Korea-Japan Symposium for Autoimmune Pancreatitis

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Korean panel: Jae Bock Chung, Seon Joo Kim, Myung-Hwan Kim, Jae Hoon Lim, Se Jin Jang, Jikon Ryu and Kyu Taek Lee

Japanese panel: Terumi Kimisawa, Tetsuhide Ito, Isao Nishimori, Kazuichi Okazaki, Shigeyuki Kawa, Masaru Koizumi, Terumi Kamisawa and Makoto Otsuki

Date: August 31, 2007

Venue: Yonsei University College of Medicine, Auditorium

The Host Organization: Korean Society of Pancreaticobiliary Disease

Korea-Japan symposium for autoimmune pancreatitis was held in Yonsei medical college, Seoul on August 31, 2007. Many pancreatologists from Korea and Japan attended to discuss about the latest findings regarding autoimmune pancreatitis (AIP). The session was divided into two parts: various features of AIP including imaging, serology, histology, other organ involvement and steroid responsiveness, and the currently used Japanese and Korean criteria with prospects of what should be included for an ideal diagnostic criteria. AIP is a relatively recently recognized disease entity that is receiving much attention by gastroenterologists because of its close clinical resemblance to pancreatic cancer and its good response to steroid therapy.

Both Japanese and Korean diagnostic criteria are similar in that characteristic imaging features of AIP-diffusely enlarged pancreas with diffuse narrowing of the main pancreatic duct- are essential for diagnosis. In recognition of more localized form of AIP, revised Japanese criteria of 2006 included localized enlargement and segmental narrowing of the duct as well. To go a step further, revised Korean criteria, which have been proposed in this symposium, also included mass forming type in the imaging criteria. Both criteria require addition of either positive serologic markers or histopathology consistent with AIP in addition to imaging for diagnosis. Serologic markers for AIP include serum IgG4, IgG and autoantibodies. Serum IgG4, although more sensitive, is more costly, takes longer for results and is not as widely available as

serum IgG. Currently, both criteria include both serum IgG and IgG4 for serologic diagnosis. Professor Kawa from Shinshu University gave lecture on serology and presented that ANA was positive in 60% of AIP patients and rheumatoid factor (RF) was positive in about 25%. Later, Professor Okazaki from Kansai Medical University presented data that positive result was obtained in 90% of patients using more specific autoantibodies combination of anti-lactoferrin and anti-carbonic anhydrase antibodies.

As for histopathologic diagnosis, Professor Se Jin Jang from Asan Medical center gave us a brief review. As widely recognized, periductal lymphoplasmacytic infiltration and fibrosis are histopathologic characteristics of AIP. In addition, vasculitis preferentially involving vein, which is termed obliterative phlebitis, is frequently observed. Another histological characteristic emphasized in the lecture was that inflammation was at similar stage for all involved sites unlike that of ordinary chronic pancreatitis, which shows some active and some inactive stage such as fibrosis and calcification differing by the region. This he named temporal and spatial homogeneity and considered important in differentiating from other causes. Also mentioned was finding of predominantly intralobular fibrosis with intact reticulin framework, and use of tissue IgG4 immunohistochemical stain for detection of IgG4 plasma cell infiltration in AIP diagnosis.

Recently, disease concept of AIP has been changing. What used to be considered associated autoimmune diseases with AIP in now considered as other organ involve-

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ment with similar pathogenesis. This concept of AIP as systemic fibroinflammatory disease was first proposed by Professor Kamisawa, who also gave lecture later in the symposium; infiltration of IgG4 positive plasma cells were not only found in pancreas, but also in other organs such as bile duct, gallbladder, stomach, salivary gland, lymph node and even bone marrow. Thus, the associated organ involvement such as salivary gland enlargement, retroperitoneal fibrosis, intra/extrahepatic bile duct strictures, and hilar lymphadenopathy are now considered to be of similar pathogenesis as pancreatic lesion, and they all respond well to steroid therapy. Professor Ito from Kyushu Univ presented a lecture on the associated other organ involvement. By studying 191 AIP patients from 7 institutions in Japan, the order of frequently involved extrapancreatic sites were sclerosing cholangitis (50%), hilar lymphadenopathy (30%), and salivary gland involvement (20%).

Professor Nishimori from Kochi Medical School presented a lecture on steroid response. Steroid responsiveness was studied by two nationwide surveys in Japan. From the data, initial steroid dose was most commonly 30-40 mg (0.6 mg/kg). Recurrence rate after steroid withdrawal was shown to be nearly 50%, and this recurrence rate was reduced to 24% by maintenance steroid dose of 5-7.5 mg continued for up to 3 years, because the recurrence of AIP usually occurs within 3 years.

In the second session, lectures centered on the diagnostic criteria of AIP and what should be included in the ideal criteria. Professor Ji Kon Ryu from Seoul National

University gave an overview of diagnostic criteria of AIP starting from historical background to the latest criteria that have been published. Professor Kyu Taek Lee from Samsung Medical Center compared the diagnostic criteria of Japanese, Korean, and one from Mayo clinic of USA. Lastly, Professor Myung-Hwan Kim from Asan Medical Center and Professor Kamisawa from Tokyo Metropolitan Komakome Hospital gave lecture on the ideal criteria and gave prospects for new Korean Criteria and Asian criteria for AIP. At the end of the second session, the most debated topic among Japanese and Korean pancreatologists was whether steroid response should be included for diagnosis of AIP. Steroid response is included in the Korean criteria, but not included in the Japanese diagnostic criteria. Japanese doctors were cautious in including it because inflammatory change associated with pancreatic cancer, the main differential diagnosis of AIP, may also show improvement with steroid trial, and facile usage of steroid trial without proper work up to rule out pancreas cancer may lead to misdiagnosis. Professor Myung-Hwan Kim remarked that although inflammatory pseudotumor associated with pancreatic cancer may improve, reversal of ductal narrowing seen in AIP is its unique feature. Japanese doctors also acknowledged that steroid responsiveness is a useful characteristic for AIP diagnosis for some cases that do not show either positive serology or histopathology.

Above has been a brief summary of some of the materials from the Korea-Japan Symposium for autoimmune pancreatitis.