Low-grade B-cell lymphoma with eosinophilia and elevated IgG4 mimicking IgG4-related retroperitoneal fibrosis: A case report

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Abstract. A 74-year-old man presented with a 4-month history of fatigue, malaise and persistent epigastric discomfort. Peripheral blood testing showed marked eosinophilia (41.5% of leukocytes; absolute eosinophil count, 2,075/μl). The patient was referred to the Department of Hematology (Toranomon Hospital Kajigaya, Kawasaki, Japan), but no hematologic disorder explaining the eosinophilia was identified, and the patient was subsequently referred to the Department of Rheumatology. Laboratory tests revealed elevated serum IgG4 (3,024 mg/dl) and soluble interleukin-2 receptor (2,820 U/ml) levels, whereas C-reactive protein was negative. Non-contrast computed tomography and ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography demonstrated increased attenuation and FDG uptake in the retroperitoneal adipose tissue. IgG4-related retroperitoneal fibrosis was initially considered, although the lesions were atypical for IgG4-related disease (IgG4-RD). Laparoscopic biopsy of a retroperitoneal lymph node was performed. Histopathological findings were not consistent with IgG4-RD. Immunohistochemistry and molecular analyses supported a clonal B-cell population, including monoclonal immunoglobulin heavy-chain gene rearrangement. The lesion was diagnosed as low-grade B-cell lymphoma, with marginal zone lymphoma considered the most likely but not definitively established. Although we initially selected to observe the patient, the serum IgG4 level continued to increase and his gastrointestinal symptoms worsened; therefore, oral prednisolone was initiated at 20 mg/day. His symptoms promptly resolved accompanied by a reduction in the serum IgG4 level. Prednisolone was tapered and discontinued over 6 months without recurrence. This case highlights the importance of sufficient radiological and histopathological evaluation, and appropriate genetic analysis in distinguishing IgG4-RD from its mimickers. We hypothesize that a lymphoma-associated type 2 immune response may underlie eosinophilia and elevated IgG4 levels.

Introduction

IgG4-related disease (IgG4-RD) is a systemic chronic inflammatory disorder characterized by elevated serum IgG4 levels and lymphoplasmacytic infiltration with IgG4-positive plasma cells and fibrosis in affected organs (1,2). The diagnosis of IgG4-RD is based on a combination of clinical, serological, and histopathologic findings, including elevated serum IgG4 levels and characteristic histologic features such as dense infiltration of IgG4-positive plasma cells, storiform fibrosis, and obliterative phlebitis (3). However, numerous disorders can mimic IgG4-RD, and careful exclusion of these conditions is essential. The 2019 ACR/EULAR classification criteria include clinical, serological, radiological, and pathological exclusion criteria (4). These criteria include marked peripheral eosinophilia (>3,000/mm^3) and radiological or pathological findings suspicious for malignancy that have not been sufficiently investigated as exclusion criteria. Nevertheless, eosinophilia and elevated serum IgE levels are relatively common in IgG4-RD, which can sometimes complicate the distinction from its mimickers (5).

Among hematologic mimickers, B-cell lymphomas are particularly important because some may show plasmacytic differentiation and prominent IgG4-positive plasma-cell

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infiltration. IgG4-producing marginal zone lymphoma (MZL) has been reported, and a systematic review has described IgG4-positive MZLs occurring both in association with IgG4-RD and as de novo lymphomas (6,7). Th2/Treg-associated immune responses have also been implicated in IgG4-related conditions and IgG4-positive MZLs (8,9). In addition, persistent eosinophilia requires careful evaluation to distinguish secondary eosinophilia from clonal hematologic disorders (10).

Here, we report a case of low-grade B-cell lymphoma, considered most consistent with marginal zone lymphoma (MZL), that showed marked eosinophilia, elevated serum IgG4 levels, and distinctive radiological findings. This case underscores lymphoma as an important mimicker of IgG4-RD. We also discuss a possible lymphoma-associated type 2 immune response as a hypothesis that could account for the concomitant IgG4 elevation and eosinophilia.

Case report

A 74-year-old man presented with a 4-month history of fatigue and general malaise. Because of persistent epigastric discomfort, he visited a local physician and marked eosinophilia, with eosinophils accounting for 41% of leukocytes, was found. He was referred to our hematology department, and a bone marrow biopsy was performed revealing 27.6% eosinophils, whereas plasma cells accounted for only 1.0%. Fluorescence in situ hybridization showed no deletion involving 4q12. No hematologic disorder accounting for the eosinophilia was identified, and the patient was referred to our department.

The day of admission to the department of Hematology was defined as day 1. The laboratory findings at presentation are summarized in Table I. The white blood cell count was 5,000/μl, with eosinophils accounting for 41.5%, corresponding to an absolute eosinophil count of 2,075/μl. No recent drug exposure, allergic disease, parasitic infection, or other identifiable secondary cause of eosinophilia was found. No monoclonal protein or light-chain restriction was detected. Serum IgG was elevated to 4,509 mg/dl, with markedly elevated IgG4 and IgE levels of 3,024 mg/dl and 797 IU/ml, respectively. The soluble interleukin-2 receptor (sIL-2R) level was mildly elevated at 2,820 U/ml. Autoantibodies, including antineutrophil cytoplasmic antibodies, were negative.

The imaging findings are shown in Fig. 1. Non-contrast computed tomography (CT) on day 4 revealed increased attenuation of the retroperitoneal adipose tissue. (Fig. 1A) The lesion was not limited to the retroperitoneum but extended into anterior mesentery and the pelvis. Areas of slightly increased attenuation were also observed within the adipose tissue, indicating that the lesion extended into the surrounding fat. Mild lymphadenopathy was also present. Magnetic resonance imaging was performed on day 14. Water-only T1-weighted image (Fig. 1B) and Fat-suppressed T2-weighted image (Fig. 1C) also showed that the lesion extended into adipose tissue. Diffusion-weighted imaging (DWI) (Fig. 1D) showed high intensity corresponding to these lesions, with decreased apparent diffusion coefficient (ADC) value on ADC map (Fig. 1E). These findings on DWI and ADC indicated restricted diffusion within the lesion, probably associated with cellular infiltration rather than simple edema. (Fig. 1D and E) ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) also demonstrated proliferation and increased attenuation of the adipose tissue in the posterior mediastinum and retroperitoneum. Increased FDG uptake was also observed in multiple lymph nodes throughout the body. (Fig. 1F and G) IgG4-related retroperitoneal fibrosis was initially considered. However, the distribution and extension of the lesions were atypical for IgG4-RD, raising concern for an underlying neoplasm, including lymphoma.

The department of gastroenterological surgery was consulted to perform a biopsy of the retroperitoneal lesion on day 25. Intraoperatively, the retroperitoneum appeared erythematous, as shown in Fig. 2A. Histopathological examination showed prominent extension of inflammatory or neoplastic cells into the surrounding adipose tissue and extranodal areas. These cells comprised plasma cells and plasmacytoid lymphocytes, including cells with abundant cytoplasm and occasional cells showing early Russell body-like changes. (Fig. 2B and C) These plasma cells were positive for IgG and IgG4, with an IgG4/IgG-positive plasma-cell ratio of over 95%. (Fig. 2D and E) However, the distribution and pattern of plasma cell infiltration and absence of fibrosis were not consistent with IgG4-RD. Immunohistochemical staining showed that the B-cell population was positive for CD20, whereas the plasma cells lacked CD20 expression. (Fig. 2F) The plasma cells were positive for CD19 (Fig. 2G) and CD138. Immunohistochemistry for CD5, CD10, CD56, BCL2, BCL6, cyclin D1, and MUM1 was negative, and EBER-ISH was negative. In situ hybridization for both Immunoglobulin kappa and lambda light chains showed no detectable expression of either light chain. (Fig. 2H and I) Immunoglobulin heavy-chain (IgH) polymerase chain reaction analysis performed using DNA extracted from the whole tissue in an unstained section detected a monoclonal rearrangement in framework region 2 (FR2). The tissue was not specifically enriched for IgG4-positive plasma cells; therefore, this finding supported the presence of a clonal B-cell process but did not establish that the IgG4-positive plasma cells belonged to the same clone. These findings supported a diagnosis of low-grade B-cell lymphoma, with MZL considered the most likely but not definitively established subtype. Based on these findings, the patient was diagnosed with low-grade B-cell lymphoma accompanied by secondary elevations in serum IgG4 and peripheral blood eosinophilia.

The clinical course is shown in Fig. 3. At our institution, the initial pathological assessment on day 71 raised concern for a plasma-cell neoplasm. However, the hematologists considered this diagnosis insufficiently supported by the available immunophenotypic findings, including the absence of detectable kappa and lambda light-chain expression. Because the pathological interpretation remained unresolved, an external hematopathologist was consulted. During this period, the patient was initially followed with no medication; however, serum IgG and IgG4 levels continued to increase, fatigue and general malaise persisted, and diarrhea worsened. No gastrointestinal biopsy was performed; therefore, eosinophilic gastrointestinal involvement could not be completely excluded. After discussion between the hematologists and we rheumatologists, glucocorticoid therapy following the treatment approach for IgG4-RD was

Table I. Laboratory findings at initial presentation.

A, Blood biochemistry

Laboratory values, unit	Patient result	Reference range
Total protein, g/dl	9.1	6.6-8.1
Albumin, g/dl	2.3	4.1-5.1
Aspartate aminotransferase, U/l	23	13-30
Alanine aminotransferase, U/l	19	10-42
Lactate dehydrogenase (IFCC), U/l	166	124-222
Alkaline phosphatase (IFCC), U/l	48	38-113
γ-glutamyl transpeptidase, U/l	10	13-64
Total bilirubin, mg/dl	0.4	0.4-1.5
Urea nitrogen, mg/dl	13	8-20
Creatinine, mg/dl	0.89	0.65-1.07
Estimated glomerular filtration rate, ml/min/1.73 m^2	64.1	>60
Uric acid, mg/dl	6.6	3.7-7.0
Sodium, mmol/l	137	138-145
Potassium, mmol/l	4.0	3.6-4.8
Chloride, mmol/l	108	101-108
Erythrocyte sedimentation rate, mm/h	111	1-10
C reactive protein, mg/dl	0.26	<0.14
Ferritin, ng/ml	170	5-120

B, Immunology

Laboratory values, unit	Patient result	Reference range
Immunoglobulin G, mg/dl	4,509	861-1,747
Immunoglobulin G4, mg/dl	3,024	<118
Immunoglobulin A, mg/dl	82.9	93-393
Immunoglobulin M, mg/dl	35.8	50-269
Immunoglobulin E, IU/ml	797	<170
CH50, IU/l	22	31-58
Complement C3, mg/dl	44	73-138
Complement C4, mg/dl	12	11-31
Rheumatoid factor, IU/ml	<5	0-15
Anti-nuclear antibody	<40	<40
M protein	Negative	
Free light chain κ, mg/l	346.8	3.3-19.4
Free light chain λ, mg/l	282.3	5.7-26.3
κ/λ ratio	1.23	0.26-1.65
Soluble IL-2 receptor, U/ml	2,820.0	157-475

C, Complete blood count

Laboratory values, unit	Patient result	Reference range
White blood cell count, x10^3/μl	5.0	3.3-8.6
Neutrophils, %	30.0	38.0-74.0
Eosinophils, %	41.5	0-8.5
Hemoglobin, g/dl	12.4	13.7-16.8

Table I. Continued.

C, Complete blood count

Laboratory values, unit	Patient result	Reference range
Hematocrit, %	37.5	40.7-50.1
Platelet count, x10^3/μl	181	158-348
Hemoglobin A1c, %	5.5	4.9-6.0

D, Bone marrow biopsy

Laboratory values, unit	Patient result	Reference range
Nucleated cell count, /μl	33,000	
Myeloid-to-erythroid ratio	4.03	
Eosinophils, %	27.6	
Plasma cells, %	1.0	
Lymphocytes, %	5.0	

CH50, 50 percent hemolytic unit of complement; IL; interleukin; IFCC, International Federation of Clinical Chemistry and Laboratory Medicine.

recommended. Prednisolone 20 mg/day (0.4 mg/kg/day) was started on day 84, followed by rapid improvement in the patient's symptoms and decreases in serum IgG and IgG4 levels as well as the IgG4/IgG ratio. The external hematopathology review subsequently supported a diagnosis of low-grade B-cell lymphoma, and the final diagnosis was established on day 140. The prednisolone dose was gradually tapered at approximately monthly intervals and discontinued on day 259. FDG-PET/CT performed on day 284 showed marked improvement, with near-complete resolution of the lesions identified at presentation. The patient has remained free of recurrence through day 465.

Discussion

We encountered a case of low-grade B-cell lymphoma that was identified during the evaluation of peripheral eosinophilia and markedly elevated serum IgG4 levels. The present case did not meet the 2020 revised comprehensive diagnostic criteria for definite IgG4-RD and did not reach the classification threshold of the 2019 ACR/EULAR criteria (3,4). According to the 2020 revised diagnostic criteria, the markedly elevated serum IgG4 level fulfilled the serological criteria; however, the biopsy fulfilled the IgG4-positive plasma-cell subitem but lacked fibrosis, storiform fibrosis, and obliterative phlebitis; therefore, the pathological item was not fulfilled. Thus, even if the retroperitoneal lesion were regarded as characteristic organ involvement, the present case would meet at most the category of possible IgG4-RD (3). Importantly, the absence of storiform fibrosis or obliterative phlebitis alone does not exclude IgG4-RD, particularly in lymph-node lesions. According to the 2019 ACR/EULAR classification criteria, the absolute

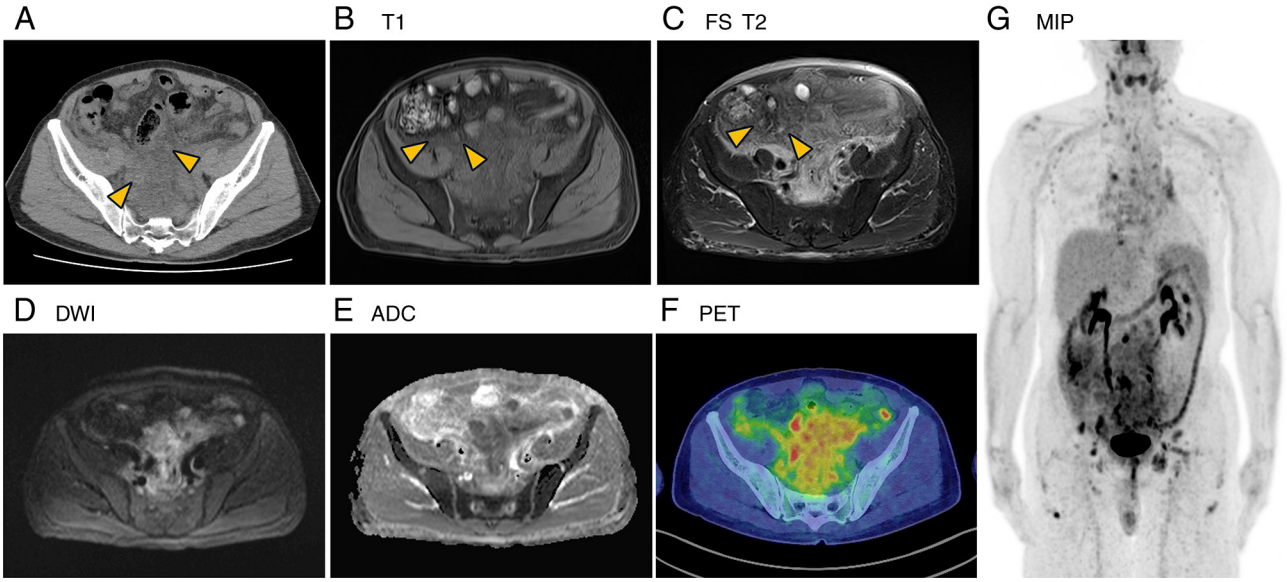


Figure 1. Radiographic imaging at initial presentation. (A) Non-contrast computed tomography showed enlargement and increased attenuation of the adipose tissue in the retroperitoneum and adjacent mesentery. Several lymphadenopathies were also present within the lesion (arrowheads). (B) T1-weighted gradient-echo image showed low intensity within the lesions, and (C) FS single-shot T2-weighted image showed high intensity within the lesions, indicating extension of the lesions into adipose tissue. (D) DWI showed high intensity lesions, and (E) ADC map showed decreased ADC value, indicating restricted diffusion consistent with cellular infiltration rather than simple edema. (F) FDG-PET/CT demonstrated proliferation and increased attenuation of the adipose tissue in the posterior mediastinum and retroperitoneum, with increased FDG uptake in multiple lymph nodes throughout the body. (G) MIP image of FDG-PET/CT. DWI, diffusion-weighted imaging; FS, fat-suppressed; ADC, apparent diffusion coefficient; FDG-PET/CT, ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography; MIP, maximum intensity projection.

eosinophil count was 2,075/μl and was therefore below the >3,000/mm^3 exclusion threshold. However, the retroperitoneal lesion lacked the typical circumferential or anterolateral periaortic distribution used for weighted inclusion, and IgG4 immunostaining of lymph-node tissue is not scored in the immunostaining domain. Serum IgG4 >5 times the upper limit of normal contributes 11 points; even if 4 points were assigned for dense lymphocytic infiltration, the total would remain below the ≥20-point classification threshold (4).

Among B-cell lymphomas, MZL, particularly MZL with plasmacytic differentiation, is the subtype most commonly associated with IgG4 expression or production. Sato *et al* (6) reported a case of MZL involving retroperitoneal lymph nodes and demonstrated that IgG4-producing cells belonged to the neoplastic clone. Nishimura *et al* (7) subsequently conducted a systematic scoping review of IgG4-positive MZLs and reported that IgG4-positive MZLs may be classified into two different types: IgG4-RD-related and de novo IgG4-positive MZLs. Although MZL was considered the most likely in our patient, definitive diagnosis was not possible. Moreover, because the IgH rearrangement analysis was performed on whole tissue in unstained sections, the relationship between the IgG4-positive plasma cells and the neoplastic clone remains unresolved. Therefore, our lymphoma could not be definitively classified as an IgG4-producing MZL. However, taken together with the histopathologic findings of our case and the previously reported association between MZL and IgG4 production, the present low-grade B-cell lymphoma was considered most consistent with MZL.

A key question in this case is whether the elevated IgG4 reflected IgG4 production by the neoplastic cells or a lymphoma-associated IgG4 reaction. One distinctive finding was that neither kappa nor lambda light-chain expression was detectable. Unlike the case reported by Sato *et al* (6), our findings do not establish that the IgG4-positive plasma cells themselves represented the neoplastic population and leave open the possibility of a reactive IgG4 response.

We hypothesize that the concomitant IgG4 elevation and eosinophilia may reflect a lymphoma-associated type 2 immune response that promoted the expansion and IgG4 class switching of plasma cells. In IgG4-RD, expansion of regulatory T cells and activation of Th2-associated immune responses have been implicated in the pathogenesis, with cytokines including IL-4 and IL-5 contributing to IgG4-related immune responses and eosinophilia (8). Ohno *et al* (9) compared ocular adnexal IgG4-positive and IgG4-negative MZLs with ocular IgG4-RD and demonstrated that IgG4-related MZLs exhibited upregulation of cytokines associated with Th2/Treg response. These findings provide a biological rationale for the possibility that a Th2/Treg-associated type 2 immune response may have promoted a lymphoma-associated IgG4 reaction in the present case. However, cytokine measurements, T-cell phenotyping, and tissue expression of IL-4, IL-5, IL-13, GATA3, or FoxP3 were not evaluated in this patient; therefore, this interpretation remains hypothetical.

This hypothesis is also supported by the presence of eosinophilia. In our case, eosinophilia was observed in both the peripheral blood and bone marrow. No apparent eosinophil-mediated organ dysfunction was identified. Because the gastrointestinal symptoms were not evaluated histologically, eosinophilic gastrointestinal involvement could not be completely excluded. Therefore, hypereosinophilic syndrome was not established (10). Eosinophilia could be promoted by type 2 immune activation through the IL-5

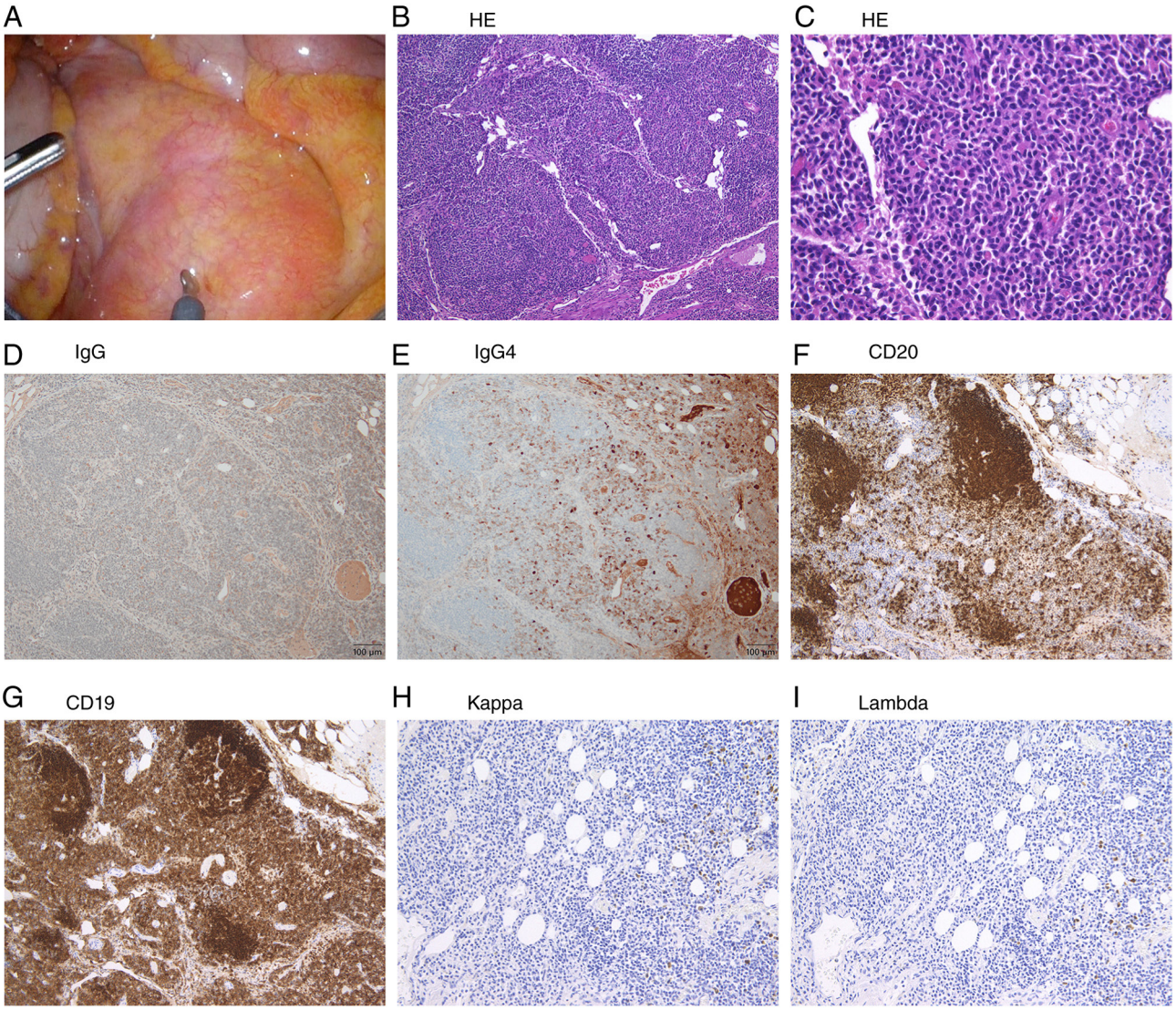


Figure 2. Pathological findings of retroperitoneum biopsy specimen. (A) Macroscopic appearance of the retroperitoneal lesion, which was erythematous. (B) Low power and (C) high power field of hematoxylin and eosin staining of lymph nodes in retroperitoneum. Inflammatory or neoplastic cells extended into the surrounding adipose tissue and extranodal areas. These cells were plasma cells and plasmacytoid lymphocytes, including cells with abundant cytoplasm and occasional cells showing early Russell body-like changes. Immunohistochemical staining for (D) IgG and (E) IgG4. These were positive in plasma cells and IgG4/IgG ratio was >95%. (F) Immunohistochemical staining for CD20. B-cell population was positive for CD20, whereas the plasma cells lacked CD20 expression. (G) Immunohistochemistry of CD19. The plasma cells were positive for CD19. (H and I) *In situ* hybridization for immunoglobulin κ and λ light chains. Although appropriate signals were detected in internal control cells, no detectable expression of either light chain was observed in the plasma-cell population. Original magnification, x10 for panels B-I.

axis. The concomitant elevation of IgG4 and eosinophils could be explained by a common type 2 immune response associated with the underlying low-grade B-cell lymphoma. Monocytoid B-cell lymphoma, a low-grade B-cell lymphoma closely related to MZL, has previously been reported in association with marked eosinophilia (11). More recently, Zhou *et al* (12) analyzed 54 cases of B-cell lymphoma with notable tissue eosinophilia and found that nodal MZL was the most frequent subtype, accounting for 26 cases, with frequent plasmacytic differentiation and a tumor microenvironment rich in T cells and histiocytes. The neoplastic B cells themselves did not express IL-5, suggesting that non-neoplastic cells within the tumor microenvironment may have contributed to the eosinophilic response. Direct IgG4 production by the lymphoma cells alone would not readily account for the eosinophilia observed in our patient. In contrast, a lymphoma-associated type 2 immune response could provide a possible explanation for the reactive IgG4 increase and eosinophilia. Watanabe *et al* (13) also reported a B-cell lymphoma in which the eosinophil counts closely paralleled lymphoma activity. This observation is consistent with the clinical course of our patient. In our patient, the eosinophil count decreased in parallel with the treatment response. These observations are compatible with, but do not prove, a lymphoma-associated type 2 immune response.

We acknowledge several limitations of this case. First, a definitive histopathologic diagnosis of MZL could not be established, and additional analyses including CD23, SOX11, PAX5, and MYD88 mutation analysis were not performed. Second, because IgH clonality was demonstrated in whole tissue in an unstained section, the relationship between the IgG4-positive plasma cells and the neoplastic clone remains

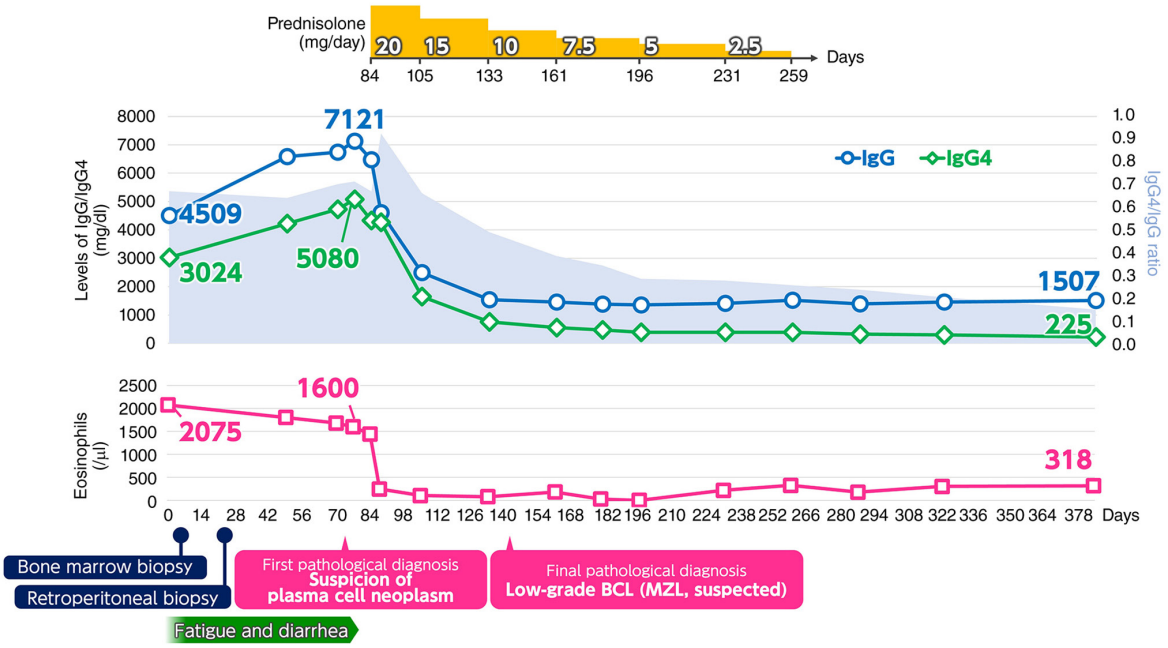


Figure 3. Clinical course, including prednisolone dose, serum IgG and IgG4 levels, the IgG4/IgG ratio, absolute eosinophil counts, and the timing of biopsies and pathological diagnoses. Serum IgG, IgG4, and eosinophil counts decreased rapidly after initiation of prednisolone and remained low during tapering. MZL, marginal zone lymphoma.

unresolved. Third, no direct assessment of type 2 immune activation was performed. Finally, the sustained remission achieved with glucocorticoid monotherapy raises some uncertainty as to whether the lesion truly represented lymphoma. Treatment strategies for indolent B-cell lymphomas vary according to the histological subtype, disease extent, and clinical symptoms, and may range from careful observation to anti-CD20 antibody-based systemic therapy (14). Glucocorticoid monotherapy is not generally regarded as definitive treatment for indolent B-cell lymphoma, making the sustained remission observed in our patient unusual. Continuing long-term follow-up is therefore warranted, and repeat biopsy with reassessment of the pathologic diagnosis will be important if the lesion recurs.

In conclusion, we reported a case of low-grade B-cell lymphoma, with MZL considered the most likely but not definitively established subtype, that initially mimicked IgG4-RD because of markedly elevated IgG4 levels, marked eosinophilia, and radiologic and pathological findings. This case highlights not only the importance of considering lymphoma and other mimickers before diagnosing IgG4-RD, but also the possibility that low-grade B-cell lymphoma may be associated with eosinophilia. We also hypothesize that the concomitant IgG4 elevation and eosinophilia raise the possibility of a lymphoma-associated type 2 immune response. Although it remains a hypothesis because no direct evidence of type 2 immune activation was obtained, recognition of a lymphoma-associated type 2 immune response may be helpful when evaluating patients with the combined findings of eosinophilia and elevated IgG4 levels.

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Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

YO conceptualized the case report, participated in the clinical management of the patient, collected and analyzed the clinical data, prepared the figures, and wrote the original draft of the manuscript. KK contributed to the hematological evaluation of the patient, interpretation of the hematological findings and treatment decisions. ST performed the retroperitoneal biopsy and contributed to the acquisition and interpretation of the surgical findings. HU, MFN and YS performed and interpreted the histopathological examination, and reviewed and interpreted the hematopathological and molecular findings. TK reviewed and interpreted the radiological findings. YU contributed to the analysis and interpretation of the clinical and pathological findings and critically revised the manuscript. HT, MYo, HM, MYa, TS and NS contributed to the clinical management of the patient and the analysis and interpretation of the clinical data. All authors critically reviewed and revised the manuscript. YO and YU confirm the authenticity of all the raw data. All authors have read and approved the final manuscript.

Ethics approval and consent to participate

Not applicable.

Patient consent for publication

Informed, voluntary and written consent for publication was obtained from the patient described in the article.

Competing interests

All the authors declare that they have no competing interests.

Use of artificial intelligence tools

During the preparation of this work, artificial intelligence tools were used to improve the readability and language of the manuscript, and subsequently the authors revised and edited the content produced by the artificial intelligence tools as necessary, taking full responsibility for the ultimate content of the present manuscript.

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