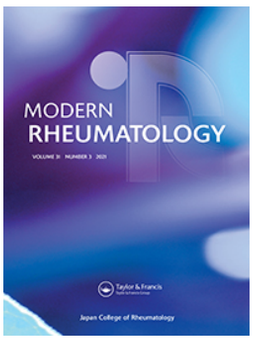

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ORIGINAL ARTICLE

The 2020 revised comprehensive diagnostic (RCD) criteria for IgG4-RD

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ABSTRACT

IgG4-related disease (IgG4-RD) is a fascinating clinical entity first reported in this century in Japan, and includes a wide variety of diseases, such as formerly named Mikulicz's disease (MD), autoimmune pancreatitis (AIP), interstitial nephritis, prostatitis and retroperitoneal fibrosis. The Japanese IgG4 team organized by the Ministry of Health, Labor and Welfare (MHLW) of Japan has published the first criteria, comprehensive diagnostic (CD) criteria for IgG-RD 2011. Thereafter, IgG4-RD has been accepted widely and many cases have been reported from all over the world. Several problems have arisen in clinical practice, however, including the difficulty obtaining biopsy samples, and the sensitivity and specificity in cut off level of serum IgG4 and impaired immunostaining of IgG4. Given these situations, the Japanese IgG4 team has updated the 2011 comprehensive diagnostic criteria for IgG4-RD and propose the 2020 revised comprehensive diagnostic (RCD) criteria for IgG4-RD, which consists of 3 domains; 1) Clinical and radiological features, 2) Serological diagnosis and 3) Pathological diagnosis. In addition, the new pathological diagnosis is composed by three sub-items including storiform fibrosis and obliterative phlebitis.

Abbreviations: IgG4-RD: IgG4-related disease; CD: comprehensive diagnostic; RCD: revised comprehensive diagnostic; MD: Mikulicz's disease; AIP: autoimmune pancreatitis; KD: kidney disease; TIN: tubulointerstitial nephritis; SS: Sjögren's syndrome; RPF: retroperitoneal fibrosis; LSG: labial salivary glands.

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IgG4-RD; diagnosis; criteria; Mikulicz's disease; autoimmune pancreatitis; sclerosing cholangitis; orbital disease

1. Introduction

IgG4 related disease (IgG4-RD) was first described in 2001 in patients with sclerosing cholangitis who had elevated serum IgG4 levels [1]. Subsequently several conditions were found to have similar clinical and laboratory features, include the disease formerly diagnosed as Mikulicz's disease (MD), autoimmune pancreatitis (AIP), hypophysitis, Riedel thyroiditis, Küttner tumor, interstitial pneumonitis, interstitial nephritis, retroperitoneal fibrosis, inflammatory aortic aneurysm and aortitis.

The clinical symptoms of IgG4-RD are dependent on the affected organs, and the severity of disease is greater in patients with serious complications, such as obstruction or compression symptoms due to organomegaly or organ dysfunction caused by cellular infiltration or fibrosis. However, elevated serum IgG4 concentration is frequently observed, and pathological examination usually reveals marked infiltration of IgG4-positive plasma cells with occasional fibrosis, a characteristic called storiform fibrosis [2,3].

The all Japan IgG4-RD team, consisting of physicians and researchers in various fields, including rheumatology, gastroenterology, nephrology, pulmonology, ophthalmology, hematology, odontology, pathology, statistics, and basic and molecular immunology, was organized by the Ministry of Health, Labor and Welfare (MHLW) of Japan. This team established comprehensive diagnostic (CD) criteria for IgG4-RD in 2011 encompassing as many aspects of IgG4-RD as possible [4]. Thereafter, IgG4-RD has been accepted widely and many cases diagnosed by CD criteria have been reported from all over the world. However, several problems have arisen in clinical practice including the difficulty obtaining biopsy samples, and the sensitivity and specificity in cut off level of serum IgG4.

Moreover, it is often difficult to evaluate IgG4-positive and IgG-positive cells due to the inappropriate staining or high background, even though immunostaining for IgG4

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and IgG is mandatory for diagnose of IgG4-RD. While, storiform fibrosis and obliterative phlebitis are reported to be unique and characteristic feature for IgG4-RD in addition to lymphoplasmacytic infiltration [5,6]. In the case with poor IgG4 and IgG staining, storiform fibrosis and obliterative phlebitis observed in hematoxylin and eosin staining are helpful for diagnosis of IgG4-RD.Following this situation, working group of the Japanese IgG4 team (Table 1) has updated the 2011 CD criteria for IgG4-RD and proposed the 2020 revised comprehensive diagnostic (RCD) criteria for IgG4-RD.

2. Original 2011 comprehensive diagnostic criteria for IgG4-RD

The original CD criteria for IgG4-RD consisted of three major items; 1) organ involvement such as diffuse/localized swelling; 2) elevated serum IgG4 concentrations greater than135 mg/dl; and 3) marked plasmacyte infiltration, defined as >10 IgG4+ cells per high powered field (HPF) and a > 40% ratio of IgG4+/IgG + cells, accompanied by fibrosis on histopathological examination [4]. Patients who fulfilled all three criteria were diagnosed with definite IgG4-RD; those who fulfilled items 1) and 3), but without increased serum IgG4 concentration, were diagnosed with probable IgG4-RD; and those who fulfilled criteria 1) and 2), with negative results on histopathology or without histopathologic examination, were diagnosed with possible IgG4-RD [4].

Several problems with these criteria have arisen in clinical practice, including the difficulty obtaining biopsy samples from some patients, especially those with AIP or retroperitoneal fibrosis, and the limited sensitivity and specificity of serum IgG4 concentrations [7,8]. These problems have been addressed by organ-specific criteria for IgG4-RD. These have included diagnostic criteria for AIP [9], IgG4-related lacrimal gland and saliva adenitis [10], IgG4-related kidney disease [11], IgG4-related sclerosing cholangitis [12], IgG4-related ophthalmic disease [13], IgG4-related respiratory disease [14], and IgG4-related large periarteritis/periarteritis and retroperitoneal fibrosis [15]. Patients with a possible or probable diagnosis of IgG4-RD could be re-diagnosed by organ-specific criteria and patients who fulfilled at least one of the organ-specific criteria could be diagnosed with definite IgG4-RD [16]. Use of an algorithm based on both CD and organ-specific criteria increased the sensitivity and specificity of diagnosing IgG4-RD in Japanese study [17].

3. The 2020 revised comprehensive diagnostic (RCD) criteria for IgG4-RD

3.1. Clinical and radiological features

Patients with IgG4-RD frequently present with simultaneous or metachronous lesions in multiple organs (Table 2). Although organ distribution varies among studies, the pancreas and salivary glands are the most frequently involved organs [7,18]. Fifty eight to 88% of patients show multiple organ involvement, with the remainder having isolated lesions, by radiological evaluation and clinical examination, respectively [18,19]. Thus, the first item in the revised CD-criteria includes involvement of one or more organs, as shown by swelling, mass or nodules defined clinically or radiographically (item 1 in Table 2). Although lymphadenopathy frequently occurs in IgG4-RD, lymph nodes are often affected in autoimmune diseases, multicentric Castleman's disease, hematological malignancies such as malignant lymphoma and lymph node metastasis of cancers. And infiltration of IgG4-positive lymphocytes is detected in some patients with B cell lymphoma [20] and with cancers of digestive organs [21,22]. Therefore, involvement of a single lymph node was not included as a criterion for IgG4-RD. IgG4-related lymphadenopathy should be diagnosed only in the case with other organ involvement.

3.2. Serological diagnosis

Although elevated serum IgG4 concentration is unique and commonly observed in IgG4-RD, it is not a gold standard for its diagnosis. Several studies have questioned the diagnostic utility of serum IgG4 concentration, as it showed a both low sensitivity of 51% [7] and a low specificity of 60% [8]. In contrast, large cohort studies from the United Kingdom (UK), Taiwan, China and Japan showed that elevated serum IgG4 concentration had overall sensitivities of more than 80% [18,19,23–25]. In addition, serum IgG4 concentration has been reported useful for distinguishing patients with IgG4-RD from those with pancreatic cancers [26].

Therefore, a serum IgG4 cut off level of 135 mg/dl was considered a unique and reliable marker predictive of IgG4-RD, and was included in the original CD criteria for IgG4-RD [4] as well as other organ-specific criteria [9–15]. Similarly, a serum IgG4 cut off level of 135 mg/dl was adopted in the 2020 RCD criteria (item 2 in Table 2).

3.3. Pathological diagnosis

Although tissue biopsies are difficult to obtain from some organs, such as the pancreas, retroperitoneum, and ocular cavity, histopathological findings are important hallmarks of IgG4-RD and indispensable for its diagnosis [2,3,5,27]. One of characteristic histopathologic features of IgG4-RD is dense infiltration of lymphocytes/plasma cells and fibrosis (sub-item 3-1 in Table 2). However, there are some pathological differences among organs. For example, abundant lymphocytes/plasma cells are observed more frequently in lymph nodes, lacrimal/salivary glands and skin than in the pancreas, lungs, bile ducts, kidneys, aorta and retroperitoneum. In contrast, fibrosis and obliterative phlebitis are rare in lymph nodes and lacrimal and salivary glands [2,3,5].

Another characteristic pathological feature is high numbers of IgG4-positive plasma cells in affected organs. However, the numbers of these cells differ among organs [17]. IgG4-positive cells are usually more abundant in

Table 1. Revised comprehensive diagnostic criteria for IgG4-RD working group, the research program for intractable disease by the ministry of health, labor and welfare (MHLW) Japan.

Kazuichi Okazaki (Chairman)	Gastroenterology	Kansai Medical University
Hisanori Umehara (Leader)	Rheumatology	Nagahama City Hospital
Shigeyuki Kawa	Gastroenterology	Matsumoto Dental University
Hiroki Takahashi	Rheumatology	Sapporo Medical University
Hiroshi Goto	Ophthalmology	Tokyo Medical University
Shoko Matsui	Respirology	University of Toyama
Nobukazu Ishizaka	Cardiology	Oosaka Medical College
Takashi Akamizu	Endocrinology	Wakayama Medical University
Yasuharu Sato	Pathology	Okayama University
Mitsuhiro Kawano	Rheumatology and Nephrology	Kanazawa University

Table 2. The 2020 Revised comprehensive diagnostic (RCD) criteria for IgG4-RD.

[Item 1] clinical and radiological features
One or more organs show diffuse or localized swelling or a mass or nodule characteristic of IgG4-RD. In single organ involvement, lymph node swelling is omitted.
[Item 2] serological diagnosis
Serum IgG4 levels greater than 135 mg/dl.
[Item 3] pathological diagnosis
Positivity for two of the following three criteria:
① Dense lymphocyte and plasma cell infiltration with fibrosis.
② Ratio of IgG4-positive plasma cells /IgG-positive cells greater than 40% and the number of IgG4-positive plasma cells greater than 10 per high powered field
③Typical tissue fibrosis, particularly storiform fibrosis, or obliterative phlebitis
Diagnosis:
Definite: 1) +2) +3)
Probable: 1) +3):
Possible: 1) +2)

Explanatory note 1: Combination of organ-specific diagnostic criteria*.
Patients with a possible or probable diagnosis by comprehensive diagnostic criteria who fulfill the organ-specific criteria for IgG4-RD are regarded as being definite for IgG4-RD.
*Diagnostic criteria according to the IgG4-related organ:.
① International consensus diagnostic criteria for autoimmune pancreatitis[7], ② IgG4-related lacrimal gland, saliva adenitis diagnostic criteria[8], ③ Diagnostic criteria for IgG4-related kidney disease[9], ④ Clinical diagnostic criteria of IgG4-related sclerosing cholangitis 2012[10], ⑤Diagnostic criteria for IgG4-related ophthalmic disease[11], ⑥Diagnostic criteria for IgG4-related respiratory disease[12], ⑦ Diagnostic criteria for IgG4-related large periarteritis/periarteritis and retroperitoneal fibrosis[13].
Explanatory note 2: exclusion diagnosis.
1) It is important to acquire tissue samples from each involved organ to distinguish malignant tumors (e.g. cancer, malignant lymphoma) and similar benign conditions (e.g. Sjögren syndrome, primary sclerosing cholangitis, multicentric Castleman's disease, secondary retroperitoneal fibrosis, granulomatosis with polyangiitis, sarcoidosis, eosinophilic granulomatosis with polyangiitis).
2) It is important to exclude an infectious- or inflammation-related disease in patients with high fever, highly elevated CRP and neutrophilia.
Explanatory note 3: pathologic diagnosis.
1) The numbers of IgG4-positive cells are usually more abundant in resected organs and partially enucleated tissue than in tissue samples obtained by needle biopsy or endoscopic biopsy. Thus, it is important to not be too particular about cell number and to provide a precise judgment.
2) Storiform fibrosis is defined as spindle-shaped cells, inflammatory cells and fine collagen fibers forming a flowing arrangement. Obliterative phlebitis is defined as fibrous venous obliteration with inflammatory cells. Both are helpful for a diagnosis of IgG4-RD. ① and ③ without ② can only be applied in a case with poor IgG4 and/or IgG staining.
Explanatory note 4: steroid reactivity.
Steroid trial is not recommended. However, if patients do not respond to initial steroid therapy, the diagnosis should be reconsidered.

lymph nodes, lacrimal and salivary glands and skin than in the pancreas, lung, bile ducts, kidneys, aorta and retroperitoneum [5,17]. In addition, the numbers of IgG4-positive cells may depend on tissue size, as samples obtained by needle biopsy are much smaller than those obtained by tissue resection. Moreover, infiltration by IgG4-positive cells has been observed in patients with conditions other than IgG4-RD, such as rheumatoid arthritis, anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis, and atopic dermatitis [28]. Therefore, the ratio of IgG4-positive to IgG-positive cells is more diagnostic than the absolute numbers of IgG4-positive cells. In almost all patients with IgG4-RD, the ratio of IgG4-positive plasma cells among IgG-positive cells are more than 40% (sub-item 3-2 of Table 2) [4]. Although immunostaining of IgG4 and IgG is important for diagnose of IgG4-RD, there are no standard for antibodies or staining procedure for IgG4 immunostaining. Therefore, it is often difficult to evaluate IgG4-positive and IgG-positive cells due to the background staining, especially in minute samples [6]. On the other hand, storiform fibrosis and obliterative phlebitis are reported to be unique and characteristic feature for IgG4-RD in addition to lymphoplasmacytic infiltration [5,6]. Therefore, storiform fibrosis and obliterative phlebitis observed in hematoxylin and eosin staining are helpful for diagnosis of IgG4-RD, especially in the case with poor IgG and/or IgG4 staining. The 2020 RCD criteria includes additional pathological criterion of storiform fibrosis and obliterative phlebitis (sub-item 3-3 in Table 2). Storiform fibrosis is defined as spindle-shaped cells, inflammatory cells and fine collagen fibers forming a flowing arrangement. Obliterative phlebitis is defined as fibrous venous obliteration with inflammatory cells. Examples of each are shown in Figure 1. Thus, a pathological diagnosis was dependent on the three afore-mentioned independent items, and 2 out of 3 sub-items must be satisfied for diagnosis of definite IgG4-RD.

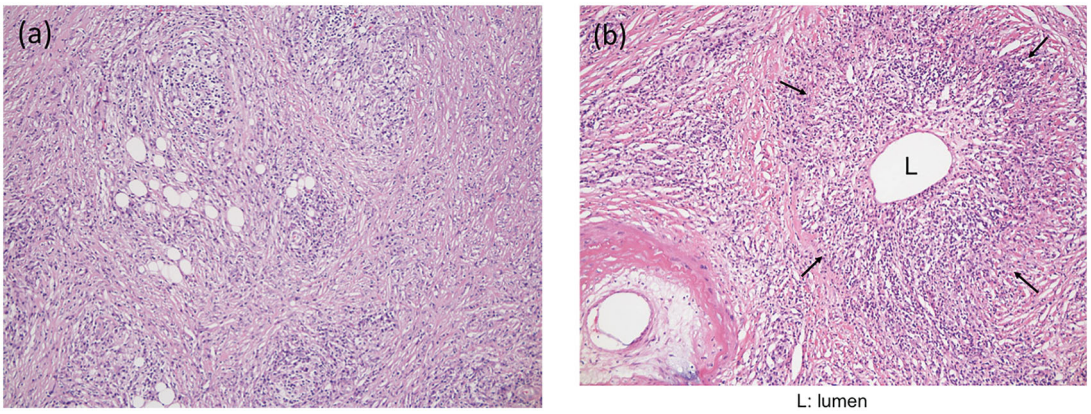

Figure 1. Storiform fibrosis and obliterative phlebitis. (a) Storiform fibrosis: spindle-shaped cells, inflammatory cells and fine collagen fibers are forming a flowing arrangement. The infiltration of plasma cells is easy to recognize. Eosinophils are also intermingled (hematoxylin and eosin stain). (b) Obliterative phlebitis: fibrous venous obliteration with inflammatory cells (arrows). L:lumen.

Eventually, IgG4-RD is definitively diagnosed in patients who fulfilled all three RCD criteria: 1) organ involvement, such as diffuse/localized swelling; 2) serum IgG4 concentration >135 mg/dl; 3) positive for 2 out of 3 pathological subitems. As the same as CD criteria, patients who fulfilled items 1) and 3) are diagnosed with probable IgG4-RD and those who fulfilled items 1) and 2) with possible IgG4-RD. In addition, patients diagnosed with probable or possible and who met the organ-specific criteria for IgG4-RD are subsequently diagnosed with definite IgG4-RD. The combination of CD and organ-specific criteria reinforce each other, increasing the sensitivity and specificity of IgG4-RD diagnosis [16,17].

4. Final thoughts

Recently, the American College Rheumatology (ACR) and European League Against Rheumatism (EULAR) formulated the 2019 ACR/EULAR IgG4-RD Classification Criteria for international use [29,30]. Although patients with IgG4-RD frequently present with simultaneous or metachronous lesions in various organs, the Classification Criteria were developed only for patients with frequently involved 10 organs (e.g. pancreas, bile ducts, orbits, lacrimal glands, major salivary glands, retroperitoneum, kidney, aorta, pachymeninges, thyroid gland), and excluded organs that were not frequently affected (prostate, meninges and skin, etc.) to maximize the specificity. In addition, the ACR/EULAR classification criteria have defined exclusion criteria before analysis to eliminate patients with infections, malignancies and hematological and immunological diseases. Exclusion criteria include specific autoantibodies such as anti-dsDNA, anti-SSA/Ro or SSB/La antibody and MPO- or PR3-ANCA. Thereby, patients with some autoantibodies were excluded prior to analysis of IgG4-RD, even though they are not accurate autoimmune diseases.

The overall purpose of classification criteria is to maximize the specificity of classification, even if sensitivity is not good. However, physicians must diagnose exactly and treat all patients in actual clinical practice. Therefore, it is important to understand the general features of IgG4-RD such as organ involvement, clinical symptoms, radiological abnormalities, laboratory data and pathological characteristics, and to diagnose patients carefully using either of the criteria. In addition, the RCD criteria shouold be validated for sensitivity and specificity in future study.

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Conflict of interest

None.

Additional Information

We pursued and obtained public comments for The 2020 RCD criteria for IgG4 from Japanese Society of Internal Medicine, Japan College of Rheumatology, The Japanese Society of Gastroentelogy, Japan Pancreas Society, Japanese Society for Sjogren's Syndrome, and Japanese Association of IgG4-related disease.

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