Supplemental Table S1. Analysis of thoracic CT scans of IgG4-RD patients with and without notified thoracic involvement

Thoracic CT scans features	IgG4-RD Group 1 (n=28)	IgG4-RD Group 2 (n=20)	p value (chi2)	All patients (n=48)
Bronchiolitis (tree in bud)	3 (11%)	3 (15%)	0.62	6 (13%)
Bronchiolitis (alveolar nodules)	3 (11%)	4 (20%)	0.41	7 (15%)
Bronchiectasis	9 (32%)	5 (25%)	0.66	14 (19%)
GGO	6 (21%)	0 (0%)	**0.03**	6 (13%)
Area of consolidations	9 (32%)	3 (15%)	0.23	12 (25%)
Pleural effusion	4 (14%)	0 (0%)	0.07	4 (8%)
Pleural thickening	2 (7%)	0 (0%)	0.66	2 (6%)
Peribronchovascular thickening	16 (57%)	10 (50%)	0.61	26 (54%)
Solid nodules	12 (43%)	4 (20%)	0.09	16 (33%)
Mucoid impactions	6 (21%)	2 (10%)	0.32	8 (16%)
Mediastinal lymph nodes	15 (54%)	11 (55%)	0.84	26 (54%)
Septal thickening	5 (18%)	2 (10%)	0.42	7 (15%)
Honeycombing	0 (0%)	0 (0%)	NA	0 (0%)
Reticulations	7 (25%)	0 (0%)	**0.01**	7 (15%)
Architectural distortion	4 (14%)	0 (0%)	0.08	4 (8%)
Retromediastinal fibrosis	2 (7%)	0 (0%)	0.22	2 (4%)
Patterns of IgG4-RD thoracic involvement				
Peribronchovascular involvement	16 (57%)	11 (55%)	0.91	27 (56%)
Lymph nodes enlargement	8 (29%)	7 (35%)	0.62	15 (31%)
Nodular	7 (25%)	0 (0%)	**0.02**	7 (25%)
Interstitial disease	7 (25%)	0 (0%)	**0.02**	7 (25%)
GGO	5 (18%)	0 (0%)	**0.05**	5 (10%)
Pleural disease	4 (14%)	0 (0%)	0.07	4 (8%)
Retromediastinal fibrosis	2 (7%)	0 (0%)	0.22	2 (4%)

GGO: ground-glass opacities

Supplemental Table S2. Characteristics of biopsies performed in the patients with definite thoracic involvement
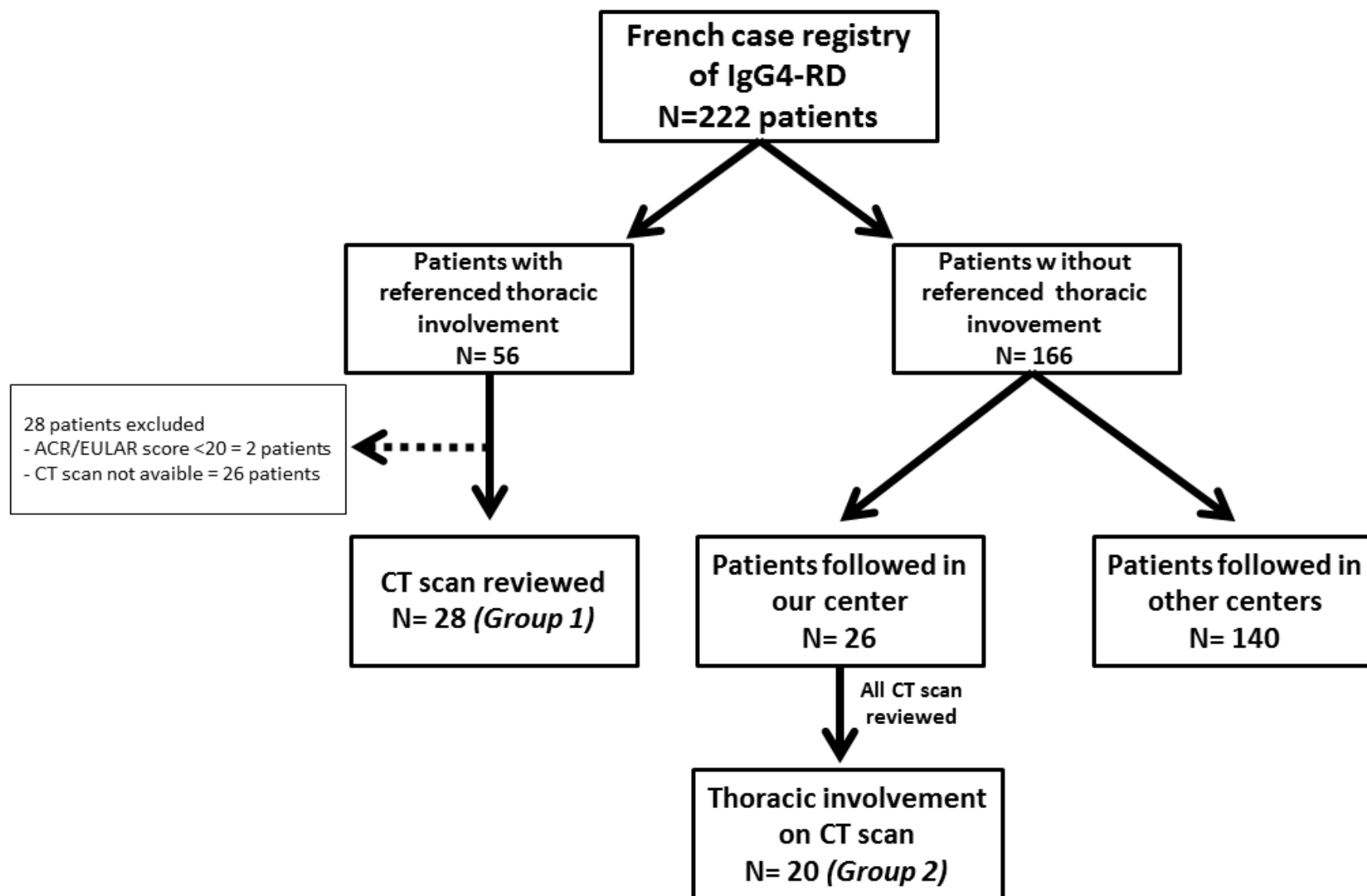
Patients	Biopsied tissue	Biopsy method	Histological feature					
			Storiform fibrosis	lymphoplasmacytic infiltration	Obliterative phlebitis	Absence of granuloma and clonal proliferation	IgG4+ plasma cells / high power field (HPF)	IgG4+/IgG+ ratio
1	Thoracic lymph node	EBUS (endobronchial ultrasound)	×	✓	×	✓	70	70
	Pancreas	Endoscopic retrograde cholangiopancreatography	✓	✓	×	✓	50	40
2	Pulmonary pleurae	Percutaneous core cutting needle	✓	✓	×	✓	60	60
	Abdominal lymph node	Laparoscopy	×	✓	×	✓	30	70
	Salivary glands	Surgical sampling	×	✓	×	✓	25	60
3	Thoracic lymph node	EBUS	×	✓	×	✓	60	80
4	Lung	Surgical sampling	✓	✓	×	✓	30	50
	Thoracic lymph node		×	✓	×	✓	40	45
5	Lung	Surgical sampling	×	✓	×	✓	100	41
	Thoracic lymph node		×	✓	×	✓	50	70
6	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	60	50
	Salivary gland	Surgical sampling	✓	✓	✓	✓	40	80
7	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	80	45
8	Thoracic lymph node	EBUS	×	✓	×	✓	50	48
	Pancreas	Endoscopic retrograde cholangiopancreatography	✓	✓	×	✓	30	40
9	Lung	EBUS	✓	✓	×	✓	80	60
10	Lung	Surgical biopsy	✓	✓	✓	✓	60	80
	Salivary gland	Surgical sampling	×	✓	×	✓	30	30
11	Lung	EBUS	✓	✓	×	✓	70	90
12	Pulmonary pleurae	Percutaneous core cutting needle	✓	✓	×	✓	56	45
	Pancreas	Endoscopic retrograde cholangiopancreatography	×	✓	×	✓	70	80
13	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	70	90
	Salivary gland	Surgical sampling	✓	✓	×	✓	60	50

Patients	Biopsied tissue	Biopsy method	Histological feature					
			Storiform fibrosis	lymphoplasmacytic infiltration	Obliterative phlebitis	Absence of granuloma and clonal proliferation	IgG4+ plasma cells / high power field (H*PF*)	IgG4+/IgG+ ratio
14	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	50	50
	Prostate	Transrectal biopsy	×	✓	✓	✓	40	40
15	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	150	80
	Pancreas	Endoscopic retrograde cholangiopancreatography	✓	✓	×	✓	70	30
16	Thoracic lymph node	EBUS	×	✓	×	✓	50	50
	Liver	Percutaneous needle biopsy	✓	✓	×	✓	60	90
17	Thoracic lymph node	EBUS	×	✓	×	✓	80	80
	Salivary gland	Surgical sampling	✓	✓	×	✓	40	45
18	Pulmonary pleurae	Surgical sampling	✓	✓	✓	✓	60	45
19	Thoracic lymph node	EBUS	×	✓	×	✓	70	70
	Salivary gland	Surgical sampling	✓	✓	✓	✓	55	65
20	Thoracic lymph node	EBUS	×	✓	×	✓	60	80
	Pancreas	Endoscopic retrograde cholangiopancreatography	✓	✓	×	✓	80	80
	Salivary gland	Surgical sampling	×	✓	×	✓	60	70
21	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	40	45
	Prostate	Transrectal biopsy	✓	✓	×	✓	30	40
22	Thoracic lymph node	Mediastinoscopy	×	✓	×	✓	60	50
	Retroperitoneal fibrosis	Surgical sampling	✓	✓	×	✓	70	60
23	Thoracic lymph node	EBUS	×	✓	×	✓	60	70
	Liver	Percutaneous needle biopsy	✓	✓	×	✓	30	40
	Kidney	Percutaneous needle biopsy	×	✓	×	✓	60	40

Supplemental Table S3. Comparison of clinical and biological characteristics of IgG4-RD patients with and without IgG4-related thoracic involvement

	IgG4-RD patients without thoracic involvement (n=146) n (%)	IgG4-RD patients with thoracic involvement (N=48) n (%)	p-value
Demographic data			
Male	106 (72.6)	39 (81)	p = 0.23
Median age at onset (years) [range]	58 [12 ;89]	60 [23;83]	p = 0.86
Median diagnostic delay (months) [range]	18 [1 ;292]	6 [1; 240]	**p < 0.01**
Organ involvement			
Mean number of organs affected [range]	2.5 [1-6]	3 [1-6]	**p = 0.049**
Lymph nodes	74 (50.7)	30 (62.5)	p = 0.14
Autoimmune pancreatitis	51 (34.9)	15 (31.2)	p = 0.64
Tubulointerstitial nephritis	38 (26)	15 (31.2)	p = 0.48
Sialadenitis	45 (30.9)	15 (31.2)	p = 0.84
Retroperitoneal fibrosis	33 (22.6)	10 (20.8)	p = 0.80
Sclerosing cholangitis	38 (26)	10 (20.8)	p = 0.47
Aortitis	19 (13)	8 (16.6)	p = 0.53
Dacryoadenitis	14 (9.6)	6 (12.5)	p = 0.57
Prostatitis	10 (6.8)	3 (6.2)	p = 0.86
Testicular involvement	6 (4.1)	0 (0)	p = 0.15
Hypophysitis	2 (1.3)	0 (0)	p = 0.41
Thyroiditis	5 (3.4)	0 (0)	p = 0.19
Biological characteristics			
Elevated serum IgG (> 1400 mg/dL)	89/128 (69.5)	43 (89.5)	**p < 0.01**
Serum IgG4 level >135 mg/dL	112/136 (82.4)	43 (89.5)	p = 0.24
Serum IgG4 level >2xN	84/136 (61.8)	32 (66.7)	p = 0.55
Median serum IgG4 level (mg/dL) [range]	590 [4; 8970]	430 [23; 2040]	p = 0.61
Low C3 (<80 mg/dL)	22/81 (27.2)	13 (27)	p = 0.90
Low C4 (<15 mg/dL)	21/77 (27.3)	13 (27)	p = 0.91
Low CH50 (<70%)	19/72 (26.4)	13 (27)	p = 0.89
Elevated CRP level (>5 mg/L)	92/125 (73.6)	39 (81.2)	**p = 0.02**
Positive ANA (>1/160)	22/107 (20.5)	9 (18.8)	p = 0.80

IgG: Immunoglobulin G, CRP: C reactive protein, ANA: antinuclear antibodies

Supplemental figure S1. Flow chart showing the search strategy

Supplemental figure S2. Evolution of lung CT scan in a patient with IgG4-RD before and after treatment with prednisone and rituximab

Axial and coronal enhanced thoracic CT scan from a patient with IgG4-related lung involvement with ground glass opacities pattern. A. B. axial slices showing round shape ground glass opacities with centrolobular distribution (arrows) C. coronal reconstruction (arrows). D. E. F. After treatment with prednisone and rituximab, complete regression of ground glass opacities.