CLINICAL MANIFESTATIONS AND THERAPEUTIC CHALLENGES OF HYPOCOMPLEMENTEMIC URTICARIAL VASCULITIS SYNDROME

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Hypocomplementemic urticarial vasculitis syndrome (HUVS) is a distinct type of urticarial vasculitis with multiorgan involvement, whose etiology and link with SLE and other immune diseases are still unknown. The major manifestations of HUVS are chronic urticarial vasculitic lesions, angioedema, laryngeal edema, ocular inflammation, arthritis, arthralgia, obstructive lung disease, recurrent abdominal pain and glomerulonephritis. We present 4 patients who showed the typical signs of hypocomplementemic urticarial vasculitis syndrome, including urticarial lesions persisting more than 24 hours, recurrent angioedema, ocular inflammation, arthritis and arthralgia. Two of them had progressive obstructive lung disease, the third patient had glomerulonephritis, and the fourth patient did not show any further systemic involvement. In the autoimmune laboratory results we found hypocomplementemia and rheuma factor positivity in all cases, in one case ANA positivity with SS-A, SS-B positivity, in one case borderline ds-DNA positivity, in other one dsDNA and ANA borderline positivity, while the last patient had only a mild ENA positivity. The skin histological findings were leukocytoclastic urticarial vasculitis in all cases with immune complex deposits contains IgG, IgM and c3, and in 2 cases we stained for c1q and found strong positivity at the basal membrane. The therapeutic outcome with usual immunosuppressive treatment was insufficient in every case, skin flares and angioedema recurred irrespectively of therapy. HUVS is considered by some to be a SLE-associated immunological disease, whereas many others consider it a distinct disease entity. Among our patients, 3 fulfilled the diagnostic criteria for SLE, but 1 patient did not. The fact that not all HUVS patients have SLE, and only a minor fraction of SLE patients develop HUVS, indicates a distinct pathomechanism for developing HUVS.