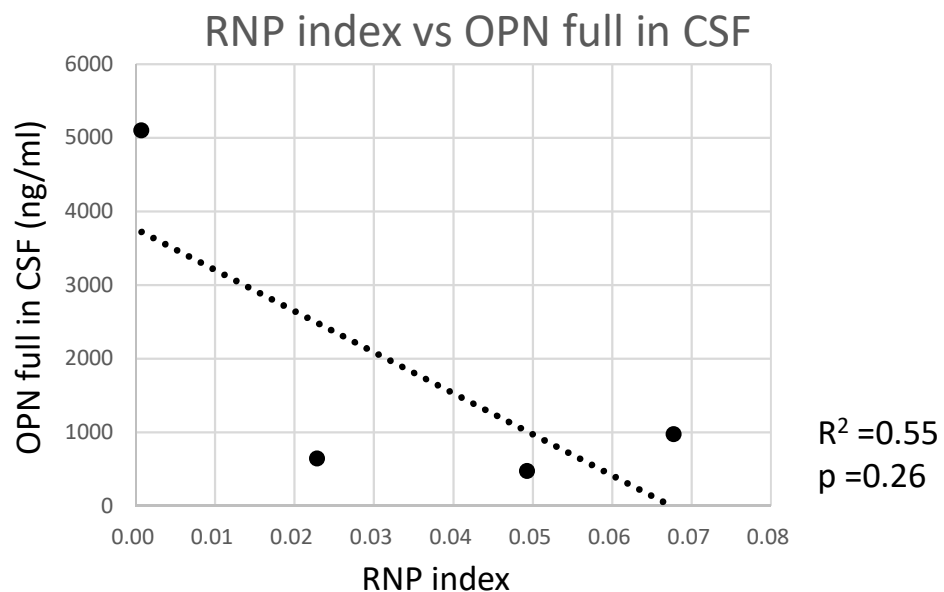


Correlation of OPN full in CSF with RNP index in NPSLE. In the present study, there were 4 patients whose anti-RNP antibody in CSF was positive. No significant correlation was found between OPN full in CSF and RNP index by a linear progression analysis. RNP index is calculated as ([CSF anti-U1 RNP antibodies/serum anti-U1 RNP antibodies]/[CSF IgG/serum IgG]), according to the study by Sato et al[20).

Suppl. Table 1. Classification of NPSLE

| NPSLE (N = 11) | ACR Classification of NPSLE* |
|---|---|
| No.1 | Psychosis, Seizure disorders |
| No.2 | Acute confusional state, Demyelinating syndrome, Seizure disorders |
| No.3 | Cognitive dysfunction, Mood disorder |
| No.4 | Acute confusional state |
| No.5 | Demyelinating syndrome, Polyneuropathy |
| No.6 | Acute confusional state, Demyelinating syndrome |
| No.7 | Headache |
| No.8 | Acute confusional state, Psychosis, Seizure disorders |
| No.9 | Aseptic meningitis |
| No.10 | Demyelinating syndrome |
| No.11 | Acute confusional state, Psychosis |

*Arthritis Rheum. 1999; 42: 599-608.

Suppl. Table 2. Diagnosis of non-NPSLE

| Non-NPSLE (N = 7) | Diagnosis | OPN measurement | Multiplex analysis |
|---|---|---|---|
| No. 1 | Dizziness due to anemia | ◯ | ◯ |
| No. 2 | Mononeuropathy due to cubital tunnel syndrome | ◯ | ◯ |
| No. 3 | Depression | ◯ | ◯ |
| No. 4 | Temporal lobe epilepsy | ◯ | ◯ |
| No. 5 | Depression | ◯ | |
| No. 6 | Neurosis | ◯ | ◯ |
| No. 7 | Steroid myopathy | ◯ | ◯ |

Suppl. Table 3. Profiles of patients whose OPN, cytokine, and chemokine levels in the CSF were examined

| | Non-NPSLE [1]<br>(N = 6) | NPSLE<br>(N = 8) |
|---|---|---|
| Age | 47.2 ± 19.2 | 35.0 ± 15.0 |
| Sex ratio<br>(male : female) | 0:6 | 1:7 |
| Anti-dsDNA Ab (IU/L)<br>(Min-max) | 8.5 ± 14.1<br>(0 - 37) | 62.5 ± 104.5<br>(0 - 300) |
| SLEDAI<br>(Min-max) | 2.3 ± 3.9<br>(0 - 10) | 17.9 ± 6.0<br>(10 - 26) |
| PSL (mg/day) | 8.8 ± 6.6 | 8.7 ± 4.7 |
| Immunosuppressants | TAC:1 | AZA:1<br>MZB:1<br>CyA + MZB:1 |

[1]SLE patients without NPSLE. Values are expressed as means ± SD and ranges (minimum to maximum). Ab: antibody, AZA: azathioprine, CyA: ciclosporin A, MZB: mizoribine, NPSLE: neuropsychiatric SLE, PSL: prednisolone, SLE: systemic lupus erythematosus, SLEDAI: SLE disease activity index, TAC: tacrolimus.