8 Urticarial Vasculitis Syndrome

8.1 General Aspects

The usual clinical presentation of necrotizing vasculitis is a palpable purpura; other possible features are macules, papules, pustules, ulcers, or scars. A few decades ago, attention was called to the fact that urticaria can also be a manifestation of cutaneous vasculitis (McCoombs 1965). The clinical aspects and underlying pathomechanisms of urticarial vasculitis were studied two decades later (McDuffie et al. 1973, Sissons et al. 1974, Soter et al. 1974, Agnello et al. 1975 and 1976), and by the early eighties, more than 100 patients with the disease had been reported in the literature (Sanchez et al. 1982, Wanderer et al. 1983).

By then, it was apparent that vasculitis can be an accidental finding in chronic urticaria, but that such changes can also be associated with severe disease such as systemic lupus erythematosus (SLE), serum hepatitis, or malignant neoplasms. It is thus appropriate to speak of urticarial vasculitis as a syndrome that encompasses a broad spectrum of systemic symptoms and laboratory changes, with the common features being certain histological alterations and a relative refractoriness to treatment.

Synonyms that have been in use for the disease are vasculitis with urticaria (Feig et al. 1976), hypocomplementemic urticaria (McDuffie et al. 1973), and “unusual SLE-related syndrome” (Agnello et al. 1975).

8.2 Epidemiology

Urticarial vasculitis affects preferably young to middle aged women (range 6–68 years). The incidence is relatively low, but varies widely, depending on how strictly the disease is defined. Jones et al. (1983) diagnosed hypocomplementemtic urticaria (McDuffie et al. 1973), and “unusual SLE-related syndrome” (Agnello et al. 1975).

leukocytoclastic vasculitis in only one of 43 patients with chronic urticaria, while four had decreased serum complement levels and three had circulating immune complexes. Synkowski et al. (1979) found no histologic evidence of vasculitis at all in 14 randomly selected patients with acute and chronic urticaria. Among 231 patients with chronic urticaria and arthritis, only eight were found to also have vasculitis in urticarial skin lesions (Small et al. 1982).

The incidence of urticaria among patients with proven necrotizing vasculitis is also relatively low. Phanuphak et al. (1980) observed wheals in 12 of 79 patients, Ekenstam and Callen (1984) among 17 of 82 patients in a private practice, and Fauci (1983) reported an incidence of 9%. These authors emphasize that urticaria is typically associated with chronic and not with acute vasculitis.

Urticaria is observed in 0–50% of the patients with systemic lupus erythematosus in different series. Tuffanelli and Dubois (1964) observed it in 7% of their 520 patients and O'Loughlin et al. (1978) more recently in 22% of 54 patients. Urticaria is never a feature of discoid (cutaneous) lupus erythematosus (Provost et al. 1980).

8.3 Clinical Aspects

8.3.1 Cutaneous Manifestations

Urticarial lesions in patients with vasculitis are generalized in their distribution and may even involve the palms and mucous membranes (Soter et al. 1974). Individual lesions are raised, indurated, faintly red (Fig. 8.1), or purple red with an elevated, red margin (Fig. 8.2). A fine, punctate purpura is often observed within the lesions. After healing, a residual slight hyperpigmentation, scaling, or purpura may remain at the site of previous lesions (Callen and Kalbfleisch 1982). Wheals generally persist for 24–72 h, and according to Provost et al. (1980), even up to 1 week. Symptoms can be entirely absent, although most patients experience severe pruritus or even burning at the site of lesions. Hyperesthesia on application of light pressure to the wheals has been noted in some patients (Provost et al. 1980).

Beside urticarial lesions, other types of skin lesions can coexist, such as bullous lesions, erythema multi-
forme-like eruptions, or livedo reticularis (Table 8.1). New crops of lesions can appear at daily or monthly intervals.

8.3.2 Systemic Manifestations

Next to the skin, the most frequently involved organs are the joints (Table 8.1). Arthralgias may present as pain, stiffness, or more rarely swelling, may be transient, and may or may not coincide with cutaneous lesions. Symptoms usually last for no more than 72 h; persistent arthritis is relatively seldom. Multiple joints may be affected, with knees, ankles, wrists, and fingers being most frequently involved.

Vascular lesions of the kidneys are often present in urticaria vasculitis, but are missed unless patients are checked for hematuria, proteinuria, or creatinine clearance. Less frequent symptoms and organ involvements are listed in Table 8.1. Mild fever, weight loss, fatigue, pleuritic chest pain, and other symptoms also known to occur in connective tissue diseases are even more rare (Falk 1984).

8.4 Histopathology

There is general agreement that urticarial vasculitis is characterized by the following features: edema of the upper dermis; a neutrophilic infiltrate in and around the walls of the small superficial venules, with scattered nuclear fragments (nuclear dust) and extracellular red blood cells in the dermis; and deposition of fibrinoid material in and around the vessel walls (Table 8.2).