## **IgG4-Related Disease**

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IgG4-related disease (IgG4-RD) is a novel clinical disease entity characterized by an elevated serum IgG4 concentration and tumefaction or tissue infiltration by IgG4-positive plasma cells. IgG4-RD encompasses a wide variety of diseases, formerly diagnosed as Mikulicz's disease (MD), autoimmune pancreatitis (AIP), hypophysitis, Riedel thyroiditis, tubulointerstitial nephritis, prostatitis, retroperitoneal fibrosis, inflammatory aortic aneurysm and inflammatory pseudotumor. However, like a crow flying on a dark night, IgG4-RD passed through the history of medicine, until the concept of an overarching of these systemic disorders was established in Japan at the beginning of the present century. In addition, most clinical practitioners are not yet familiar with IgG4-RD, and thus one cannot diagnose what one does not know.

The purpose of the book [1] is to raise awareness of this disease and its diagnostic pitfalls. Another goal was to delineate the characteristic features of individual organs in IgG4-RD radiologically and histopathologically with many color photographs. Most of the authors are members of the IgG4 team of the Japanese the Ministry of Health, Labor and Welfare (MHLW), and experts who have published numerous important papers in the field. Thus, this book will be useful to physicians in various disciplines such as gastroenterology, rheumatology, ophthalmology, otolaryngology, urology, hematology, respiratory medicine and oral medicine, not only as a textbook but also as an authoritative and comprehensive reference work.

The book consists of four parts with 33 chapters. Part I includes an overview, history of type I autoimmune pancreatitis and Milulicz's disease, comprehensive diagnostic criteria for IgG4-RD and therapies of IgG4-RD such as steroid and rituximab. Diagnosis of IgG4-RD is the most important. As all clinicians should become aware of this new disease entity, comprehensive diagnostic criteria (CD criteria) for IgG4-RD have been established in Japan for practical use and to differentiate among malignancies. The

CD criteria consist of only three parts: 1. organ involvement, 2. serum IgG4 $\geq$ 135mg/dL, 3. histopathological findings, and IgG4+ cells  $\geq$ 10/HPF and IgG4+ cells/IgG+ cells  $\geq$ 40%. Thus, the criteria are: definite (1. and 2. and 3.); probable (1. and 3.); and possible: (1. and 2.).

Part II covers characteristic radiological features of individual organs such as pancreas, bile duct, lacrimal glands and periocular lesions, salivary glands, lung, kidney and urinary tract, periarterial lesions, nervous system, etc. and images of scintigraphy and PET-CT with clear photographs.

Part III covers characteristic pathological features of each organ such as pancreas, bile duct, lacrimal and salivary glands, lung, kidney, retroperitoneal and arterial lesion, lymph nodes, skin and liver with vivid photographs.

Part IV provides descriptions of four actual informative cases: severe IgG4-related kidney disease, IgG4-RD with malignant tumor, IgG4-RD with multiple organ involvement and IgG4-RD with diabetes mellitus.

In conclusion, this is the first book on IgG4-related disease to appear in English and a concise but complete textbook in many aspects including clinical symptoms, radiological findings and characteristic pathological features. This book will be helpful for both specialists and practical physicians who seek a general outline and a more detailed examination of IgG4-RD.

## **Conflict of Interest**

Authors declare to have no conflict of interest.

## References

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<sup>1.</sup> Umehara H, Okazaki K, Stone JH, Kawa S, Kawano M. IgG4-Related Disease. 2014; Springer: Tokyo, Japan. [ISBN: 978-4-431-54227-8; 978-4-431-54228-5 (eBook)]