VASCULITIS AND THE KIDNEY

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The term "vasculitis" denotes a heterogeneous group of diseases characterized by inflammation and necrosis of blood vessels and a variable clinical picture, according to the organs or tissues mainly involved by the vasculitic lesions.

Renal involvement, often characterized by progressive impairment of renal function, frequently occurs in these patients where it represents one of the most pronounced clinical features, and a frequent cause of death.

The prevalence of renal vasculitis is difficult to assess since diagnosis may not be obvious owing to the variable clinical picture and possibly unspecific clinical features. Moreover, typical vasculitic lesions may be absent in renal biopsy material where more frequently a focal segmental necrotizing glomerulonephritis is observed. This histological picture, particularly when scanty or no deposits are present at immunofluorescence, is considered strongly suggestive of vasculitis and deserves careful examination of the patients to identify signs confirming the diagnosis.

It has been suggested that most crescentic glomerulonephritis with negative immunofluorescence, which accounts for approximately 40% of extracapillary glomerulonephritis, may actually represent the renal consequences of systemic vasculitis. Among the renal biopsies carried out over 30 years in our Institute, vasculitis accounts for 7.4% of vascular nephropathies, but it is likely that the actual prevalence is higher, on account of the increased carefulness nowadays used in diagnosis.

CLASSIFICATION

No satisfactory classification exists of the various form of vasculitis because their etiology and pathogenesis are still largely unknown and there is a large overlap among the various diseases. It is clear that the ultimate classification will depend on identification of the various antigens responsible.
TABLE I. Classification of vasculitis

A) Systemic vasculitis

a) Non granulomatous vasculitis

1) Polyarteritis Nodosa
   - Classic Polyarteritis Nodosa
   - Microscopic Polyarteritis Nodosa
   - Overlap syndrome

2) Hypersensitivity Vasculitis

3) Associated with other systemic diseases
   - Schönlein-Henoch purpura
   - Systemic lupus erythematosus
   - Cryoglobulinemia
   - Rheumatoid arthritis
   - Relapsing polychondritis
   - Malignancies

b) Granulomatous vasculitis

1) Wegener's granulomatosis
2) Allergic granulomatosis (Churg-Strauss syndrome)
3) Lymphomatoid granulomatosis

c) Giant cell arteritis

1) Temporal arteritis
2) Takayasu's disease

B) Renal vasculitis

- Associated with renal transplant rejection

The table I reports a possible classification which includes some well-defined clinical pictures that can be considered discrete nosographic entities (such as Wegener's granulomatosis) and other diseases whose separation in distinct categories could be questionable.

The distinction between hypersensitivity vasculitis and the microscopic form of polyarteritis nodosa, for example, is not always easy and the two forms are hardly distinguishable from each other on a pathological basis, the former being now generally applied to cutaneous vasculitis where an allergic reaction to an antigen (most commonly drugs) is evident.

Likewise, the separation between granulomatous and non granulomatous vasculitis, on the assumption that histological features may reflect different pathogenetic mechanisms, could be a matter of controversy.

The following discussion will focus on the main features of vasculitis of the Polyarteritis Nodosa group, Wegener's granulomatosis, and vasculitis associated with renal graft rejection since they are among the most frequent forms of renal vasculitis and their course is often characterized by irreversible renal damage leading to end stage renal failure.