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Management of IgAN and IgAV

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IgA nephropathy (IgAN) and IgA vasculitis with nephritis (IgAVN), formerly known as Henoch-Schönlein purpura nephritis (HSPN) are the most frequent childhood glomerular diseases and both have wide range of clinical manifestations, pathological presentations, and long-term outcomes. IgA vasculitis is diagnosed on the basis of palpable purpura, abdominal pain, and arthritis/arthralgia. The proportion of IgA vasculitis patients with renal involvement ranges from 18-81%. And these two diseases are considered to be related diseases because both can be encountered consecutively in the same patient, have been described in twins, and bear identical pathological and biological abnormalities.

About 70% of pediatric IgAN patients are found with asymptomatic hematuria and/or proteinuria by an annual school screening program since 1974 in Japan while those are discovered with macroscopic hematuria in Europe and the United States. The prevalence of IgAN is estimated to be 40% of all native Japanese biopsies. Although initially it seemed to be asymptomatic and had a good clinical course, 30 to 40% of patients reached ESKD in 20 years without treatments. The Japanese Pediatric IgAN Treatment Study Group involving 20 Japanese pediatric renal centers conducted several clinical trials to establish optimal treatments for childhood IgAN since 1990. Based on the results of clinical trials, a 2-year combination therapy including prednisolone, mizoribine, and lisinopril for patients with severe childhood IgAN showing diffuse mesangial proliferation (WHO), and 2-year lisinopril for mild cases are recommended in the treatment guidelines 2020 for childhood IgAN by the Japanese society for pediatric nephrology. Though there are differences between the Japanese treatment guideline and the international guidelines such as KDIGO 2021 guideline, it is necessary to modify the treatment regimen according to the situation of each country, considering differences in race, medical examination, medical system, etc.

About IgAVN, the timing of kidney biopsy and the start of treatment remains controversial. No treatment strategy for IgAVN has been established in Japan. A nationwide survey has been currently conducted, and the Japanese IgAV guideline is being prepared.

As an example, we perform a kidney biopsy before any therapeutic intervention in children with IgAVN accompanied by acute nephritic and/or nephrotic syndrome, rapidly progressive glomerulonephritis (RPGN), or in children with persistent heavy proteinuria (urine protein-to-creatine ratio [UP/C] ≥ 1.0 g/gCr) for ≥ 1 month or mild proteinuria (UP/C ≥ 0.15, < 1.0 g/gCr) for ≥ 3 months. We define IgAVN cases with moderate severity as International Study of Kidney Disease in Children (ISKDC) classification grade II–IIIa and serum albumin (Alb) ≥ 2.5 g/dL. Moderately severe IgAVN cases receive RAS inhibitors as an initial treatment. Severe IgAVN cases are defined as ISKDC classification grade IIIb–V and/or serum Alb < 2.5 g/dL, and receive treatment with combination therapy (previously: prednisolone, mizoribine/azathioprine, warfarin, and dipyridamole; recently: prednisolone, mizoribine, and RAS inhibitors) similar to the treatment for the patients with severe IgAN, with good outcomes.

In this session, I would like to talk about the clinicopathological characteristics and present clinical situations of IgAN and IgAVN in Japan.