

Summary

Introduction

Acute postinfectious glomerulonephritis (APGN) is a multifactorial immune-related kidneys' inflammation with initial damage of renal glomeruli with further involvement of all nephron structures into the pathological process. Most cases of APGN related to streptococcal etiology. There are two main streptococcal antigens responsible for development of APGN: nephritis strain-associated protein (NSAP) and nephritis-associated plasmin receptor, NAPIr. The pathogenesis of the APGN related to deposition of bacterial antigens with further in situ immune complexes formation and deposition of circulating immune complexes in renal glomeruli.

Polycystic kidney disease (PCKD) is heterogenous entity comprising various genetically determined lesions characterized by development of cysts in renal parenchyma. PCKD develop due to mutations of different genes encoding proteins of primary cilia, basal bodies, centrosomes that plays essential role in mechanoreception, Wnt-, Hedgehog signaling pathways and cell polarity.

Case presentation

Patient H., 20 years old, complained on swelling of face and lower limbs, fatigue, headache, hypertension after maxillary sinusitis. Despite the symptomatic therapy clinical symptoms, hypoproteinemia, azotemia, proteinuria progressively increased and 23.01.2015 kidney biopsy was performed. Conclusion of histopathological and immunohistochemical investigation: acute postinfectious glomerulonephritis with marked tubular and moderate interstitial components. Regarding rapid progression of disease, the pathogenetic therapy by glucocorticoids has been performed, but nephrotic syndrome with heavy protein loss were still developing. The disease progressed with further alignment of uremia, disseminated intravascular coagulation, vein thromboses and haemorrhages. Described homeostatic disorders caused development of fatal respiratory failure, hypotension and tachycardia which lead to patient's death.

Discussion

Acute postinfectious glomerulonephritis (APGN) is a multifactorial immune-related kidneys' inflammation with initial damage of renal glomeruli with further involvement of

all nephron structures into the pathological process. APGN characterized by abrupt onset of nephritic syndrome (NS) 1-3 weeks after bacterial infection. In most patients NS underlies complete remission. In some patients the symptoms of APGN persists for a long time.

APGN characterized by presence of glomerular hypercellularity, lobular appearance, subepithelial deposition of immune complexes, IgG, C3.

Various mechanisms triggers alternative complement pathway activation leading to destruction of glomerular basal membrane, increased basal membrane permeability, loss of plasma proteins. Some patients with persistent APGN possess congenital or acquired disruption of complement regulation leading to uncontrolled persistent alternative complement pathway activation.

In presented case histopathologic examination of kidney biopsy shows cystic dilation of cortical and medulary tubules of undefined type. This tubular abnormality has been asymptomatic during patient`s lifetime and complicated the clinical course and outcome of APGN.

The clinical course of APGN in presented case characterized by critical loss of plasma proteins, uremia leading to disseminated intravascular coagulation, vein thrombosis, multiple haemorrhages on skin and mucous membranes.

Unfortunately, APGN showed marked resistance to therapy by glucocorticoids which in some cases can be explained by rare genetically determined glucocorticoid resistance syndrome. On the other hand, inexorable decline of renal function can be related to renal tubules abnormality – polycystic kidney disease of undefined type.