HYPOCOMPLEMENTEMIC URTICARIAL VASCULITIS SYNDROME WITH KIDNEY INVOLVEMENT

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BACKGROUND

Hypocomplementemic urticarial vasculitis syndrome (HUVS) is a rare autoimmune hypocomplementemic small vessels vasculitis, affecting skin, joints, eyes, lungs and kidneys. It is mediated by anti-C1q antibodies and renal manifestations vary from microscopic hematuria to nephrotic syndrome and acute kidney injury. The objective of the study is to report a case of a patient with severe renal involvement due to HUVS and initial suspicion of systemic lupus erythematosus.

CASE REPORT

A 43-year-old woman was admitted to the hospital due to a generalized pitting edema, a new-onset arterial hypertension and dyspnea with progressive worsening. She had a history of refractory chronic urticaria, for more than one year, that was treated with omalizumab, an anti-IgE monoclonal antibody, but showed poor response. She also complained about arthralgias. At physical examination, she had important lower limbs and palpebral edema, ascites and urticarial rashes on her face and extremities. Laboratory tests showed increased serum creatinine (1.6 mg/dL) and nephrotic syndrome (proteinuria 5.5 g/day and serum albumin 2.8 g/dL). Urinalysis revealed microscopic hematuria and leukocyturia. The patient had inflammatory syndrome (C-reactive protein = 27.7 mg/L and erythrocyte sedimentation rate = 45 mm/h) and moderate, normocytic anemia (Hb: 9.4 g/dL). Immunological work-up—ANA, anti-ds-DNA antibodies, anti-Ro antibodies, anti-La antibodies, pANCA, cANCA, anti-GBM antibodies, rheumatoid factor and cryoglobulins—was negative. Complement fractions were low (C3 = 36 mg/dL; C4 = 4.4 mg/dL). Tests for HBV, HCV and HIV were negative. A kidney biopsy was performed, and the clinical and pathological features were consistent with an immune complex mediated membranoproliferative glomerulonephritis with positive C1q immunostaining and complement activation. In this context, the diagnosis of HUVS was established. Indeed, the patient fulfilled both major criteria, namely urticaria for more than 6 months and hypocomplementemia, and two of the six minor criteria which were arthralgias and renal involvement. Induction treatment with intravenous pulses of methylprednisolone and hydroxychloroquine was subsequently initiated. The patient evolved with quick improvement of the urticarial rash, arthralgias and full recovery of renal function in the course of time.

CONCLUSION

This case highlights the importance of HUVS as a cause of an immune-complex mediated glomerulonephritis and as a differential diagnosis with other autoimmune diseases such as systemic lupus erythematosus. Therefore, it is important for physicians to recognize this condition to start aggressive immunosuppressive treatment early to avoid a poorer prognosis.

KEYWORDS

Hypocomplementemic urticarial vasculitis, Glomerulonephritis, Anti-C1q antibodies.