Supplementary table 1. Number of the IgG4-RD and MCD patients with satisfying comprehensive diagnostic criteria for IgG4-RD.

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| --- | --- | --- |
| **Cases with pathological diagnosis** | IgG4-RD (n =45) | MCD (n =33) |
| Definite diagnosis | 41 | 1 |
| Probable diagnosis | 4 | 0 |
| Possible diagnosis | 0 | 5 |
| **Cases with clinical diagnosis** | IgG4-RD (n =10) | MCD (n =10) |
| Definite diagnosis | 0 | 0 |
| Probable diagnosis | 0 | 0 |
| Possible diagnosis | 10 | 1 |

Supplementary table 2. Patient characteristics of IgG4-RD and MCD with clinical diagnosis.

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| --- | --- | --- |
| **Clinical characteristics** | **IgG4-RD (n = 10)** | **MCD (n = 10)** |
| Age (years) | 61.8 ± 14.7 | 55.4 ± 9.6 |
| Male : female ratio | 5:5 | 7:3 |
| Atopic history\* (%) (n) | 60.0 (6) | 10.0 (1) |
| Biopsy sites | None  | 6 skin, 3 bone marrow, 1 lung |
| **Organ involvement (%) (n)** |  |  |
| Orbits and lacrimal glands | 50.0 (5) | 0 (0) |
| Salivary glands | 70.0 (7) | 0 (0) |
| Lymph nodes | 30.0 (3) | 100 (10) |
| Lungs | 30.0 (3) | 70.0 (7) |
| Aorta and large blood vessels | 10.0 (1) | 10.0 (1) |
| Retroperitoneum | 10.0 (1) | 10.0 (1) |
| Pancreas | 40.0 (4) | 0 (0) |
| Kidney | 30.0 (3) | 30.0 (3) |
| Skin | 0 (0) | 60.0 (6) |
| Splenomegaly | 0 (0) | 30.0 (3) |
| **Laboratory data** |  |  |
| WBC (cells/μL) | 6410.0 ± 1581.5 | 6980.0 ± 2100.7 |
| Hb (g/dL) | 13.3 ± 1.2 | 11.0 ± 2.6 |
| Plt (×104/μL) | 20.7 ± 6.5 | 28.3 ± 6.2 |
| Eosinophil (%) | 5.6 ± 0.8 | 2.5 ± 0.4 |
| Alb (g/dL) | 3.8 ± 0.4 | 2.8 ± 0.7 |
| IgG (mg/dL) | 2028.4 ± 615.5 | 4196.3 ± 1532.3 |
| IgA (mg/dL) | 157.6 ± 113.3 | 689.4 ± 252.8 |
| IgM (mg/dL) | 46.6 ± 22.6 | 261.8 ± 127.3 |
| IgG4 (mg/dL) | 795.2 ± 500.8 | 248.0 ± 173.9 (n = 2) |
| CRP (mg/dL) | 0.5 ± 0.9 | 4.4 ± 3.7 |