Supplementary Table S1. Demographic features, clinical characteristics and laboratory parameters of patients with IgG4-RD.

Characteristics at baseline	IgG4-RD (n=51)	
Demographic features		
Age (years), mean ± SD	53 ± 13	
Male/Female ratio	27/24	
Disease duration (month), M (Q$_1$-Q$_3$)	12 (6-30)	
History of allergy (n, %)	22 (43.1)	
Symptoms at disease onset (n, %)		
Lacrimal gland enlargement	16 (31.4)	
Submandibular gland enlargement	11 (21.6)	
Abdominal pain	9 (17.6)	
Nasal congestion	6 (11.8)	
Jaundice	6 (11.8)	
Cough	4 (7.8)	
Lymph node swelling	2 (3.9)	
Back pain	2 (3.9)	
Parotid gland enlargement	2 (3.9)	
Itching	2 (3.9)	
Nausea and vomiting	1 (2.0)	
Arthralgia	1 (2.0)	
Organ affected (n, %)		
Submandibular gland	24 (47.1)	
Lacrimal gland	21 (41.2)	
Pancreas	17 (33.3)	
Lymph node	15 (29.4)	
Paranasal sinus	9 (17.6)	
Lung	9 (17.6)	
Parotid gland	7 (13.7)	
Prostate	5 (18.5)	
Bile duct	6 (11.8)	
Aortitis/periaortitis	3 (5.9)	
Kidney	2 (3.9)	
Pituitary	2 (3.9)	
Thyroid gland	1 (2.0)	
Skin	1 (2.0)	
Liver	1 (2.0)	
Laboratory parameters	**Detected values**	**Normal ranges**
HGB (g/L), mean ± SD	137 ± 18	(110-150)
WBC (10^9/L), mean ± SD	7.3 ± 2.2	(3.50-9.50)
PLT (10^9/L), mean ± SD	270 ± 92	(100-350)
EOS (%), M (Q1-Q3)	3.3 (2.1-7.1)	(0.50-5.00)
Eosinophilia (n, %)	14 (27.5%)	<0.5×10^9/L
ESR (mm/h), M (Q$_1$-Q$_3$)	23 (14-63)	(0-20.0)
hsCRP(mg/L), M (Q$_1$-Q$_3$)	2 (0.99-6.21)	(0-8.00)
IgG (g/L), mean ± SD	20.75 ± 9.36	(7.00-17.0)
IgA (g/L), mean ± SD	2.11 ± 1.06	(0.70-4.00)
IgM(g/L), mean ± SD	1.10 ± 1.03	(0.40-2.30)
IgG1 (mg/L), M (Q$_1$-Q$_3$)	8780 (7728-10125)	(3941-10444)
IgG2 (mg/L), M (Q$_1$-Q$_3$)	5500 (4430-6853)	(1661-8064)
IgG3 (mg/L), M (Q$_1$-Q$_3$)	434 (285-845)	(101-895)
IgG4 (mg/L), M (Q$_1$-Q$_3$)	8155 (3473-15025)	(36-2090)
T-IgE (KU/L), M (Q1-Q3)	344.0 (154.0-596.0)	(0-60.0)
ALT (U/L), mean ± SD	28±25	(7.00-40.0)
Cr (μmmol/L), mean ± SD	72±15	(45.0-84.0)
Disease activity, M(Q1-Q3)		
Baseline IgG4-RD RI	6 (3-9)	
IgG4-RD RI in disease remission	0 (0-1)	

M: median; Q1: quartile 1; Q3: quartile 3; WBC: white blood cell; HgB: haemoglobin; PLT: platelet; EOS: eosinophils; ESR: estimated sedimentation rate; hsCRP: hypersensitive C-reactive protein; Ig: immunoglobulin; C3: complement 3; C4: complement 4; Cr: creatinine; IgG4-RD RI: Immunoglobulin G4-related Disease Responder Index; T-Ig: total immunoglobulin.
Blood eosinophil count higher than 0.5×10^9/L was defined as eosinophilia.
Normal range of serum IgG was 7–17 g/L; normal range of serum IgG4 0–1400 mg/L; normal range of serum IgE was 0–60 KU/L.

Supplementary Table S2. Clinical parameters and treatment strategy of IgG4-RD patients with disease remission.

Number	Age	Gender	RI	Treatment
Patient 1	32	M	1	LEF 20 mg qd
Patient 2	67	M	1	Pred 7.5 mg qd+ MMF 0.5 g bid
Patient 3	68	F	0	Tripterygium 20 mg qd
Patient 4	41	M	0	Pred 5 mg qd+ MTX 15 mg qw
Patient 5	67	M	1	Pred 10 mg qd+ CTX 50 mg qd
Patient 6	46	M	0	Pred 5 mg qd+ MMF 0.75 g qd
Patient 7	45	F	1	LEF 10 mg qd
Patient 8	54	F	0	MP 4 mg qd
Patient 9	36	M	0	Pred 10 mg qd+ CTX 50 mg qd
Patient 10	64	M	1	MP 6 mg qd+ CTX 50 mg qod
Patient 11	42	F	1	MP 4 mg qd+ AZA 50 mg qd
Patient 12	49	F	0	Iguratimod 25 mg qd
Patient 13	67	M	0	Pred 5 mg qd
Patient 14	66	M	0	Pred 10 mg qd
Patient 15	33	M	0	Pred 7.5 mg qd
Patient 16	63	M	0	MP 6 mg qd+ CTX 50 mg qd
Patient 17	53	F	0	MMF 0.25 g qd
Patient 18	37	F	1	LEF 20 mg qd

M: male; F: female; Pred: prednisone; LEF: leflunomide; MMF: mycophenolate mofetil; MTX: methotrexate; CTX: cyclophosphamide; AZA: Aazathioprine; MP: methylprednisolone.
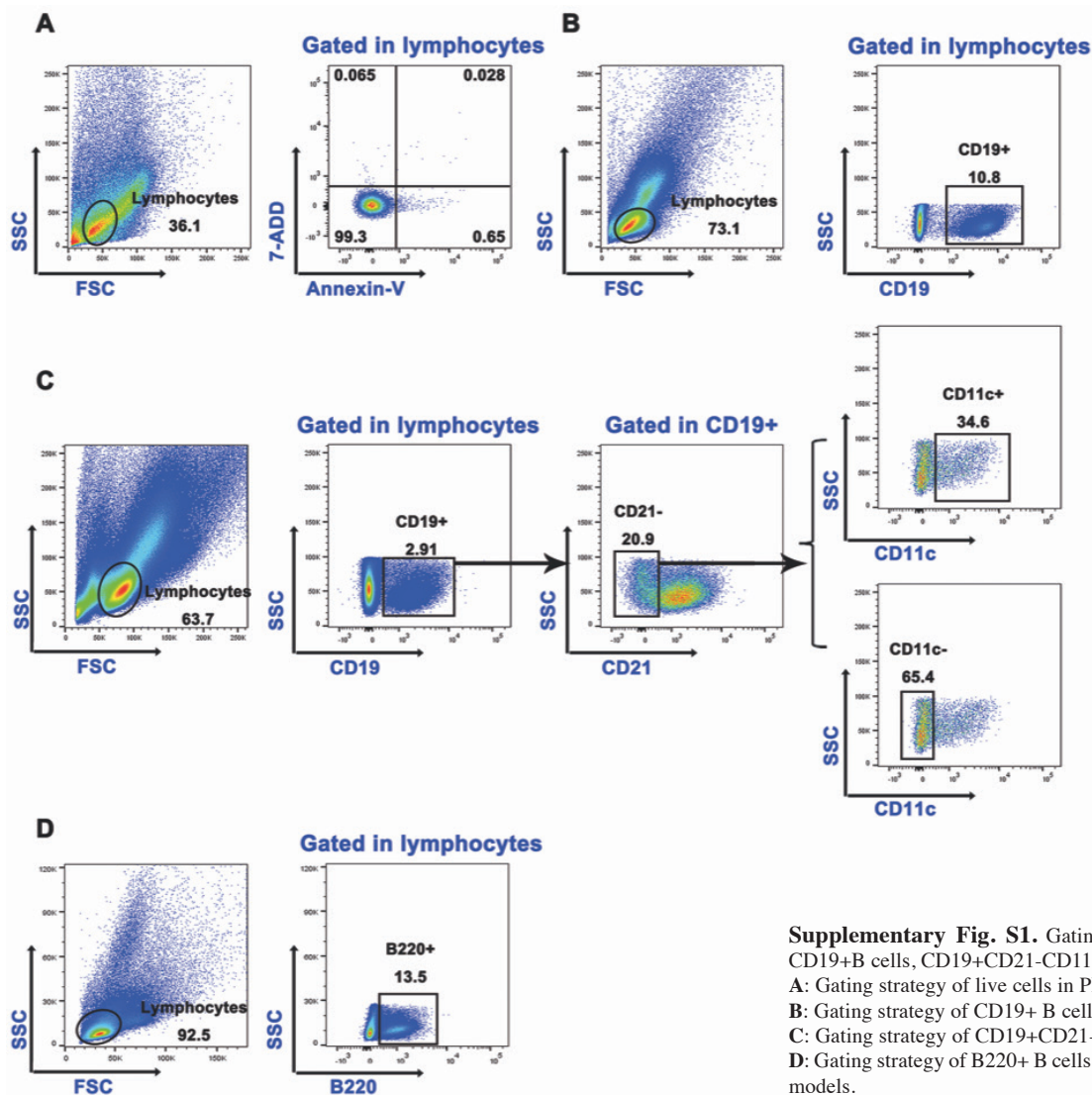

Supplementary Fig. S1. Gating strategy of lymphocytes, CD19+B cells, CD19+CD21-CD11c+/- cells and B220+ cells.
A: Gating strategy of live cells in PBMCs.
B: Gating strategy of CD19+ B cells from lymphocytes.
C: Gating strategy of CD19+CD21-CD11c+/- cells.
D: Gating strategy of B220+ B cells from lymphocytes in mouse models.

A

B

C

D

E
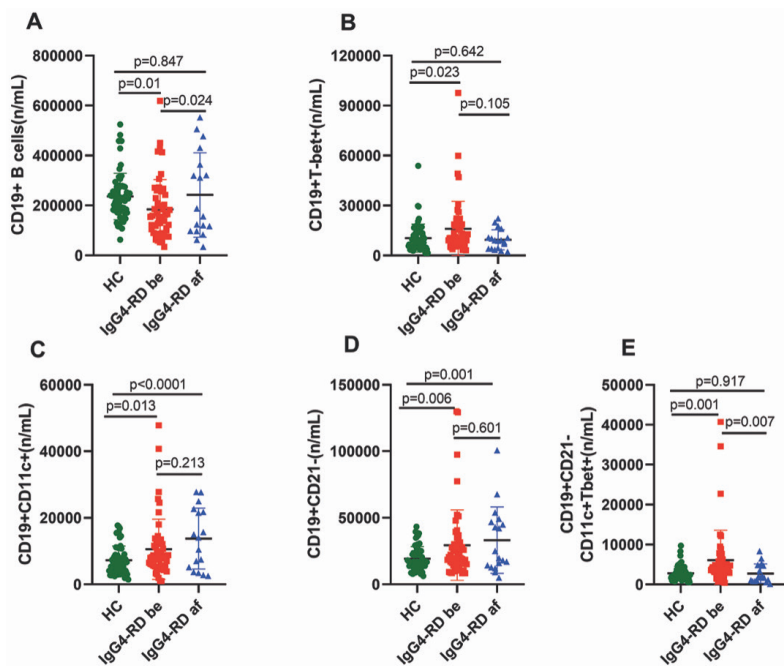
Supplementary Fig. S2. Absolute numbers of CD19+ B cells, CD19+ T-bet+ cells, CD19+CD11c+ cells, CD19+CD21- cells and ABCs in IgG4-RD.
A: Absolute numbers of CD19+ B cells in HC, IgG4-RD before and after treatment.
B-E: Represents the absolute numbers of CD19+T-bet+, CD19+CD11c+, CD19+CD21- and CD19+CD21-CD11c+T-bet+ cells in HC, IgG4-RD before and after treatment.

A

B

C

D

E

F

G

H
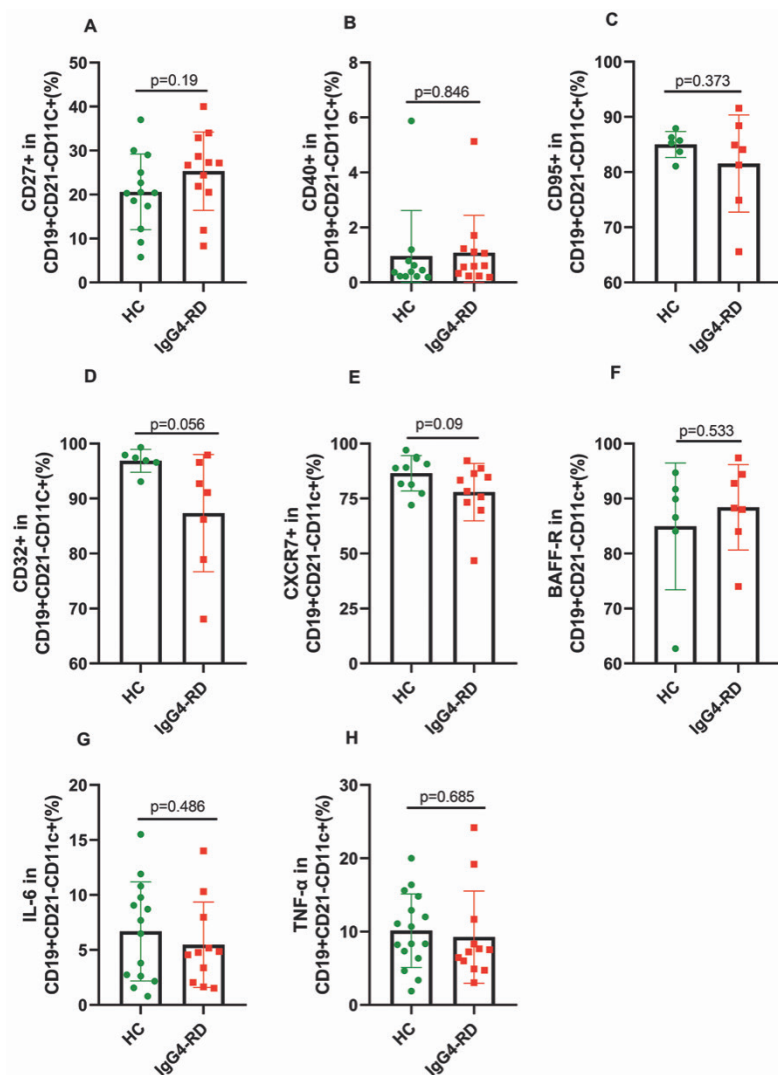
Supplementary Fig. S3. Correlation of ABCs with serum IgG4 levels.

Supplementary Fig. S4. Surface markers and intracellular cytokines staining in CD19+CD21-CD11c+ cells.
A-F: Represents CD27, CD40, CD95, CD32, CXCR7 and BAFF-R expressed in CD19+CD21-CD11c+ cells respectively.
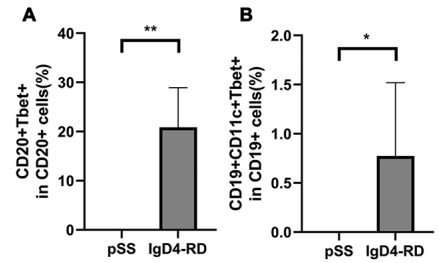**G-H**: Represents intracellular staining of IL-6 and TNF-α in CD19+CD21-CD11c+ cells in IgG4-RD patients and HCs.
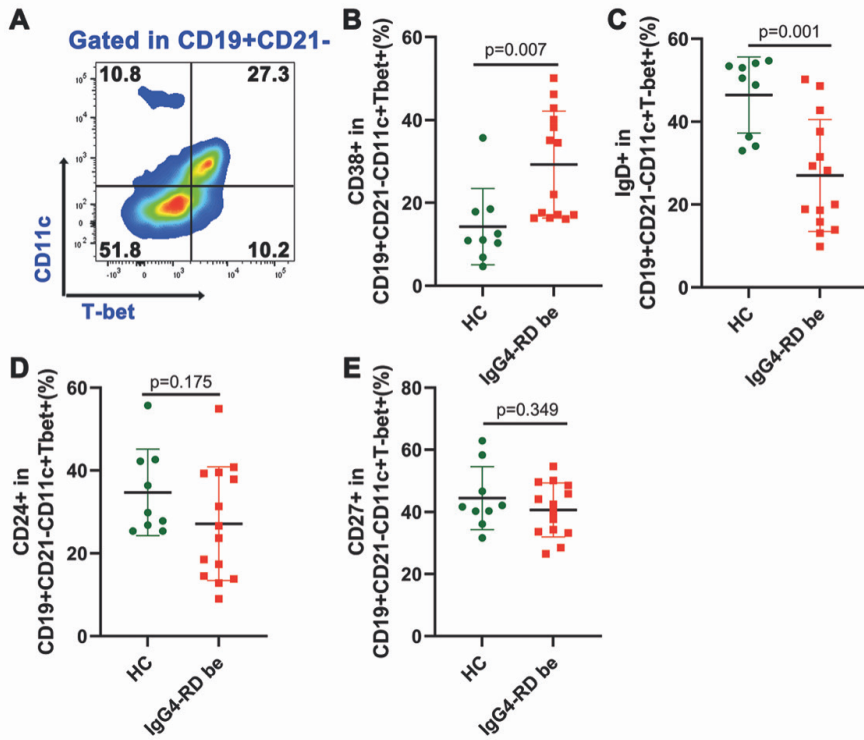
Supplementary Fig. S5. Expression of CD38, IgD, CD24 and CD27 in ABCs of IgG4-RD.
**A**: Representative pictures of T-bet+ co-expression with CD11c+ cells in CD19+CD21- cells of one IgG4-RD patient.
B-E: Represents CD38, IgD, CD24 and CaD27 expression in ABCs of IgG4-RD and HCs.

Supplementary Fig. S7 semi-quantification of CD20+Tbet+ cells and CD19+CD11c+Tbet+ cells in involved organs of IgG4-RD.
A: The semi-quantification of CD20+Tbet+ cells in affected organs of IgG4-RD.
B: The semi-quantification of CD19+ CD11c+ Tbet+ cells in involved organs of IgG4-RD.
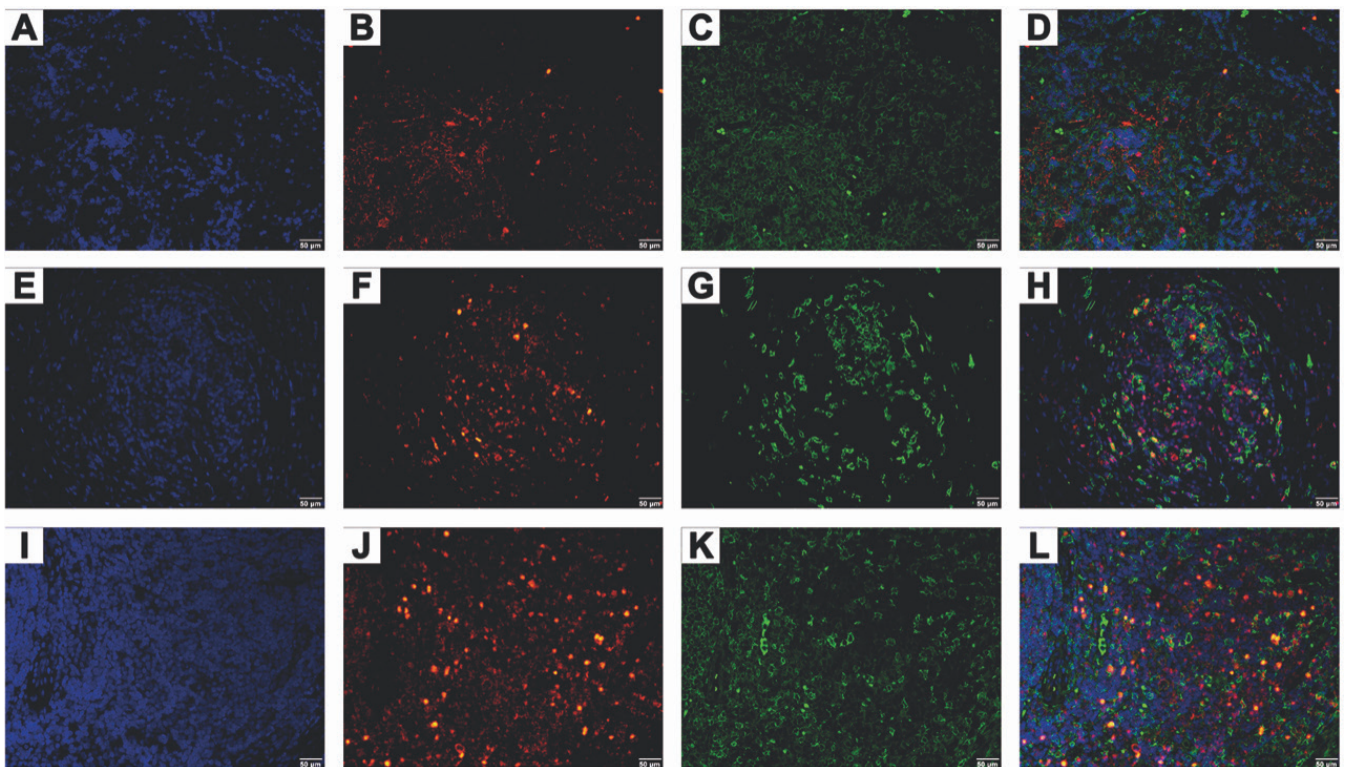
Supplementary Fig. S6. Infiltration of CD20+Tbet+ cells in SMGs of IgG4-RD and controls.
A - D: shows the stain of DAPI, T-bet, CD20 and merge of the three markers in one patient with sialadenitis. **E-H**: shows the stain of DAPI, T-bet, CD20 and the merging of the three markers in patient 1 with IgG4-RD. **I-L**: shows the stain of DAPI, T-bet, CD20 and merge of the three markers in patient 2 with IgG4-RD.