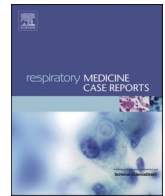


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## Respiratory Medicine Case Reports

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## Case report

# IgG4-related disease with elevated adenosine deaminase in pleural effusion diagnosed clinically using thoracoscopy under local anesthesia and FDG-PET-CT



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## ABSTRACT

In general, we have to assume tuberculous pleurisy when a patient presents with pleural effusion and elevated adenosine deaminase (ADA). However, other diseases need to be considered, including immunoglobulin (Ig)G4-related disease (IgG4-RD). This case involved a 65-year-old asymptomatic man with right pleural effusion showing elevated ADA. He had no articular findings or rashes. Results were negative for all autoantibodies. Pleura, mediastinal lymph nodes, and areas around the aorta and vertebra showed high uptake of <sup>18</sup>F-fluorodeoxyglucose (FDG) on positron-emission tomography-computed tomography (PET-CT). These findings were specific for IgG4-RD. Based on the results of FDG-PET-CT, we performed thoracoscopy under local anesthesia and bronchoscopy. Pleural biopsy and culture, and other examinations including sputum and blood yielded negative findings for tuberculous pleurisy. A pleural biopsy specimen showed IgG4-positive plasma cells and fibrosis without obliterative phlebitis or storiform fibrosis, and serum IgG4 was also high. The ratio of IgG4 to IgG-positive plasma cells was under 40%, and >10 IgG4-positive cells were seen in high-power fields. This case was classed as 'possible IgG4-RD' on the comprehensive diagnostic criteria for IgG4-RD, but did not meet the diagnostic criteria for IgG4-related respiratory disease. Prednisolone proved effective against the pleural effusion. We therefore clinically diagnosed IgG4-RD with pleural effusion based on the 2019 classification criteria for IgG4-RD in the United States. Although few cases of IgG4-RD with pleural effusion have been reported, this disease needs to be considered among the differential diagnoses for high-ADA pleural effusion. FDG-PET-CT and thoracoscopy under local anesthesia may be helpful for diagnosis.

## 1. Introduction

Immunoglobulin (Ig)G4-related disease (IgG4-RD) was first put forward as a disease concept in Japan, and presents as swelling of the systemic organs including the pancreas, lacrimal glands, salivary glands, kidneys and retroperitoneum. This disease is also characterized by elevated serum IgG4, and infiltration and fibrosis of IgG4-positive plasma cells and lymphocytes. The diagnostic criteria for IgG4-related respiratory disease (IgG4-RRD) were proposed from the Japanese Respiratory Society in 2014 [1]. According to these criteria, differentiating

the pathology from other diseases is important, because some patients with elevated serum IgG4 and infiltration of IgG4-positive cells are not classified as showing IgG4-RRD. Thoracic lesions of IgG4-RD have various findings such as mediastinal lymphadenopathy, nodular shadows, bronchial wall thickening, interlobular thickening and consolidation. However, few reports have described IgG4-RD with pleural effusion.

We report herein a rare case of IgG4-RD with pleural effusion. This case showed exudative effusion with high levels of adenosine deaminase (ADA). Distinguishing this case from tuberculous and rheumatic pleurisy

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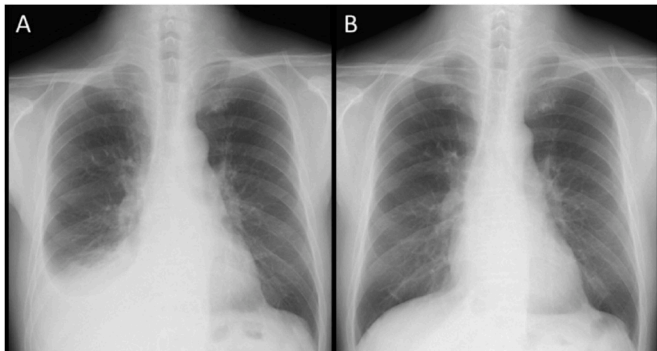
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**Table 1**  
Laboratory findings.

<Complete blood count>			<Biochemistry>		
White blood cells	4950	/ $\mu$ l	Na	141	mEq/L
Neutrophils	60.0	%	K	3.7	mEq/L
Lymphocytes	29.5	%	Cl	107	mEq/L
Monocytes	7.3	%	BNP	115.9	pg/ml
Eosinophils	2.8	%	<Serology>		
Red blood cells	401	$\times 10^4$ / $\mu$ l	C-reactive protein	1.08	mg/dl
Hemoglobin	12.6	g/dl	IgG	2183	mg/dl
Platelets	25.2	$\times 10^4$ / $\mu$ l	IgA	314	mg/dl
<Biochemistry>			IgM	91	mg/dl
Total protein	7.6	g/dl	C3	98	mg/dl
Albumin	3.2	g/dl	C4	104	mg/dl
Total bilirubin	0.4	mg/dl	rheumatoid factor	<3.0	U/ml
Aspartate aminotransferase	24	U/L	antinuclear antibody	80 (Homo)	
Alanine aminotransferase	17	U/L	anti-CCP antibody	0.9	U/ml
Lactate dehydrogenase	176	U/L	IgG4	299	mg/dl
Alkaline phosphatase	202	U/L	CEA	1.0	ng/ml
$\gamma$ GTP	42	U/L	sIL-2R	956	U/ml
Blood urea nitrogen	13.9	mg/dl	KL-6	144	U/ml
Creatinine	1.00	mg/dl	surfactant protein-D	33.5	ng/ml

BNP, brain natriuretic peptide; CCP, cyclic citrullinated peptide; CEA, carcinoembryonic antigen; KL-6, sialylated carbohydrate antigen; sIL-2R, soluble interleukin-2 receptor;  $\gamma$ GTP,  $\gamma$ -Glutamyltranspeptidase.



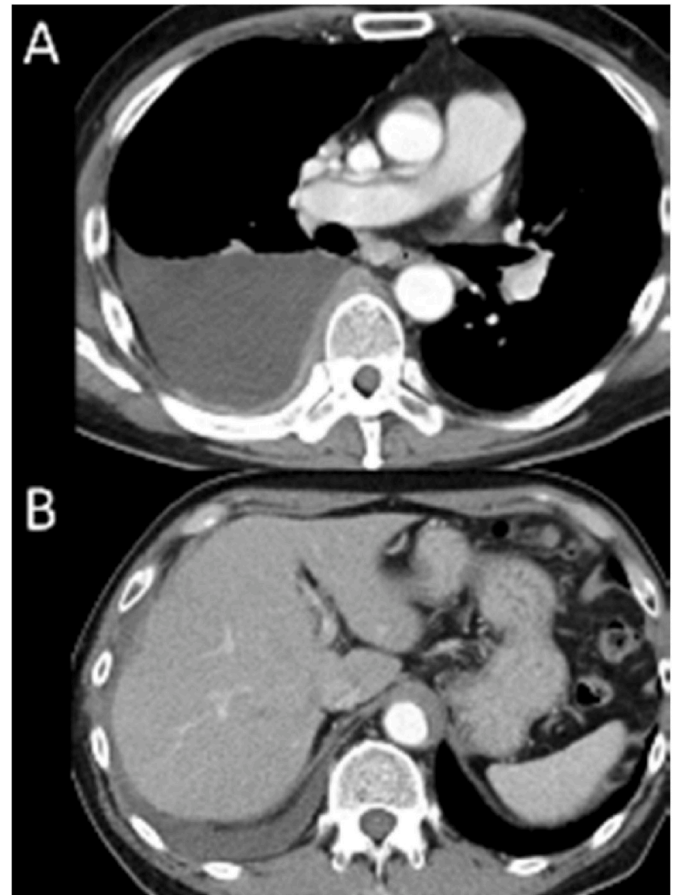
**Fig. 1.** Changes in level of pleural effusion after treatment. Chest X-ray before treatment (A) and after treatment (B).

is thus important. We diagnosed IgG4-RD clinically using thoracoscopic pleural biopsy under local anesthesia and  $^{18}$ F-fluorodeoxyglucose (FDG) positron emission tomography (PET)-computed tomography (CT), and the effectiveness of prednisolone (PSL) treatment. We should include IgG4-RD as a differential diagnosis for patients with exudative pleural effusion and especially high ADA.

## 2. Case report

A 65-year-old Japanese man presented with right pleural effusion during follow-up after percutaneous coronary intervention for angina pectoris. He was asymptomatic. Smoking history was 50 cigarettes/day for 30 years. No allergic history or family history of tuberculosis infection was elicited. The patient was employed in clerical work, with no exposure to asbestos.

On physical examination, temperature was 36.4 °C, blood pressure was 117/65 mmHg, pulse rate was 54 beats/min, respiratory rate was 18 breaths/min, and peripheral oxygen saturation was 98% in ambient air. Lung sounds on auscultation were diminished in the lower right lung field. No articular findings, rash, or swelling of the salivary or lacrimal glands were identified. According to the laboratory data, C-reactive protein (CRP) was 1.08 mg/dl, IgG was 2183 mg/dl, IgG4 was 299 mg/dl, soluble interleukin-2 receptor was 956 U/ml, and negative results were obtained for various autoantibodies, tumor markers, and the interferon-gamma release test (QuantiFERON TB; QIAGEN, Hilden, Germany) (Table 1).



**Fig. 2.** Contrast-enhanced computed tomography (CT) of the chest showed right pleural thickening, pleural effusion, mediastinal lymph node enlargement (A), and periaortitis of the abdomen and perivertebritis (B).

Chest X-ray showed right pleural effusion (Fig. 1A). Contrast-enhanced chest CT showed right pleural thickening, pleural effusion, mediastinal lymph node enlargement, periaortitis of the abdomen and perivertebritis (Fig. 2). FDG-PET-CT showed that maximum standardized uptake value (SUVmax) at these sites was 4–6. SUVmax was 2.17 at



**Fig. 3.** <sup>18</sup>F-fluorodeoxyglucose PET-CT showed that mediastinal lymph node, periaortic, and prevertebral maximum standardized uptake value (SUVmax) was 4–6. SUVmax was 2.17 at the pleura.

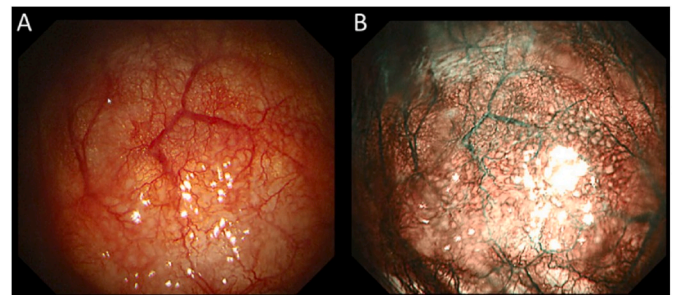
the pleura (Fig. 3). Echocardiography revealed left and right atrial dilation, estimated right ventricular pressure was 36.7 mmHg, although ejection fraction was good, and no valvular disease or synergy was evident. Thoracentesis showed a brownish, lymphocyte-predominant pleural exudate with high levels of ADA, IgG, and IgG4 (Table 2).

Smears, cultures and polymerase chain reaction (PCR) tests for tuberculosis and *Mycobacterium avium* complex all yielded negative results. Cytology was defined as Class II with lymphocytes and plasma cells predominating (Table 2). On thoracoscopy under local anesthesia, thoracoscopic findings revealed no pleural adhesion. Parietal pleura showed white and dense granular lesions (Fig. 4A). Pleural thickness showed mild and white with partly hypervascularization and redness in the cupula and lateral (Fig. 4A). Using narrow-band imaging, distention of capillaries were revealed among dense granular lesions (Fig. 4B). Visceral pleura showed no abnormalities. We obtained these granular lesions in the lateral. These findings were referred to the standardized description of thoracoscopic findings in Japan [2]. Pleural biopsy specimens showed infiltration of inflammatory cells with tiny lymphocytes and fibrosis. The ratio of IgG4-to IgG-positive plasma cells

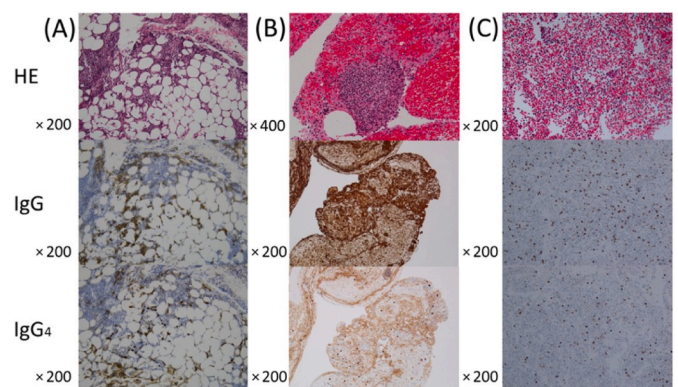
**Table 2**  
Findings from thoracentesis.

White blood cells	4425	/μl	Microbiological test	
Neutrophil	1+		smear	negative
Lymphocyte	2+		culture	negative
Eosinophil	–		PCR	
Total protein	6.2	g/dl	<i>Mycobacterium tuberculosis</i>	negative
Lactate dehydrogenase	112	U/L	<i>M. avium</i>	negative
Glucose	126	mg/dl	<i>M. intracellulare</i>	negative
Amylase	26	U/L	cytology	Class II
Triglycerides	12	mg/dl		
CEA	<0.5	ng/ml		
Adenosine deaminase	46.6	U/L		
Hyaluronate	6000	ng/ml		
IgG	2869	mg/dl		
IgG4	492	mg/dl		

CEA, carcinoembryonic antigen; PCR, polymerase chain reaction.



**Fig. 4.** Thoracoscopic findings; mild pleural thickening with white and dense granular lesions, hypervascularization and redness in the cupula and lateral parietal pleura (A). Distention of capillaries on narrow-band imaging (B).



**Fig. 5.** (A) Pleural biopsy. Infiltration of inflammatory cells with tiny lymphocytes and fibrosis. (B) Mediastinal lymph node biopsy. Infiltration of inflammatory cells with small and large lymphocytes and some plasma cells. (C) Cell block of pleural effusion. Hematoxylin and eosin staining in the top image, immunohistochemical staining for IgG in the middle image, and immunohistochemical staining for IgG4 in the bottom image. The ratio of IgG4-to IgG-positive plasma cells (IgG4/IgG) is 22.4% (A), about 50% (B), and could not be evaluated (C).

(IgG4/IgG) was 22.4%, and 10 IgG4-positive cells were seen on a high-power field (HPF) (Fig. 5A). Transbronchial needle aspiration of a subcarinal lymph node on endobronchial ultrasonic bronchoscopy showed inflammatory cell infiltration with both small and large

**Table 3**  
Clinicopathological features of patients with IgG4-RD pleurisy.

Case	Age years/ Sex	Symptoms	Pleural effusion	Extrathoracic lesions	ADA (U/ L)	Serum IgG4 (mg/dl)	Intrathoracic findings	References
1	81/M	DOE	both	none	85	233		12
2	70/M	DOE, cough	right	none	75.6	1030	diffuse pleural thickening	13
3	43/F	DOE	right	PC, RP		125	partial pleural thickening with multiple nodules and redness	14
4	79/M	None	right	SG	54.6	2040		15
5	78/M	None	right	PC		760	milky pleural plaque	16
6	70/M	DOE	both	PC, PA		437		17
7	70/M	DOE	right	none		224		17
8	32/M	DOE, malaise	both	PC		550		18
9	81/M	DOE	left	SG	61.7	820		19
10	50/F	fever, chest pain, malaise	left	PC		428		20
11	70/M	None	left	PA	56.7	352		21
12	16/M	None	both	none	10.7	1650		22
13	48/M	DOE, fever	both	N				23
14	71/F	DOE, cough	right	PC, PA		684		24
15	69/M	chest pain	right	N	70.6	2380		25
16	29/F	DOE, chest pain	right			136		26
17	78/M	fever, malaise	both	none	46.7	483		27
18	71/M	DOE	left	pituitary		240		28
19	73/M	DOE	right	PC, RP	59.8	1500		29
20	68/M	Cough	left	SG, N	104.4	372		29
21	85/M	Cough	left	orbit, SG, N, stomach, gallbladder	122	2740		30
22	65/M	None	left	SG, PA		279		31
This case	65/M	None	right	PA, RP	46.6	299	mild pleural thickness with white and dense granular lesions, hypervascularization, and redness	

ADA, adenosine deaminase; DOE, dyspnea on exertion; F, female; M, male; N, neck lymph nodes; P, pancreas; PA, periaorta; PC, pericardium; RP, retroperitoneum; SG, salivary glands.



lymphocytes, and some plasma cells. IgG4/IgG could not be evaluated (Fig. 5B). A cell block of pleural effusion suggested that IgG4/IgG was about 50% (Fig. 5C). No obliterative phlebitis or storiform fibrosis was identified. No granulomas or acid-fast bacteria were evident from any biopsy specimens. According to the comprehensive diagnostic criteria for IgG4-RD proposed in 2001, this case would be classed as “possible IgG4-RD” [3]. On the other hand, this case did not meet the diagnostic criteria for definitive IgG4-RRD. Since no findings specific for tuberculosis or rheumatoid arthritis were identified, we clinically diagnosed IgG4-RD with pleural effusion.

After we initiated PSL at 40 mg/day (0.6 mg/kg/day), pleural effusion was improved. The dose of PSL was gradually tapered every 2 weeks. As of the time of writing, we are continuing to administer PSL at 2.5 mg/day with no apparent recurrence (Fig. 1B). This responsiveness to PSL supported the diagnosis of IgG4-RD. Based on the 2019 classification criteria for IgG4-RD in the United States, this case met the inclusion criteria, and did not meet any exclusion criteria [4]. Total score for inclusion was 35 points, over the 20 needed for diagnosis of IgG4-RD [4]. We finally diagnosed IgG4-RD with pleural effusion.

### 3. Discussion

In this case, reaching a definitive pathological diagnosis of IgG4-RD was difficult. However, we reached a diagnosis of IgG4-RD based on thoracoscopic pleural biopsy and PET-CT, and the responsiveness to PSL. This case was characterized by lymphocyte-predominant exudative pleural effusion with high levels of ADA. We have to consider differential diagnoses other than tuberculous pleurisy. Thoracoscopy under local anesthesia may be useful for reaching a diagnosis. PET-CT may also be useful as an adjunctive tool for diagnosis, because some specific findings for IgG4-RD will help decide on biopsy sites.

Sensitivity and specificity of ADA over 50 U/L for tuberculous pleurisy are reportedly 91% and 81%, respectively [5]. However, a previous report found that cases with ADA over 50 U/L were mainly tuberculous pleurisy, although some cases were diagnosed as rheumatic pleurisy, pleurisy associated with systemic lupus erythematosus, bacterial pleurisy, or malignant lymphoma [6]. Thoracoscopy is useful to rule out tuberculous pleurisy and malignancy, and to decide on sample sites according to thoracoscopic findings. Tuberculous pleurisy is difficult to diagnose, but we have previously reported that tissue culture of samples obtained by thoracoscopy might be useful for diagnosis [6]. We performed thoracoscopy under local anesthesia. We can perform this examination safely for patients who cannot be given general anesthesia because of complications as this patient had a history of cardiac problems. Biopsy specimens in this case did not allow us to reach a diagnosis of IgG4-RD, but we were able to rule out tuberculous pleurisy and malignancy from pleural biopsy and culture. Pathological differentiation from multicentric Castleman’s disease was extremely difficult, because this disease showed infiltration of IgG4-positive plasmacytes [7]. However, multicentric Castleman’s disease shows systematic inflammation including fever, high CRP, and intractable reliance on PSL, distinguishing those clinical findings from the present case [7].

Findings from PET-CT for IgG4-RD are characterized by high SUVmax in the involved organs [8]. In addition, the distribution of organs with high SUVmax is helpful for diagnosing IgG4-RD [9]. As seen in this case, a review of 37 cases with PET-CT reported that 41% showed high SUVmax on and around the aorta [10]. No reports have described findings around the vertebrae on PET-CT, but thoracic paravertebral lesions are a specific finding for IgG4-RD on CT [11]. In this case, we could not sample these lesions because of the excessive level of invasiveness. However, we estimate that high SUVmax on mediastinal lymph nodes and the pleura represented the same etiology as periaortitis and perivertebritis. PET-CT may therefore be useful as an adjunctive tool for making a diagnosis and deciding sites for sampling, as we can evaluate findings for all organs and detect lesions which we cannot sample due to invasiveness.

We reviewed 22 cases for the purpose of revealing clinical characteristics of IgG4-RD with pleural effusion (Table 3) [12–31]. Some cases showed symptoms accompanied by pleural effusion including dyspnea on exertion, but few cases had symptoms accompanied by pleurisy including fever and chest pain. Five cases were asymptomatic as in this case. These findings supported IgG4-RD, rather than multicentric Castleman’s disease. Pleural effusion occurred mainly on the right side, as in this case. This might be because thoracic paravertebral lesions occur mainly on the right side [11]. ADA >50 U/L assumed to represent tuberculous pleurisy has been reported in 9 of 11 cases (82%). Some cases showed extrathoracic lesions, but the correlations with serum levels of IgG4 and ADA are unknown. Thoracoscopic findings are also unknown. Only 3 cases were observed, and the findings were various and non-specific. Mild pleural thickness with white and dense granular lesions, hypervascularization, and redness was thoracoscopic findings in this case, which were different from those findings in other cases. However, these findings might have been related to IgG4-RD. As almost no knowledge of these findings has been accumulated, further reports will be required in the future.

In conclusion, IgG4-RD should be included among the differential diagnoses for pleural effusion with elevated ADA, but is extremely difficult to definitely diagnose. PET-CT is helpful both to reach a diagnosis and to choose appropriate biopsy sites. Thoracoscopy under local anesthesia is less invasive than that under general anesthesia and is also helpful for making a diagnosis, and for ruling out tuberculous pleurisy and malignancy.

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### Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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