EXCLI Journal 2012;11:651-658 – ISSN 1611-2156
Received: May 18, 2012, accepted: September 09, 2012, published: September 17, 2012

Original article:

IGG4-RELATED DISEASE AND THE CURRENT STATUS OF DIAGNOSTIC APPROACHES

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ABSTRACT

IgG4-related disease is a newly recognized systemic disease characterized by involving a wide range of organs. It includes the pancreas, biliary tree, salivary glands, periorbital tissues, upper aerodigestive tract, retroperitoneum, mediastinum, aorta, soft tissue, skin, central nervous system, breast, kidneys, prostate, lungs and lymph nodes. The elevated serum titer of immunoglobulin G4 (IgG4), which is the least common (3 % to 6 %) of the 4 subclasses of IgG, is a special marker for IgG4-related disease. However, its entity is still unknown. This article reviewed the literature to learn the IgG4-related diseases and their current status of diagnostic approaches.

Keywords: IgG4-related disease, autoimmune pancreatitis, IgG4-related sclerosing cholangitis, IgG4-related retroperitoneal fibrosis

INTRODUCTION

IgG4-related disease is a newly recognized fibroinflammatory condition characterized by tumefactive lesions, a dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells, storiform fibrosis, often but not always elevated serum IgG4 concentration and a favorable response to steroids (Stone et al., 2012). IgG4-related disease affects predominantly middle-aged and elderly patients, with male predominance. The patients present with symptoms referable to the involvement of one or more sites either simultaneously or serially, usually in the form of mass lesions, including the pancreas, biliary tree, salivary glands, periorbital tissues, kidneys, lungs and lymph nodes. In this review, we will discuss the new information on autoimmune pancreatitis,sclerosing cholangitis, retroperitoneal fibrosis and their current diagnostic approaches in detail.

Autoimmune pancreatitis

Autoimmune pancreatitis (AIP) is a unique and rare form of chronic pancreatitis (less than 5 % of all chronic pancreatitis). While it's wildly known was increasing worldwide. In 1961, Sarles et al. reported one case of pancreatitis associated with obstructive jaundice and hyperglobulinemia, suggestive of an underlying autoimmune process (Sarles et al., 1961). Until 1995, Yoshida et al. reported the term of 'Autoimmune Pancreatitis', which described a diffuse enlargement of the pancreas and irregular narrowing of the pancreatic duct, serologically associated with hyperglobulinemia or IgG levels, anti-nuclear antibody (ANA) positivity and fibrotic change with lymphocyte infiltration (Yoshida et al., 1995). Since then, many AIP cases have been reported in Japan and other countries, and some researchers have paid attention to the AIP. However, its definite pathogenesis

remains unclear. Therefore, AIP was once named as sclerosing pancreatitis, primary inflammatory pancreatitis, lymphoplasmacytic sclerosing pancreatitis, autoimmune pancreatitis, chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct, and sclerosing pancreaticocholangitis et al. In 2001, Hamano et al. obtained serum samples from 20 patients with sclerosing pancreatitis, and the corresponding contrast group patients, and the result showed IgG4 was significantly increased in AIP patients; but the concentrations of IgG1, IgG2, IgG3 or IgA, IgM, or IgE had not significantly difference between autoimmune pancreatitis and normal subjects (Hamano et al., 2001). With the more cases of AIP were observed, the Japan Pancreas Society firstly proposed the diagnostic criteria of AIP in 2002 according to the serologic findings and imaging (Members of the Criteria Committee for Autoimmune Pancreatitis, 2002). Since then AIP diagnostic criteria have undergone several changes. In 2006, Mayo Clinic College of Medicine summarized the criteria as HISORt (Table 1) based on the five cardinal features of AIP in histology, imaging, serology, other organ involvement, and response to steroid therapy. The third Japan–Korea symposium on AIP in 2008 obtained the consensus and set the final Asian criteria (Table 2). Currently, the criteria summarized as HISTOR and the final Asian criteria are the main criteria for the diagnosis of AIP (Chari, 2007; Divatia et al., 2012). At the AIP Summit 2008, evaluation of patients and the pathology of autoimmune pancreatitis suggest that AIP should be classified into two types. Type 1, the pathological synonym of which is lymphoplasmacytic sclerosing pancreatitis (LPSP) characterized by predominantly lobular involvement with “storiform” fibrosis, obliterative phlebitis, and infiltration by many IgG4+ plasma cells, typically seen in older males; Type 2, which is identified based on the histological features of neutrophilic infiltration into the pancreatic duct epithelium (granulocytic epithelial lesion: GEL). The differences between Type 1 and Type 2 autoimmune pancreatitis are listed in Table 3 (Zen et al., 2011). Type 1, which is very similar to pancreatic cancer, is a pancreatic manifestation of igG4 related disease. While Type 2 represents the main pattern in western series, and no association with extrapancreatic involvement has been described except for inflammatory bowel disease (IBD) in 20 %–30 % of the cases (Rebours et al., 2012). In 2010, Moon and coauthors proposed that expression of IgG4 in duodenal papillary biopsy specimens may be useful for the diagnosis of AIP. In 2012, Rebours et al. reported IgG4-positive plasma cells from the major papilla or pancreatic tissue are useful for the diagnosis of AIP. While, in other digestive tissues such as gastric mucosa and colonic mucosa IgG4-positive cells were not specific for AIP diagnosis, especially in patients with IBD (Moon et al., 2010).

IgG4-related sclerosing cholangitis

IgG4-related disease includes IgG4-related sclerosing cholangitis (IRSC or IgG4-SC), which involves the biliary duct and gallbladder. In 1963, Bartholomew et al. reported two cases of primary sclerosing cholangitis (PSC) with pancreatic involvement in the New England Journal of Medicine. These might have been the first two cases of IRSC reported (Bartholomew et al., 1963). Since then, it was always named by IgG4-related disease or autoimmune pancreatitis in literature. The pathogenesis of the IgG4-related sclerosing cholangitis was still unknown, and therefore it was once named by inflammatory pseudotumor from sclerosing cholangitis (Jafri et al., 1983), primary sclerosing cholangitis mimicking chronic pancreatitis (Laszik et al., 1988), pancreatic pseudotumor with multifocal idiopathic fibrosclerosis (Clark et al., 1988), lymphoplasmacytic sclerosing pancreatitis with cholangitis (Kawaguchi et al., 1991), sclerosing pancreato-cholangitis (Erkelens et al., 1999), atypical primary sclerosing cholangitis associated with unusual pancreatitis (Nakazawa et al., 2001),

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lymphoplasmacytic sclerosing cholangitis without pancreatitis (Nakazawa et al., 2004), immunoglobulin G4-related lymphoplasmacytic sclerosing cholangitis, autoimmune pancreatitis-associated sclerosing cholangitis. And recently it was named IgG4-associated cholangitis (IAC) (Björnsson et al. 2007). Since 2009, IAC has received more attention, owning to its steroid-responsive nature, which is similar to IgG4-related disease (Deshpande et al., 2009). The followings will discuss the new findings of IAC in recent years. IAC is characterized by obstructive jaundice, coexistence of AIP(74–96 %), lymphoplasmacytic infiltration and abundant IgG4-positive plasma cells, an immune reaction predominantly mediated by Th2 cells and Tregs, response well to steroid therapy or biliary drainage (Nishimori et al., 2009). Indeed, IAC shows various cholangiographic features which is similar to those of pancreatic cancer (PCa), PSC, and cholangiocarcinoma (CC) on cholangiography and clinical features. In 2006, Nakazawa et al. reported that IgG4-SC can be classified into four types based on the region of strictures revealed by cholangiography.These types include: Type 1, in which stenosis is located only in the lower part of the common bile duct; Type 2, in which stenosis is diffusely distributed throughout the intra- and extrahepatic bile ducts; Type 3, in which stenosis is detected in both the hilar hepatic region and the lower part of the common bile duct; and Type 4, in which strictures of the bile duct are detected only in the hilar hepatic region. Cholangiographic findings in Type 1 of our classification can often lead to a misdiagnosis of pancreatic carcinoma, those in Type 2 as PSC, and those in Types 3 and 4 as cholangiocarcinoma (Nakazawa et al., 2006). The comparison of IAC, PSC and CC is listed in Table 4. In 2011, Nakazawa et al. made diagnostic procedures for IgG4-related sclerosing cholangitis (Nakazawa et al., 2011). In 2012, Nakazawa et al. established diagnostic criteria for IgG4-SC based on cholangiographic classification (sensitivity 100 %, specificity 96.3 %) (Nakazawa et al., 2012). Diffuse or segmental stenosis of bile duct with wall thickening, together with one of criteria 1, 2, or 3.

Table 1: Mayo Clinic diagnostic criteria for autoimmune pancreatitis (IgG4-related sclerosing pancreatitis): The HISORt criteria

Criterion H-Histology (at least one of the following)	1. Periductal lymphoplasmacytic infiltrate, obliterative phlebitis, storiform fibrosis 2. Lymphoplasmacytic infiltrate, storiform fibrosis, abundant IgG4+ cells (≥10 HPF)
Criterion I-Imaging of pancreas	1. Typical-diffusely enlarged gland with delayed (rim) enhancement; diffusely irregular, attenuated main pancreatic duct 2. Others-Focal pancreatic mass / enlargement; focal pancreatic duct stricture; pancreatic atrophy; pancreatic calcification; pancreatitis
Criterion S-Serology	Elevated serum IgG4 (normal: 8-140 mg/dl)
Criterion O - Other organ involvement (can be confirmed by biopsy or resolution/ improvement with steroid therapy)	Hilar/intrahepatic biliary strictures; persistent distal biliary stricture; parotid/lacrimal gland involvement; mediastinal lymphadenopathy; retroperitoneal fibrosis
Criterion R-Response to steroid therapy	Resolution or marked improvement of pancreatic/extrapancreatic manifestation with steroid therapy
Diagnostic of autoimmune pancreatitis when any of the following is fulfilled	1. Criterion H 2. Criterion I+S 3. Strong clinical suspicion of autoimmune pancreatitis (idiopathic pancreatic disease + Criterion S and/or O) + Criterion R

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Table 2: Asian diagnostic criteria (Japan-Korea Consensus) for autoimmune pancreatitis

Criterion I - Imaging (both required)	1. Pancreatic parenchyma-diffuse / segmental / focal enlargement of the gland, occasionally with a mass and/or hypoattenuation rim; 2.Pancreaticobiliary ducts-diffuse / segmental / focal pancreatic duct narrowing, often with stenosis of bile duct
Criterion II - Serology (1 required)	1. High levels of serum IgG or IgG4 2. Detection of autoantibodies
Criterion III - Histopathology of pancreatic biopsy	Lymphoplasmacytic infiltration with fibrosis and abundant IgG4+ cells
Criterion IV - Histopathology of resected pancreas	Lymphoplasmacytic sclerosing pancreatitis storiform fibrosis, lymphoplasmacytic infiltration, periductal inflammation, obliterative phlebitis, numerous IgG4+ cells)
Optional criterion - Response to steroid therapy	Diagnostic trial of steroid therapy should be conducted only in patients fulfilling criterion I alone with negative work-up results for pancreatobiliary cancer
Diagnostic of autoimmune pancreatitis when any of the following is fulfilled:	1. Criterion I+II 2. Criterion I+III 3. Criterion I+II+III 4. Criterion IV

Table 3: Comparison of type1 and type2 autoimmune pancreatitis (AIP)

	Type 1 AIP	Type 2 AIP
Age	Adult	Child and adult
Gender predominantly	Male	Almost equal
Serum IgG4 levels	Elevated (usually >10/hpf)	Normal
Histological nomenclature	Lymphoplasmacytic sclerosing pancreatitis	Idiopathic duct-centric pancreatitis
Ducts	Periductal inflammation preserved epithelium	intraepithelial neutrophils damaged epithelium
Lobule	Lymphoplasmacytic inflammation with storiform fibrosis	Patchy inflammation, often with neutrophils
Veins	Phlebitis very common	Phlebitis unusual
IgG4+ plasma cells	Many	Rare
Granulocytic epithelial lesion	Absent	Present
Relapse rate	High	Low
Extra-pancreatic lesions	IgG4-related disease	Inflammatory bowel disease

Table 4: Discrimination between IAC, PSC and CC

	IAC	**PSC**	**CC**
Clinical features			
Age（years）	62.7 ± 10.7	42.4 ± 19.5	70.4 ± 11.7
Gender	male	no differences	no differences
Presenting complaint or abnormality	Obstructive jaundice	Liver test abnormalities	Obstructive jaundice
Association with AIP	74–96 %	less frequently	less frequently
Association with IBD	less frequently	60 %-80 %	less frequently
Serology			
IgG4 >135 mg/dl	83 %-100 %	5 %-9 %	22 %
IgG >1800 mg/dl	67 %-93 %	58 %	17 %
Tumor markers			
CA199 (U/ml)	101 ± 191	—	303 ± 524
CEA (ng/dl)	3.3 ± 2.9	—	12.9 ± 42.0
Clinical course	no progressive	progressive	progressive
Steroid therapy	responsive	resistant	resistant
Treatment	steroid therapy	liver transplantation	hepatectomy

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1. Association with autoimmune pancreatitis or other organ involvement

2. Group A (include patients with a Type 1 cholangiogram): Ruling out pancreatic cancer with high serum level of IgG4; Group B (include those with Type 2): Ruling out IBD with higher serum level of IgG4 or characteristic cholangiogram (discriminant analysis) or liver biopsy; Group C (include those with Type 3 and Type 4): Ruling out cholangiocarcinoma and characteristic IDUS (intraductal ultrasonography) findings [wall thickness (non-stricture) > 0.8 mm]

3. Evident fibrosis and prominent infiltration of lymphocytes and IgG4-positive plasma cells were observed in the bile duct of biopsy specimen (Nakazawa et al., 2012).

Retroperitoneal fibrosis

Retroperitoneal fibrosis (RF) is a chronic inflammatory fibrosis condition. Many factors including infection, radiation therapy, drugs, malignant tumors, trauma, surgery, and autoimmune lesions could result in retroperitoneal fibrosis (Stone, 2011). In this part we will discuss the autoimmune retroperitoneal fibrosis. In the last decade some cases of retroperitoneal fibrosis were intermittently reported in association with IgG4-related disease such as autoimmune pancreatitis, reticular lung lesion, periaortitis in thoracic aorta. IgG4-related retroperitoneal fibrosis often primarily involved the abdominal aorta, the kidney, the urethra or no specific lesions with a mass of involvement. The patients always have some atypical clinical pictures such as fever, low abdominal pain, backache, lumbar pain or edema in lower extremities. In 2009, Zen selected 17 cases (10 cases of IgG4-related and 7 cases of non-IgG4-related) of retroperitoneal fibrosis to perform clinic pathologic analysis, and discovered the following characteristics of IgG4-related retroperitoneal fibrosis. From the pathological specimens, the number of IgG or IgG4-positive plasma cells, and the ratios of IgG4/IgG-positive plasma cells in IgG4-related retroperitoneal fibrosis were significantly higher than that in Non-IgG4-related retroperitoneal fibrosis cases. From the serological examinations, the concentration of IgG4 and the IgG4/IgG ratio were significantly higher between the above two groups, but there were no significant differences in the IgG and antinuclear antibodies. IgG4-related retroperitoneal fibrosis always involves other organs or tissues, which includes pancreas, lung, sialaden and so on. Furthermore, the IgG4-related retroperitoneal fibrosis responds well to steroid therapy and the prognosis is good (Zen et al., 2009; van Bommel et al., 2009). However, there are no standard diagnostic criteria for retroperitoneal fibrosis until now. When we make a diagnosis, we should take the findings of CT scanning and magnetic resonance imaging, serological examination (essentially IgG4 should be greater than 135 mg/dl) and other manifestations of IgG4-related disease into consideration.

Other manifestations of IgG4-related disease

Sclerosing cholangitis, sclerosing cholecystitis, and retroperitoneal fibrosis are the peripancreatic inflammations in the abdomen. Besides these, there are other manifestations of IgG4-related disease. When the lesion involves the salivary gland, it can cause Mikulicz's disease (MD). MD is a part of IgG4-related disease, and the parotid gland involved either solely or along with the submandibular glands. MD was firstly reported by Johann von Mikulicz-Radecki in 1888. After that, MD was considered similar with SS (Sjogren's syndrome) for many years, indeed MD was different from SS. The main differences of salivary gland in MD are listed as following: elevated serum IgG4 concentrations and infiltration of plasmacytes expressing IgG4, the lack of anti-SS-A and anti-SS-B antibodies, good responsiveness to steroids, the persistent enlargement of lacrimal and salivary glands and not detectable or slight secretory dysfunction (Yamamoto et al., 2006). The IgG4-related interstitial lung disease can

present as interstitial lung disease (ILD), pulmonary inflammatory pseudotumor, and lymphomatoid granulomatosis (LYG). Zen et al. reported a case of IgG4-related pulmonary inflammatory pseudotumor (Zen et al., 2005). Shigemitsu reported a case of IgG4-related ILD and reviewed related literature in 2009 (Shigemitsu et al., 2009). It more frequently affects males aged about 60 years, sometimes with or without other organs involvement, into the lung of which were characterized by many IgG4-positive plasma cells infiltration, dense lymphoplasmacytic infiltrates with fibrosis, eosinophilic infiltration, and irregular narrowing of bronchioles. The chest X-ray and the chest CT may show diffuse ground-glass opacity, interlobular thickening, and bronchial wall thickening with hilar and mediastinal lymph-node swelling. And it also responds well to steroids (Shigemitsu et al., 2009; Zen et al., 2005).

CONCLUSION

IgG4-related disease has been known for many years. However, its definite pathological mechanism is still unclear. Currently, IgG4-related disease has been recognized as an immunological process, but there was no specific antibody could be one marker for it, especially in pediatric cases. With the more cases of IgG4 related diseases have been identified in adult or pediatric patients (Mannion and Cron, 2011), we believe people could discover the real entity of IgG4-related disease in the future.

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