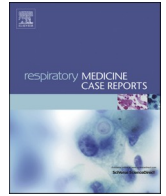




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Long-term spontaneous remission with active surveillance in IgG4-related pleuritis: A case report and literature review

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ABSTRACT

Pleural effusion is a relatively rare feature of IgG4-related disease (IgG4-RD). Here, we report a case of a 72-year-old woman who presented with pleural effusion. Although the pleural adenosine deaminase level was increased, surgical biopsy of the pleura and left inguinal lymph node indicated that the effusion was due to IgG4-RD. Active surveillance was initiated because serum IgG4 and pleural effusion naturally decreased and then completely disappeared. The patient has shown no recurrence for >4 years. This case suggests that pleural biopsy can be used to distinguish IgG4-RD from tuberculosis; moreover, some cases with pleural effusion could improve without treatment.

1. Introduction

Immunoglobulin G4-related disease (IgG4-RD) is a chronic disease that manifests with inflammation and fibrosis of involved tissue. It is characterized by the presence of IgG4-positive plasma cells and lymphocytic infiltration into tissues. IgG4-RD was first reported in 2001 [1], and the Ministry of Health, Labor and Welfare, Japan (MHLW Japan) reported the diagnostic criteria for IgG4-RD in 2011 [2]. However, IgG4-RD shows various clinical features that are not included in these criteria.

Here, we describe a unique case of IgG4-RD accompanied by unilateral pleural effusion that exhibited spontaneous remission. Pleural effusion is a relatively rare manifestation of IgG4-RD; in this case, an elevated level of adenosine deaminase (ADA) was also observed in the pleural effusion. We performed surgical pleural biopsy to distinguish the disease from tuberculous pleurisy, and the pathological analysis

supported a diagnosis of IgG4-RD. The serum IgG4 level and pleural effusion gradually decreased without any treatment, and remission has been maintained for >4 years. Additionally, we have reviewed the literature from 2001 to 2018 in the MEDLINE database based on a search for IgG4-RD with pleural effusion and its clinical presentations.¹

2. Case report

A 72-year-old woman was referred to our hospital for evaluation of left pleural effusion of unknown origin. She had a history of bronchial asthma with noteworthy findings in her family history, including autoimmune diseases and tuberculosis. She reported never smoking or drinking. She had general fatigue for 6 months and had reported left dominant pleural effusion to her previous doctor; however, the cause of pleural effusion was unclear.

On physical examination, her temperature was 37.0 °C, blood

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¹ Abbreviations: Immunoglobulin G4-related disease, IgG4-RD; Ministry of Health, Labor and Welfare, Japan, MHLW Japan; adenosine deaminase, ADA; percutaneous oxygen saturation, SpO₂; computed tomography, CT; fluorodeoxyglucose, FDG; type 2 helper T, Th2; interleukin, IL.

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pressure was 149/92 mmHg, and pulse was 110 beats per minute. Percutaneous arterial oxygen saturation (SpO₂) was 93% on room air. Her right axillary lymph node was palpable. Lung sounds were diminished in the left lung, without the presence of piping rale. Laboratory analysis showed that the erythrocyte sedimentation rate was elevated to 118 mm/h and serum IgG level was 5310 mg/dL, including IgG4 > 1500 mg/dL. Both rheumatoid factor and antinuclear antibody were negative. Analyses of tumor markers and the interferon-gamma release test revealed negative findings (Table 1). Chest radiograph showed left pleural effusion (Fig. 1a). Neck to pelvis computed tomography (CT) revealed swelling of the mediastinal and inguinal lymph nodes as well as left pleural effusion with local pleural thickness (Fig. 1b). Fluorodeoxyglucose (FDG) positron emission tomography imaging showed accumulation of FDG in the mediastinal lymph node, left inguinal lymph

Table 1
Laboratory data on admission.

Peripheral blood			Blood biochemistry		
WBC	2800	/μL	TP	9.8	g/dL
Ne	52.5	%	Alb	2.6	g/dL
Ly	28.0	%	T-Bil	0.45	mg/dL
Mo	7.5	%	AST	23	U/L
Eo	2.0	%	ALT	20	U/L
Ba	5.5	%	ALP	99	U/L
RBC	379	*10 ⁴ /μL	γ-GTP	11	U/L
Hb	11.3	g/dL	ChE	157	IU/L
Ht	33.9	%	LDH	166	U/L
Plt	262	*10 ³ /μL	Na	135	mmol/L
			K	3.7	mmol/L
			Cl	103	mmol/L
			Ca	8.3	mg/dL
Coagulation			BUN	8.9	mg/dL
PT	10.2	sec	Cre	0.41	mg/dL
APTT	30.5	sec	UA	4.5	mg/dL
PT-INR	0.99		T-cho	125	mg/dL
FDP	<2.5	μg/mL	Ferritin	169.3	ng/mL
D-dimer	<0.5	μg/mL	KL-6	140	IU/L
			ft3	2.45	pg/mL
			ft4	1.02	pg/mL
			TSH	2.96	μIU/mL
			ACE	11.5	IU/L
Serology					
CRP	0.09	mg/dL	Tumor marker		
ESR	118	mm/hr	CEA	0.55	ng/mL
RF	10.1	IU/mL	CYFRA	1.7	ng/mL
IgG	5309.7	mg/dL	ProGRP	28	pg/mL
IgG4	>1500	mg/dL			
IgA	89.7	mg/dL			
IgM	22.4	mg/dL			
IgE	2152	IU/mL			
C3	64.3	mg/dL			
C4	20.3	mg/dL			
CH50	37	IU/mL	Urinalysis		
ANA	negative		pH	6.5	
SS-A	negative		Protein	–	
SS-B	negative		Glucose	–	
IL-6	1.04	pg/mL	Occult blood	–	
s-IL2R	1513	IU/mL	Sediment	negative	
BNP	36	pg/mL			
QFT	negative				

Abbreviations: WBC, white blood cells; Ne, neutrophils; Ly, lymphocytes; Mo, monocytes; Eo, eosinophils; Ba, basophils; RBC, red blood cells; Hb, hemoglobin; Ht, hematocrit; Plt, platelets; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; RF, rheumatoid factor; IgG, immunoglobulin G; IgG4, immunoglobulin G4; IgA, immunoglobulin A; IgM, immunoglobulin M; IgE, immunoglobulin E; C3, complement component 3; C4, complement component 4; CH50, total complement activity; ANA, antinuclear antibodies; SS-A, anti-Sjögren's-syndrome-related antigen A; SS-B, anti-Sjögren's-syndrome-related antigen B; IL-6, interleukin-6; s-IL2R, soluble interleukin-2 receptor; BNP, B-type natriuretic peptide; QFT, QuantiFERON; TP, *Treponema pallidum*; Alb, albumin; T-Bil, total bilirubin; AST, aspartate transaminase; ALT, alanine transaminase; ALP, alkaline phosphatase; γ-GTP, gamma-glutamyl transpeptidase; ChE, cholinesterase; LDH, lactate dehydrogenase; Na, sodium; K, potassium; Cl, chloride; Ca, calcium; BUN, blood urea nitrogen; Cre, creatinine; UA, uric acid; T-cho, total cholesterol; KL-6, Krebs von den Lungen-6; ft3, free triiodothyronine; ft4, thyroxine; TSH, thyroid-stimulating hormone; ACE, angiotensin-converting enzyme; CEA, carcinoembryonic antigen; CYFRA, cytokeratin 19 Fragment; ProGRP, pro-gastrin-releasing peptide.

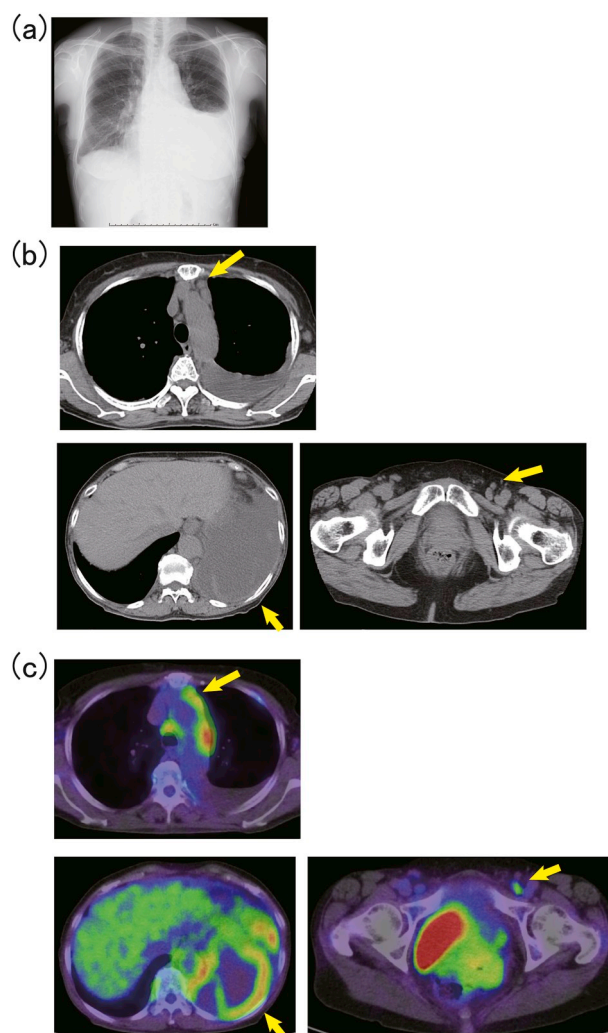


Fig. 1. Chest X-ray (a), computed tomography (CT) (b), and fluorodeoxyglucose positron emission tomography/CT (FDG-PET/CT) imaging (c) on admission. Chest X-ray showed left pleural effusion. Neck to pelvis CT imaging revealed swelling of the mediastinal and inguinal lymph nodes, as well as left pleural effusion with local pleural thickness (yellow arrows). FDG-PET/CT imaging showed FDG accumulation in the mediastinal lymph node, left inguinal lymph node, and left pleura (yellow arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

node, and left pleura (Fig. 1c).

Thoracentesis showed lymphocyte-dominant exudative pleural effusion with a high level of ADA. Culture and polymerase chain reaction analyses of pleural effusion were performed to assess the presence of tuberculosis and nontuberculous mycobacteria. Both tests were negative (Table 2).

We suspected the possibility of IgG4-RD based on clinical findings and the elevated level of serum IgG4. However, we needed to confirm the pathological diagnosis and exclude tuberculous pleurisy with elevated ADA in unilateral pleural effusion. Therefore, we performed surgical biopsies of the pleura and left inguinal lymph node (Fig. 2). Histological analysis of the biopsies revealed dense lymphocyte and plasmacyte infiltration accompanied by fibrosis. Immunohistochemical analysis showed that a large number of IgG4⁺ cells (50 cells or more per high-power field) were found in the pleural membrane and inguinal lymph node; the IgG4⁺/IgG⁺ cell ratio exceeded 40%, which met the diagnostic criteria for IgG4-RD [3]. Thus, we diagnosed the patient with definite IgG4-RD with pleural effusion and inguinal lymph node

Table 2

Thoracentesis findings.

Biochemistry			Cell counts		
TP	6.6	g/dL		5483	/ μ L
Alb	1.6	g/dL	mono	99	%
Glucose	76	mg/dL	poly	1	%
LDH	103	U/L			
ADA	80.2	U/L			
CEA	<0.20	ng/mL			
Microbiology test					
Smear	negative				
Culture	negative				
PCR					
<i>Mycobacterium tuberculosis</i>	negative				
<i>Mycobacterium avium</i>	negative				
<i>Mycobacterium intracellulare</i>	negative				

Abbreviations: TP, total protein; Alb, albumin; LDH, lactate dehydrogenase; ADA, adenosine deaminase; CEA, carcinoembryonic antigen; PCR, polymerase chain reaction.

swelling.

We then considered initiation of corticosteroid therapy; however, pleural effusion decreased without intervention, and dyspnea improved during the active surveillance period (Fig. 3a). The serum IgG/IgG4 levels gradually decreased (Fig. 3b) and the swelling of the mediastinum and inguinal lymph nodes also subsided (Fig. 3c). Therefore, we continued surveillance of the patient without administration of any drug; there has been neither obvious regrowth nor development of malignant disease for >4 years.

3. Discussion

We have herein reported long-term remission with active surveillance of IgG4-RD with pleural effusion. To clarify the characteristics of IgG4-RD with pleural effusion, we conducted a review of the English-language literature by a MEDLINE search using the following keywords: "IgG4-related disease" AND "pleural effusion" OR "pleuritis." Cases with unclear clinical data or unmatched clinical symptoms (e.g., without serum IgG4 or pleural effusion) were excluded. We ultimately

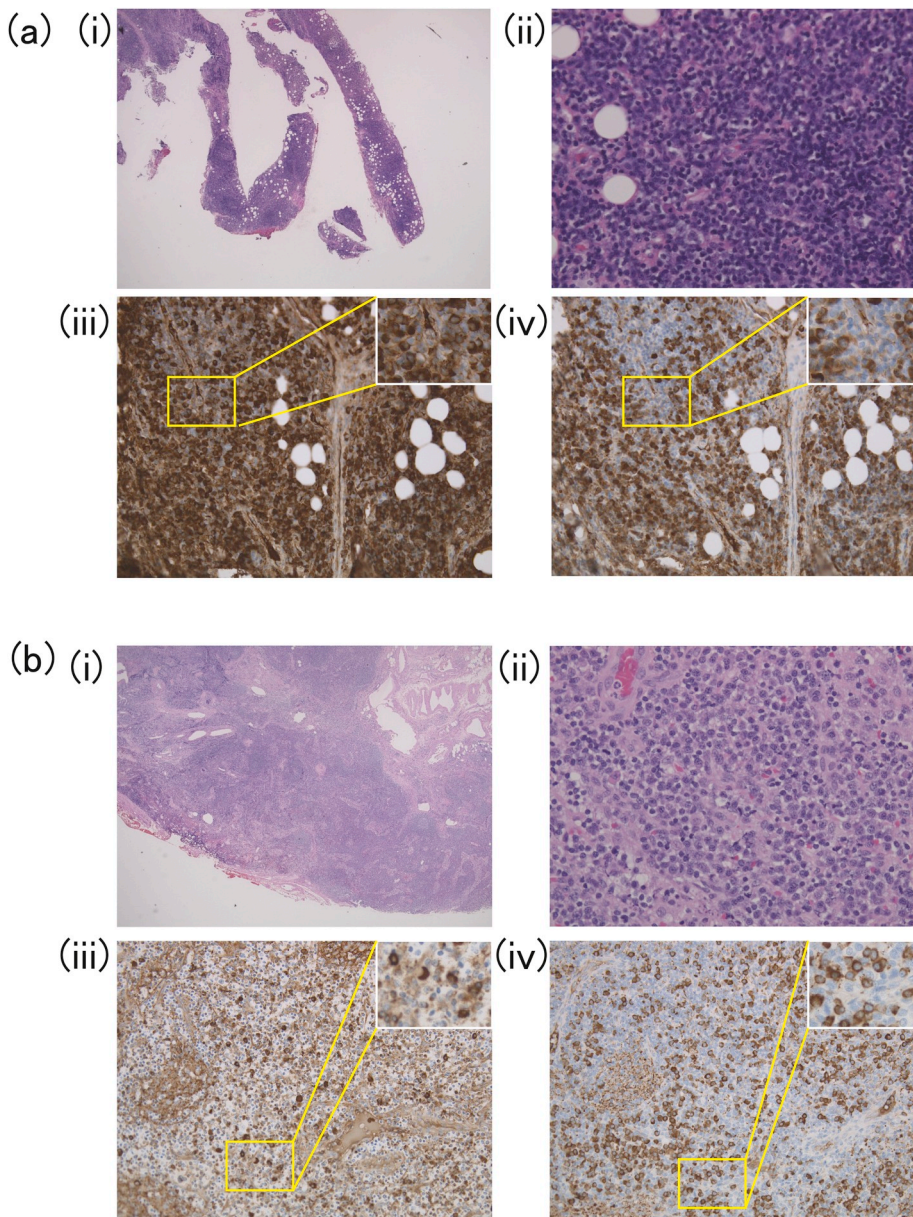


Fig. 2. Histological findings of pleura and inguinal lymph node biopsy specimens. (a): Histological analysis of the pleura revealed dense lymphocyte and plasmacyte infiltration, accompanied by fibrosis [hematoxylin and eosin (H&E) staining, magnification: $\times 10$ (upper left), $\times 400$ (upper right)]. Immunohistochemical analysis showed a large number of IgG4⁺ cells (50 cells or more per high-power field) in the pleura; the IgG4⁺/IgG⁺ cell ratio exceeded 40% [immunohistochemical (IHC) staining, magnification: $\times 200$ (IgG staining; lower left, IgG4 staining; lower right)]. (b): Histological analysis of the inguinal lymph node revealed dense lymphocyte and plasmacyte infiltration, accompanied by fibrosis [H&E staining, magnification: $\times 10$ (upper left), $\times 400$ (upper right)]. Immunohistochemical analysis showed a large number of IgG4⁺ cells (50 cells or more per high-power field) in the inguinal lymph node; the IgG4⁺/IgG⁺ cell ratio exceeded 40% [IHC staining, magnification: $\times 400$ (IgG staining; lower left, IgG4 staining; lower right)].

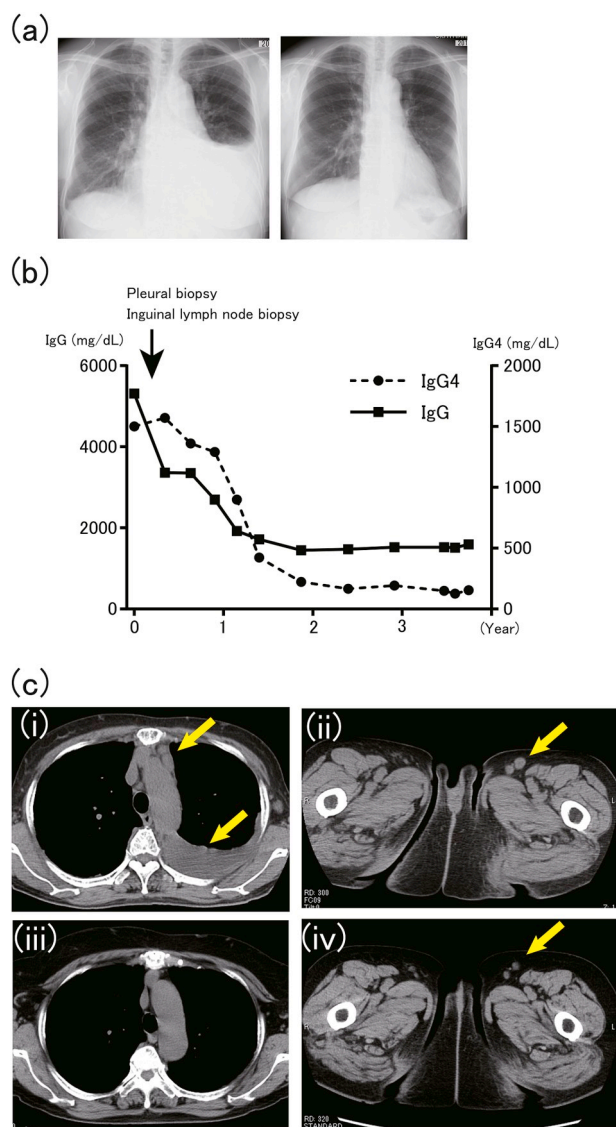


Fig. 3. (a): Clinical course after the patient was referred to our hospital. Serum IgG and IgG4 decreased without treatment. (b): Chest X-ray appearance at the time the patient was referred to our hospital (left), and after 4 years of observation (right). (c): Computed tomography imaging at the time the patient was referred to our hospital (i, ii), and after 4 years of observation (iii, iv). Mediastinal lymph nodes (i, iii) and left inguinal lymph nodes (ii, iv) are shown as yellow arrows.

analyzed 21 cases (including the present case) (Table 3) [4–22].

Review of these 21 cases revealed some recurring rare characteristics. First, the possibility of IgG4-RD should be considered even if pleural effusion is unilateral and the ADA level is elevated. Fei et al. reported that, among a prospective cohort of 248 IgG4-RD cases, 87 (35.1%) had intrathoracic lesions; only 4 cases (4.6%) had pleural effusion [23]. Thus, IgG4-RD manifesting with pleural effusion seems relatively rare. Additionally, most cases of IgG4-RD (15/20 cases) exhibited bilateral pleural effusion; however, some cases, including our case, demonstrate unilateral pleural effusion (Table 3).

As a differential diagnosis, tuberculous pleurisy must be considered, because both diseases could manifest with unilateral pleural effusion accompanied by elevated ADA levels [24–26]. The sensitivity of direct examination of pleural fluid by Ziehl-Neelsen staining or culture was low in previous reports [24]. Krenke et al. reported that ADA activity in pleural fluid is a highly sensitive and specific marker of tuberculous pleurisy (sensitivity: 100%, specificity: 93.9%; cut-off value of 40.4 U/L)

[27]. However, patients with IgG4-RD can sometimes exhibit elevation of ADA levels in pleural effusion [22]. Our review showed that ADA elevation was present in 2/5 cases (at the cut-off value of 40.4 U/L) (Table 3). The underlying mechanism associated with elevated ADA levels in pleural effusion of IgG4-RD is still unknown. In IgG4-RD, type 2 helper T (Th2) cells and regulatory T cells overexpress cytokines, including interleukins (IL-4, 5, 10, and 13), promoting eosinophilia and leading to increased serum IgG4 and IgE levels [28]. On the other hand, ADA is predominantly produced by monocytes or macrophages via the activation of cell-mediated immunity [29]. Therefore, it is suggested that the interaction of T cells and B cells may induce the activation of cell-mediated immunity in IgG4-RD, which could lead to the elevation of ADA levels in pleural effusion [22]. Consequently, it is sometimes difficult to distinguish tuberculous pleurisy and IgG4-RD in patients with unilateral pleural effusion and elevated ADA levels. In some cases, IgG4-RD and *Mycobacterium tuberculosis* infection were concurrently diagnosed [30–33]. In these reports, it is hypothesized that the Th2 activation induced by latent or reactivated tuberculosis results in IgG4-RD. To distinguish tuberculous pleurisy and IgG4-RD accurately, the pathological features and the culture results of the pleural biopsy specimen are important. In this case, the histological feature without granulomatous lesion and the negative results of the *M. tuberculosis* culture and PCR analysis also supported the diagnosis of IgG4-RD.

The second rare characteristic identified in our review was that some patients with IgG4-RD accompanied by pleural effusion showed improvement of symptoms and pleural effusion without treatment. Brito-Zeron et al. revealed that 56 (27.2%) of 206 systemic IgG4-RD patients did not undergo therapy, and only 20 patients showed remission [34]. There are few descriptions of cases manifesting as pleuritis or pleural effusion. In our review of such cases, none improved without corticosteroid therapy, with the exception of our case; some cases were refractory to corticosteroid therapy. Our case is the first report of improvement in IgG4-RD with pleural effusion solely via long-term active surveillance. However, there are currently no clear criteria for treatment decisions (i.e., whether patients should receive immunosuppressive therapy or active surveillance). Therefore, clinical symptoms, radiographs, CT images, and serum IgG/IgG4 levels must be carefully observed to determine the treatment strategy.

The pathogenesis of IgG4-RD remains to be clarified, but an excessive immune response seems to play some role in the progression [35]. A recent study suggested that IgG4-RD may be driven by a specific antigen [36]. If such pathogenesis were applicable in our patient, it may explain the spontaneous remission of IgG4-related pleuritis. Antigen exposure may be temporary, or immune tolerance might be induced for the antigen.

In conclusion, we have described a case of IgG4-RD with unilateral pleural effusion accompanied by elevated ADA levels, which improved without treatment. Unilateral pleural effusion with elevated ADA levels is a relatively rare clinical manifestation, and such cases should be distinguished from tuberculous pleurisy by surgical biopsy of the pleura. In addition, some patients with IgG4-RD can show improvement of symptoms without treatment. Prognostic factors—those that support good response to either corticosteroid therapy or active surveillance—are unknown. Further cases are needed to evaluate such factors in order to provide adequate treatment strategies for patients with IgG4-RD in the future.

Declarations of interest

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Table 3
Literature review of IgG4-RD cases with pleural effusion and their clinical findings.

Year	First Author	Age	Sex	Serum IgG4 (mg/dL)	Associated diseases	Pleural effusion		Corticosteroid therapy	Dose (mg)	Response
						Side	ADA (IU/L)			
2008	Miyake K	65	M	1194	Mikulicz's disease, lymph node swelling	L	NR	PSL	30	Improved
2009	Rossi G	63	F	420	Pericardial effusion, lymph node swelling, Autoimmune pancreatitis, Hashimoto's disease	BL	NR	Steroid	NR	Improved
2011	Yamamoto H	78	M	483	–	BL	34.1–46.7	Surveillance	–	No change
2012	Sekiguchi H	29	F	136	–	BL	NR	PSL	40	Improved
2013	Kojima M	57	M	970	–	BL	NR	Steroid	NR	Improved
2014	Choi JH	48	M	248	–	BL	NR	PSL	0.6 mg/kg	Improved
2014	Ishida A	74	F	740	–	BL	Normal	PSL	25	Improved
2014	Ishida M	71	F	684	Pericardial effusion	R	NR	PSL	40	Improved
2014	Kato E	69	M	277	–	BL	39.9	PSL	30	Improved
2015	Goag EK	16	M	1650	–	BL	10.7–15	PSL + AZA + surgical obliteration	1mg/kg	Improved
2015	Ohkubo H	68	F	307	Uterus	BL	NR	PSL	0.6 mg/kg	Improved
2015	Waheed W	74	M	217	Neuromyopathy	R	NR	PSL	40	Improved
2016	Gonzalez-Moreno J	70	M	437	Pericardial effusion	BL	NR	mPSL→PSL + CyA	PSL; 1 mg/kg	Improved
2016	Gonzalez-Moreno J	70	M	224	Mediastinitis	R	NR	PSL	0.6 mg/kg	Improved
2016	Karim AF	32	M	550	Pericardial effusion	BL	NR	PSL	30	Improved
2016	Kondo T	78	M	760	Pericardium, bile duct	BL	NR	PSL	NR	Improved
2016	Lee HJ	35	M	196	Cardiomyopathy	BL	NR	PSL	125	Improved
2017	Krause ML	84	M	306	S-colon carcinoma (paraneoplastic synd. s/o)	BL	NR	PSL + S-colon tumor resection	40	Improved
2017	Tong X	43	F	125	–	R	4.6–7	PSL	30	Improved
2018	Nagayasu A	81	M	233	–	BL	85	PSL	0.6 mg/kg	Improved
2018	This case	72	F	>1500	Lymph node swelling	L	80.2	Surveillance	-	Improved

L: left, R: right, BL: bilateral, NR: not reported, PSL: prednisolone, AZA: azathioprine, CyA: cyclophosphamide.

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