Supplemental Figure 1. Immune dysregulation in RIPK1 deficiency. tSNE of Phenograph analysis of peripheral blood mononuclear cells (PBMCs) stimulated with either LPS or PMA/I as a (A) conglomerate of all samples with individual cellular population shown or as (B) density plots of the individual groups. (C) Heatmap of the markers expressed in each cluster.
| Metal | Antibody            |
|-------|---------------------|
| 89    | CD45                |
| 115   | CD11c               |
| 141   | CD66b               |
| 142   | CD19                |
| 143   | HLA-DR              |
| 144   | CD64                |
| 145   | CD16                |
| 146   | CD8α                |
| 147   | CD45RO              |
| 148   | CD103               |
| 149   | CD25                |
| 150   | CD38                |
| 151   | CD123               |
| 152   | CD14                |
| 153   | CD45RA              |
| 154   | CD163               |
| 155   | CD27                |
| 156   | CCR4                |
| 158   | CD3                 |
| 159   | CCR7                |
| 161   | CD203c              |
| 162   | CD56                |
| 163   | CD183 (CXCR)        |
| 164   | CD15                |
| 166   | CD24                |
| 167   | LOX-1               |
| 168   | CD206               |
| 169   | CD11b               |
| 171   | CD127               |
| 172   | IgM                 |
| 173   | CD335               |
| 174   | CD4                 |
| 175   | IgD                 |
| 209   | CXCR5               |

**Supplementary Table 1.** Surface antibody panel used for general phenotyping CyTOF analysis.
| Metal | Antibody   |
|-------|------------|
| 89    | CD45       |
| 115   | CD11c      |
| 141   | CD66b      |
| 142   | CD19       |
| 143   | HLA-DR     |
| 144   | TNFα       |
| 145   | CD16       |
| 146   | CD8α       |
| 147   | CD45RO     |
| 148   | CD103      |
| 149   | CD25       |
| 150   | IL-22      |
| 151   | CD123      |
| 152   | CD14       |
| 153   | CD45RA     |
| 154   | CD163      |
| 155   | CD27       |
| 156   | IL-6       |
| 158   | CD3        |
| 159   | CCR7       |
| 160   | IFNγ       |
| 161   | IL-23p19   |
| 162   | IL-1β      |
| 163   | CD183 (CXCR) |
| 164   | CD15       |
| 166   | CD24       |
| 167   | LOX-1      |
| 168   | CD206      |
| 169   | CD11b      |
| 170   | IL-8       |
| 171   | CD127      |
| 172   | PDL1       |
| 173   | CD335      |
| 174   | CD4        |
| 175   | PD1        |
| 176   | IL-10      |

Supplementary Table 2. Antibody panel used for stimulated CyTOF analysis.
| Blood test | Patient 1 | Patient 2 | Normal range        |
|------------|-----------|-----------|---------------------|
| IgG        | 669       | 951       | 700-1600 mg/dL      |
| IgA        | 270       | 77        | 70-500 mg/dL        |
| IgM        | 105       | 46        | 26-100 mg/dL        |
| IgE        | 69        |           | 0-90 mg/dL          |
| TREC       | 2,287     | 2,179     | >400 copies/0.5 mcg DNA |
| WBC (cells/mm³) | 21,000   | 6,790     |                     |
| Lymphocytes (cells/mm³) | 8,127 (38.7%) | 3,530 (52.0%) |                     |
| CD3 (cells/mm³) | 6,339 (78.0%) | 2,471 (70.0%) |                     |
| CD4 (cells/mm³) | 2,601 (32.0%) | 741 (21.0%) | 436-1394 cells/mm³ |
| CD8 (cells/mm³) | 3,576 (44.0%) | 1,377 (39.0%) | 166-882 cells/mm³ |
| CD20 (cells/mm³) | 650 (8.0%) | 812 (23.0%) | 50-300 cells/mm³   |

**Supplementary Table 3: Laboratory work-up of the patients.**