A Case of Podocytic Infolding Glomerulopathy with Primary Sjögren's Syndrome and Hashimoto's Thyroiditis

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To the Editor: Podocytic infolding glomerulopathy (PIG) was proposed as a new disease entity in 2008,[1] and great attention has been paid to PIG because of the rare pathological changes to glomeruli associated with it. PIG is characterized by specific changes to the thickened glomerular basement membrane (GBM), including microspheres, microtubular structures, and podocytic infolding. However, the clinical features and pathogenesis of PIG still remain unclear. To elucidate the characteristics of this glomerulopathy, accumulating more information from reported cases is necessary. Herein, we present a case of glomerulopathy showing podocytic infolding in association with primary Sjögren's syndrome (pSS) and Hashimoto's thyroiditis.

A 52-year-old woman, with bilateral leg edema, proteinuria, and a high antinuclear antibody level; positivity for anti-SSA and anti-SSB antibodies; and who had undergone through examination of thyroid function and a biopsy for pathological analysis of the labial salivary gland, was diagnosed with pSS in association with Hashimoto's thyroiditis. Examination of a renal biopsy sample under light microscopy revealed glomeruli with an irregularly thickened GBM, bubble-like structures in the capillary walls, and few spike formations. Immunofluorescence examination was negative for immunoglobulins, light chains, and complement components. Electron microscopy showed an irregularly thickened GBM with numerous microspheres, suggesting PIG [Figure 1].

The causes and mechanisms underlying the development of PIG are unknown. However, with the help of pathological observations, the following possible causes were considered. First, immune abnormalities with hyperactivation of the complement pathway may play an important role in the pathogenesis of PIG. Fujigaki *et al*.[2] demonstrated that C5b-9 was positive along the entire epithelial side of the GBM and in some microstructures, and they suggested that podocyte and GBM injuries might be caused by attack of podocytes by C5b-9, which might contribute partly to podocytic infolding and intra-GBM microstructures. Moreover, most previously reported cases of nephrotic PIG associated with autoimmune disease achieved complete remission with corticosteroid therapy. These results provide support for the hypothesis of immune abnormalities, in which hyperactivation of the complement pathway will cause PIG in autoimmune diseases.

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Second, these lesions were a reaction to podocyte injury. Matsuo *et al*.[3] reported a case of podocytic infolding lesions correlated with focal segmental glomerulosclerosis secondary to vesicoureteral

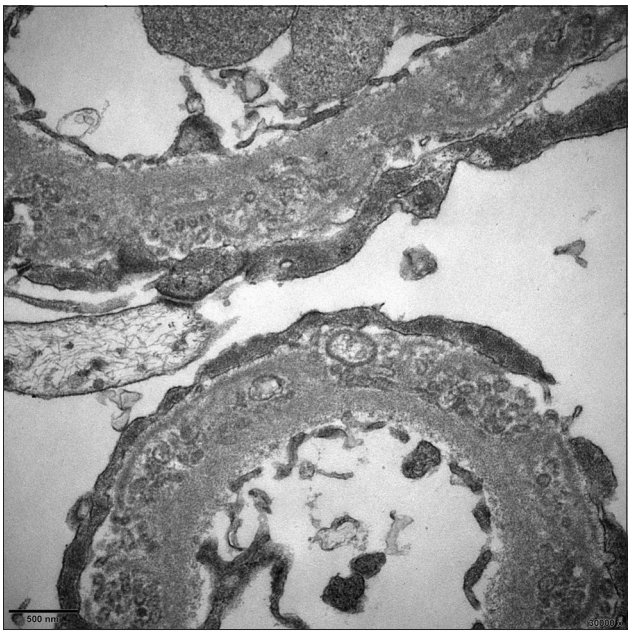

Figure 1: Glomerular findings of renal biopsy specimens. Electron microscopy showed extensive effacement of the epithelial foot process and irregularly thickened GBM (1200 nm) with numerous microspheres. These microspheres were encircled by a unit membrane. Most of them were located very close to the epithelial side of the GBM. There were no dense deposits suggesting immune complexes (original magnification ×30,000). GBM: Glomerular basement membrane.

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reflux (VUR). In this case, the second biopsy showed less podocytic infolding than the first, and proteinuria decreased with improving VUR. They speculated that podocyte damage because of hydronephrosis may lead to the development of the lesions. Podocyte injury, breaking the balance between biosynthesis and degradation of the matrix of the podocytes, may contribute to the development of PIG.

According to relevant references, PIG is heterogeneous both clinically and morphologically. Light microscopic characteristics of PIG may include different pathological types, immune deposits may be present or absent, and it may or may not be associated with collagen diseases. Patients with PIG always present with proteinuria and often have kidney dysfunction. For most patients with a previously reported PIG, the proteinuria decreased promptly with corticosteroid therapy. Thus, irrespective of whether PIG is a novel disease entity or a transient morphological finding of a well-known disease, increasing awareness of these lesions in view of how to promptly treat the disease will be beneficial. Therefore, further analysis of several cases is necessary to establish stricter diagnostic criteria and to formulate management strategies for the diagnosis in the future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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