Comparison of Diagnostic Criteria for AIP

Korean Criteria : Comparison with Those of Japan and USA

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Autoimmune pancreatitis (AIP) is a type of chronic pancreatitis characterized by an autoimmune manifestations revealed on imaging, laboratory, histologic, and clinical study. In the past 10 years, interest and reports of autoimmune pancreatitis has grown rapidly because of an increasing ability to diagnose it with growing awareness of the disease entity and the use of new markers of disease and pancreatic biopsy. Autoimmune pancreatitis is a rare systemic fibroinflammatory disease which can affect not only the pancreas, but also a variety of organs such as the bile ducts, salivary glands, retroperitoneum, and lymph nodes. Organs affected by AIP have a lymphoplasmacytic infiltrate rich in IgG4-positive cells. This inflammatory process responds dramatically to oral steroid therapy.

The overwhelming majority of reports of AIP have come from Asian countries (e.g. Japan and Korea), and the prevalence of AIP in Japan was estimated to be 0.82 per 100,000. So, it is reasonable to establish a common diagnostic criteria in Asia which has a lot cases and clinical experience.

In 2002, Japan Pancreas Society (JPS) proposed the following diagnostic criteria for AIP (Table 1) (1) diffuse irregular narrowing of the main pancreatic duct (MPD) and enlargement of the pancreas (mandatory); (2) serum elevation of α1-globulin and/or IgG, and/or the presence of autoantibodies; and (3) fibrosis with lymphoplasmocytic infiltration in pancreatic tissue. Several groups cite the diagnostic criteria proposed by JPS (AIP Criteria 2002), whereas others including Korea use their own criteria in reporting AIP. The absence of consistent, uniform criteria has made it difficult to compare studies from different centers and to establish evidence characterizing AIP.

The purpose of this review is to make comparisons among different diagnostic criteria and to help in making a common diagnostic criteria between Japan and Korea.

Diagnostic Criteria for AIP

1. Japan Criteria

As previously mentioned, the Japan Pancreas Society proposed diagnostic criteria for AIP in 2002 (Table 1). The 2002 criteria were based on the results of imaging studies, laboratory tests, and histological findings of pancreatic tissue. This diagnostic criteria has been debated because, with increased experience, a significant number of patients have been diagnosed with AIP who do not meet the above criteria. Particularly, AIP has been diagnosed in a number of patients who have only focal narrowing of the main pancreatic duct that takes up less than one-third of the pancreatic gland, in addition, patients’ IgG4 concentration might be elevated without the total IgG concentration being elevated. For these reasons,
Table 1. Diagnostic criteria for AIP of 2002, by the Japan Pancreas Society (AIP Criteria 2002)

1. Diffuse or segmental narrowing of the MPD with an irregular wall (more than one-third the length of the entire pancreas) along with diffuse or localized enlargement of the pancreas by imaging studies
2. High serum γ-globulin and/or IgG, or the presence of autoantibodies such as antinuclear antibodies and rheumatoid factor
3. Marked interlobular fibrosis and prominent infiltration of lymphocytes and plasma cells in the periductal area, occasionally with lymphoid follicles in the pancreas

For diagnosis, criterion 1 must be present together with criterion 2 and/or 3.

Table 2. Clinical diagnostic criteria for AIP (AIP Criteria 2006)

1. Diffuse or segmental narrowing of the MPD with irregular wall and diffuse or localized enlargement of the pancreas by imaging studies, such as abdominal US, CT, and magnetic resonance imaging
2. High serum γ-globulin, IgG, or IgG4, or the presence of autoantibodies such as antinuclear antibodies and rheumatoid factor
3. Marked interlobular fibrosis and prominent infiltration of lymphocytes and plasma cells in the periductal area, occasionally with lymphoid follicles in the pancreas

Diagnosis of AIP is established when criterion 1 and criterion 2 and/or 3 are fulfilled. However, it is necessary to exclude malignant diseases such as pancreatic or biliary cancers.

Table 3. Diagnostic criteria for AIP in Asan Medical Center

Criterion I. Pancreatic imaging (essential)
   (1) Diffuse enlargement (swelling) of the pancreas on CT and
   (2) Diffuse or segmental irregular narrowing of the MPD on ERCP

Criterion II. Laboratory findings
(1) Elevated levels of IgG and/or IgG4 or
(2) Detected autoantibodies

Criterion III. Histopathologic findings
Fibrosis and lymphoplasmacytic infiltration

Criterion IV. Response to steroid
Definite diagnosis: Criterion I and any of criterion II–IV

members of the Japan Pancreas Society proposed revised criteria in 2006, which introduced local ductal narrowing and elevation of IgG4 levels alone as diagnostic criteria (Table 2). They expanded the diagnostic criteria to include more patients and presented a system for stratifying patients on the basis of the strength of the evidence for AIP into (1) definite, (2) probable, or (3) possible cases of AIP. They include a diagnostic trial of steroid therapy even when serologic markers or pathologic findings do not fulfill the Japanese AIP Criteria 2002 or are not available, providing that pancreatic images are typical of AIP. Patients with possible or probable AIP are reassigned to the definite AIP class if dramatic resolution of pancreatic ductal narrowing is observed after steroid therapy.

In 2007, Korean AIP committee of Korean Pancreaticobiliary Society made a new diagnostic criteria to develop improved diagnostic criteria (Table 4). This new criteria adopted a concept of extra-pancreatic involvement and gave more chances to...
**Table 4. Korean criteria for autoimmune pancreatitis**

<table>
<thead>
<tr>
<th>Category</th>
<th>Criteria</th>
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<tbody>
<tr>
<td><strong>Definite Diagnosis: Criterion I together with any of criterion II to IV</strong></td>
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<tr>
<td>Criterion I. Imaging (Both required)</td>
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<tr>
<td>1. Imaging (CT or MRI) of pancreatic parenchyma;</td>
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<tr>
<td>Diffusely/segmentally/focally enlarged gland, occasionally with mass and/or hypoattenuation rim</td>
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<tr>
<td>2. Imaging (ERCP or MRCP) of pancreaticobiliary ducts;</td>
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<tr>
<td>Diffuse/segmental/focal pancreatic ductal narrowing, often with the stenosis of bile duct</td>
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<td>Criterion II. Serology (One required)</td>
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<tr>
<td>1. Elevated level of serum IgG or IgG4</td>
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<tr>
<td>2. Detected autoantibodies</td>
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<tr>
<td>Criterion III. Histopathology of pancreatic/extrapancreatic Lesions (One required)</td>
<td></td>
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<tr>
<td>1. Lymphoplasmacytic infiltration &amp; fibrosis, often with obliterative phlebitis</td>
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</tr>
<tr>
<td>2. Presence of abundant (&gt;10 cells/HPF) IgG4-positive plasma cells</td>
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<tr>
<td>Criterion IV. Response to steroids</td>
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<tr>
<td>1. Resolution/marked improvement of pancreatic/extrapancreatic lesion with steroid therapy</td>
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<tr>
<td><strong>Probable Diagnosis: Criterion V or VI</strong></td>
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<td>Criterion V.</td>
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<td>1. Unexplained pancreatic disease but only with characteristic pancreatic histology</td>
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<tr>
<td>Criterion VI. (Both required)</td>
<td></td>
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<tr>
<td>1. Other organ involvement and/or serologic abnormalities</td>
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<tr>
<td>2. Various atypical pancreatic imaging suggesting chronic pancreatitis with negative workup for known etiologies</td>
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**Table 5. Diagnostic criteria for AIP (HISORt criteria)**

<table>
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<tr>
<th>Category</th>
<th>Criteria</th>
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<tr>
<td>Histology fibrosis (LPSP)</td>
<td>At least one of the following:</td>
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<tr>
<td>(1) Periductal lymphoplasmacytic infiltration with obliterative phlebitis and storiform fibrosis (LPSP)</td>
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<tr>
<td>(2) Lymphoplasmacytic infiltrate with storiform fibrosis showing abundant (≥ 10 cells/HPF) IgG4-positive cells</td>
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<tr>
<td>Pancreatic imaging</td>
<td>Typical: diffusely enlarged gland with delayed (rim) enhancement; diffusely irregular, attenuated MPD</td>
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<td>Others:</td>
<td>Focal pancreatic mass/enlargement; focal pancreatic duct stricture; pancreatic atrophy; pancreatic calcification; or pancreatitis</td>
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<tr>
<td>Serology</td>
<td>Elevated serum IgG4 level (normal, 8 – 140 mg/dl)</td>
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<tr>
<td>Other organ involvement</td>
<td>Hilar/intrahepatic biliary strictures, persistent distal biliary stricture, parotid/lacrimal gland involvement, mediastinal lymphadenopathy, retroperitoneal fibrosis</td>
</tr>
<tr>
<td>Response to steroid therapy</td>
<td>Resolution/marked improvement of pancreatic/extrapancreatic manifestation with steroid therapy</td>
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<tr>
<td>Diagnosis</td>
<td>1. Group A: diagnostic histology alone</td>
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<tr>
<td>2. Group B: typical imaging features and elevated serum IgG4</td>
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<tr>
<td>3. Group C: unexplained pancreatic disease with serology or other organ involvement and response to a steroid</td>
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HPF, high power field

*With negative work-up for known etiologies for pancreatic disease, especially pancreatic/biliary cancer

*Radiologic evidence of organ involvement can be confirmed by biopsy showing lymphoplasmacytic infiltrate with abundant IgG4-positive cells or resolution/improvement with steroid therapy

*Steroid therapy should be given only to patients with a negative work-up for known etiologies for pancreatic disease and only to those in whom the response can be objectively assessed. It should not be used as a substitute for a thorough search for etiology
3. USA Criteria

Most recently, Chari et al.\textsuperscript{15} of the Mayo Clinic proposed new diagnostic criteria (HISORt criteria) for diagnosis of AIP (Table 5). The HISORt criteria consist of five categories: (1) histology, (2) imaging, (3) serology, (4) the presence of other organ involvement, and (5) response to steroid therapy. In response to the wide variation of imaging features and diverse clinical presentation, the HISORt criteria particularly emphasize the need for acquiring tissue specimens to enhance the diagnostic accuracy of AIP. The HISORt criteria are considered to be unique among the various diagnostic criteria in that they can be completely satisfied by histological findings, including those derived from immunohistochemistry. The imaging criteria are divided into typical and atypical features. The typical imaging features are a diffusely enlarged gland with delayed (rim) enhancement on CT/MR, and a diffusely irregular, attenuated main pancreatic duct on ERCP. Atypical imaging features are focal pancreatic mass, focal pancreatic duct stricture, pancreatic atrophy, and calcification. Even with atypical imaging, a case which responds to a steroid can be diagnosed as AIP in the HISORt criteria. The measurement of the serum IgG4 level is only included in the serological criteria. The involvement of other organs, such as hilar/intrahepatic/distal biliary strictures, parotid/lacrimal gland involvement, mediastinal lymphadenopathy, retroperitoneal fibrosis, and the response to steroid therapy are also included in the diagnostic criteria.

Korean Criteria vs Japan Criteria

1. Imaging

Although the USA (HISORt) criteria place emphasis on histological features, the Korean and revised Japanese criteria are image based and thereby can be argued to be more clinically applicable, given the widespread practice and availability of pancreatic imaging (ie, computed tomographic scan and ERCP) in comparison with pancreatic biopsy. The diagnosis of autoimmune pancreatitis is most commonly considered when findings on imaging are highly suggestive of the disease. In patients whose findings on imaging are suggestive of autoimmune pancreatitis, alcohol-induced chronic pancreatitis or pancreatic cancer are the most common conditions that should be differentiated from autoimmune pancreatitis. However, when cross-sectional imaging shows diffuse pancreatic enlargement and ERCP reveals a long attenuated segment of the pancreatic duct, autoimmune pancreatitis should be strongly considered. Although a direct pancreatography is mandatory to satisfy the imaging criterion of the Korean and Japanese diagnostic criteria, an additional pancreatography is not considered troublesome because most patients with AIP show obstructive jaundice, and ERCP examination is necessary for evaluation and treatment of obstructive jaundice. Both the Korean and Japanese criteria concur on the point that the imaging criterion is essential for diagnosis. Typical features found on imaging studies are diffuse enlargement of the pancreas, and diffuse or segmental irregular narrowing of the main pancreatic duct.\textsuperscript{16-19} The concurrence of these two features is hardly ever seen in other pancreatic disorders, and is therefore highly specific for AIP. Therefore, the presence of this unique combination of pancreatic features on computed tomographic scan and ERCP should trigger a
high index of suspicion of AIP.

The primary difference between the former Japanese (2002) and Korean criteria lay in the extent of main pancreatic duct involvement necessary for diagnosis. The former Japanese criteria required that the extent of the pancreatic ductal narrowing should be larger than one third of the full length of the main pancreatic duct to avoid the possible inclusion of pancreatic cancer. This was based on a study showing that the segments of narrowed main pancreatic duct in cases of pancreatic inflammatory mass were longer than those cases of pancreatic malignancy.\(^\text{20}\) The revised Japanese criteria (2006), however, deleted the extent limitation of main pancreatic duct involvement, although they still comment that more than one third of the entire length of the pancreatic duct is narrowed in typical AIP. The Korean criteria discarded the minimum extent of ductal involvement requirement on the basis of commonly reported cases of AIP with less than one third of the main pancreatic duct being involved. Even without this stipulation, the exclusion of pancreatic cancer may be feasible by noting imaging and serological features, including delayed homogenous enhancement of the pancreatic parenchyma, none-to-mild upstream duct dilatation despite the presence of a long stricture visualized on direct pancreatogram,\(^\text{21,22}\) and increased serum levels of IgG4.\(^\text{23}\)

Actually, the revised Japan criteria and new Korean criteria in imaging have a close similarity on the essential need of pancreatic imaging for the diagnosis and similar findings of pancreatic imaging.

2. Serology

The former Japanese serological criterion for the diagnosis of AIP included serum \(\gamma\)-globulin and IgG measurements except IgG4. In contrast, measurement of serum IgG4, a subtype of IgG, level is now used in the Korean, USA, and revised Japanese criteria. This is based on the reports demonstrating superior sensitivity of serum IgG4 level compared with serum IgG or \(\gamma\)-globulin level for the diagnosis of AIP. Serum IgG4 level was found to be elevated in AIP patients despite normal IgG or \(\gamma\)-globulin level.\(^\text{24-27}\) Kawa and Hamano\(^\text{25}\) also reported that the sensitivity of \(\gamma\)-globulin and IgG was 59.1% and 70.5%, respectively, whereas the sensitivity of IgG4 alone was 90.9%. In contrast to the Japanese criteria, the measurement of serum \(\gamma\)-globulin level was not included in the Korean criteria based on the study showing that the sensitivity of serum \(\gamma\)-globulin was much inferior to that of serum IgG or IgG4 level, which was consistently elevated in the presence of elevated serum \(\gamma\)-globulin levels.\(^\text{25}\)

3. Histology

In the Korean and Japan histological criteria, the characteristic features of AIP are fibrosis and lymphoplasmacytic infiltration of the pancreas. These histological features can be present in alcoholic chronic pancreatitis, and may not be specific to AIP.\(^\text{28}\) But, in the recently proposed HISORt criteria of Mayo Clinic, AIP can be diagnosed on histological findings alone.\(^\text{15,29}\) The pathognomonic histological features of AIP [which is also known as lymphoplasmacytic sclerosing pancreatitis (LPSP)], are a periductal lymphoplasmacytic infiltrate associated with storiform fibrosis, and obliterative phlebitis wherein the lymphoplasmacytic infiltrate surrounds pancreatic venules while sparing arterioles.\(^\text{30}\) The lymphoplasmacytic infiltrate in AIP has abundant IgG4-positive cells on immunostaining.\(^\text{31}\)

The diagnosis of AIP could not be made from histological findings alone in the Kim and JPS systems, but should be made in combination with the essential imaging criteria listed (Table 2, 3). The new Korean criteria (2007) in histopathology adopted a concept of LPSP, IgG4 immunohistochemical
staining, and extrapancreatic involvement according to the accumulation of new information about histopathologic findings in AIP. The new Korean criteria still needs a combination of imaging and histopathologic finding for the definite diagnosis of AIP, but AIP can be diagnosed as a probable diagnosis if it has a characteristic pancreatic histology.

4. Response to steroid

This criteria is a notably different point between Korean and Japan criteria. The response to oral steroids may provide circumstantial evidence of an underlying autoimmune pathogenesis. The relief of pancreatic ductal narrowing by steroid administration may be a unique and specific finding that cannot be seen in any other type of chronic pancreatitis or pancreatic cancer. Recovery of the duct narrowing should be accompanied by resolution of periductal fibrosis in addition to remission of pancreatic inflammation. Well-preserved collagen type IV, a component of the intact basement membrane, may be attributable to accurate regeneration of tissue and reversible fibrosis in AIP. Because marked improvement of pancreatic ductal narrowing can be observed as early as 2 weeks after initiation of steroid therapy, steroid trial may be a practical diagnostic tool and a therapeutic one. The response to the steroid is defined as improvement in clinical symptoms, negative conversion of detected autoantibodies, normalization of elevated levels of IgG, and reversion of abnormal pancreatic imaging, including CT and endoscopic retrograde pancreatography.

In contrast to the Korean and USA criteria for the diagnosis of AIP, the revised Japanese criteria (2006) do not include the response to steroid therapy as a diagnostic component because inclusion of steroid responsiveness in the criteria may encourage the use of easy therapeutic diagnostic techniques just to distinguish AIP from pancreatic cancer. This is probably owing to the possible improvement in diffuse pancreatic swelling in patients with pancreatic cancer when treated with steroids. There is also concern about the increased delay in the diagnosis of malignancy if weeks are spent trying steroid therapy. Although pancreatic swelling developed by upstream pancreatitis associated with pancreatic cancer may be improved, pancreatic ductal narrowing associated with ductal adenocarcinoma is not relieved even with steroid therapy. Another reason for the inclusion of steroid response in the Korean criteria is that a considerable proportion of AIP patients may not fulfill the serological and/or histological criteria, depending on the stage and activity of the disease.

The most crucial issue when caring for patients with suspected autoimmune pancreatitis is to differentiate autoimmune pancreatitis from pancreatic cancer, because pancreatic cancer requires surgery, whereas autoimmune pancreatitis responds dramatically to steroid treatment. Especially, a focal type of AIP has focal swelling with a localized stenosis of the main pancreatic duct and upstream dilatation without diffuse swelling of pancreas. It is very hard to differentiate pancreatic cancer and AIP in these cases. In these cases, the patients inevitably had to take major operation such as Whipple procedure or pancreaticoduodenectomy if there were no supporting evidence of serology and histopathology about AIP. Now, Korean criteria recommend short term (2 weeks) steroid trial to diagnosis AIP with strict regulation. When the presumptive diagnosis of AIP is made on the basis of imaging alone without supporting evidence of serology or histopathology about AIP, EUS±FNA is recommended to exclude malignancy before steroid therapy. For these cases, short-interval (2 weeks) imaging after the initiation of steroid therapy must be also used to assess steroid responsiveness. In the assessment of steroid responsiveness, relief of
pancreatic ductal narrowing and/or resolution of the pancreatic mass is critical. If imaging studies fail to show improvement, the diagnosis of AIP should be reevaluated and consider operation. Two weeks delay of operation is not critical for the prognosis of pancreatic cancer patients.

Korean Criteria vs USA (HISORt) Criteria

1. Imaging

In contrast to the Korean and Japanese imaging criteria, the HISORt is divided into typical and atypical features. The typical feature criterion is essentially the same as the Korean and Japanese imaging criteria, whereas the atypical features include focal pancreatic mass and duct stricture and pancreatic calcification and atrophy. The AIP with focal pancreatic mass with concomitant localized stricture constitutes what is now referred to as the “focal” type of AIP or “tumefactive” autoimmune pancreatitis.

The focal type of AIP is characterized by focal swelling of the pancreas and localized stenosis with resultant upstream duct dilatation, and thereby is commonly mistaken for pancreatic cancer. Pancreatic calcification and atrophy are comparable to rare calcification and diffuse enlargement of the pancreas in the typical form of AIP. In recent cases of AIP, and in particular in relapsed ones, pancreaticolith formation in the pancreatic duct has been reported, suggesting that late-stage AIP may show pancreatic calcifications, atrophy, and pancreaticolith formation, as seen in ordinary chronic pancreatitis.

The imaging criterion of the HISORt criteria is not essential for the diagnosis of AIP compared to Korean and Japan criteria. But, the diagnosis of AIP is most commonly considered when findings on imaging are highly suggestive of the disease. This may be because HISORt criteria originally based on the histopathologic finding instead of clinical and practical finding of Korean and Japan criteria.

2. Serology

The HISORt criteria rely only on serum IgG4 level as a serological criterion without consideration of IgG or γ-globulin measurements. The logic of this sole reliance on IgG4 levels is supported by a report stating that the overall sensitivity of combined γ-globulin, IgG, and autoantibodies was comparable to that of IgG4 alone. According to this report, the sensitivity of serum IgG4 level measurement alone seems to be similar to that of the combination of other serological measurements. This may be the reason that the HISORt criteria include the measurement of only the serum IgG4 level for the serological criterion. However, this IgG4 measurement alone may remain far from perfect because the incidence of elevated serum IgG4 level in AIP patients has been recently reported to be at most around 70%. In Kim’s report, the sensitivity of serum IgG4 was 73.3% (22/30); in other reports, the sensitivity was 71%, 68%, and 67%, respectively. Kamisawa et al. reported that patients with elevated IgG4 tended to involve more extrapancreatic organs in addition to the pancreas itself when comparing the patients in relation to presence or absence of elevated serum IgG4 concentrations. This means that the serum IgG4 level is influenced by the activity of the disease in patients with AIP.

In the Korean criteria, although serum IgG4 level was more sensitive than serum IgG level, the measurement of both serum IgG and IgG4 levels is included for the following reasons (Table 4). First, values for serum IgG are available more readily than those for serum IgG4, allowing for quicker clinical decision making. Second, despite superior sensitivity reported for serum IgG4 level, the serum IgG level showed the similar specificity to the serum IgG4 level for distinguishing AIP from pancreatic cancer.
In the HISORt criteria, autoantibody measurement is not included for the serological criterion (Table 5). In the Korean criteria, however, the rationale for additional inclusion of autoantibody measurement is based on the improved diagnostic sensitivity when using a combination of serum immunoglobulin and autoantibody levels. In Kim’s report, 2 of 31 AIP patients showed positive autoantibody including antinuclear antibody and rheumatoid factor despite normal serum IgG and IgG4 levels. Therefore, in the diagnosis of AIP, simultaneous measurement of autoantibodies in addition to serum immunoglobulin levels may serve a complementary role.

3. Histology

In the recently proposed HISORt criteria, however, AIP can be diagnosed on histological findings alone (Table 5). In the HISORt criteria, the primary histological finding is lymphoplasmacytic sclerosing pancreatitis (LPSP), which is characterized by dense periductal lymphoplasmacytic infiltration, marked perilobular and intralobular fibrosis, and obliterative phlebitis. Equally important to a diagnosis of LPSP is the lack of changes which are typically associated with other types of chronic pancreatitis, including duct dilatation, stones, or fat necrosis. The second histological component of the HISORt criteria is the abundant (≥10 cells/high power field (HPF)) presence of IgG4-positive cells within the lymphoplasmacytic infiltrate of the pancreas.

The criteria emphasizing the histological findings of LPSP may have some limitations from a practical point of view. The histological diagnosis of LPSP has most commonly been made on specimens obtained by surgical resection. The duct-centric nature of the inflammatory process and the phlebitis may not be evident on the core biopsy specimens. Weber et al. reported that a specific diagnosis of LPSP required a larger tissue sample, at least more than that obtained in a wedge biopsy. The requirement for a larger specimen may considerably diminish the clinical practicality of the HISORt histological criteria. Without a laparotomy, obtaining a pancreatic specimen can be difficult. Since the manifestations of the disease have a patch distribution, such sampling may yield false negative results. Therefore, the role of a histopathological examination of the pancreas in patients with suspected autoimmune pancreatitis may be to exclude other diseases such as cancer rather than to provide definitive evidence for a diagnosis of autoimmune pancreatitis. In addition, the correct pathological diagnosis of LPSP requires a skilled pathologist.

4. Other organ involvement

AIP has traditionally been reported to be “associated” with a number of other disorders such as primary sclerosing cholangitis (PSC), Sjögren’s syndrome, and retroperitoneal fibrosis. However, more recent studies suggest that the “associations” are in fact manifestations of AIP that mimic other well-described diseases. Kamisawa et al. have proposed that AIP is a systemic disease which predominantly afflicts the pancreas, and have called it IgG4 disease. This concept is supported by the findings of abundant infiltration with IgG positive cells in a number of organs in AIP. The organs most commonly involved are the biliary ductal system, where it causes PSC-like intra- and extrahepatic biliary strictures, the salivary glands, where it produces a Sjogren’s syndrome-like picture, and the retroperitoneum, where it causes retroperitoneal fibrosis.

The prevalence of the other organ involvement may be ethnically different. It was around 20% and retroperitoneal fibrosis was the most commonly associated autoimmune disease in Korean. The new Korean criteria adopted a concept of extrapancreatic involvement of AIP (Table 4).
5. Response to steroid therapy

The pancreatic, \(^{24}\) biliary, \(^{51}\) and salivary gland diseases \(^{54}\) associated with infiltration with IgG4-positive cells respond, often dramatically, to steroid therapy, whereas the diseases they mimic do not. This unique response to steroid therapy may be useful in the diagnosis of AIP. The Korean criteria also use this criteria as a useful diagnostic criteria for the diagnosis of AIP.

Conclusions

This review has discussed and compared three sets of criteria for the diagnosis of AIP. Japanese AIP criteria 2006 are based on the minimum consensus to avoid misdiagnosing malignancy, but Korean AIP criteria 2007 are aimed high sensitivity to diagnose the wide spectrum of manifestations of AIP. A diagnostic criteria for AIP is still evolving. An integrated diagnostic criteria is strongly advocated so that more patients can have an opportunity to receive medical treatment which will avoid any unnecessarily invasive procedure.

REFERENCES