Preface

This volume seeks to provide a comprehensive overview of the systemic vasculitides, an extremely heterogeneous group of diseases characterized by inflammation and necrosis of different-sized blood vessels. With a few exceptions (e.g., HCV-related cryoglobulinemic vasculitis and HBV-positive polyarteritis nodosa), the etiology of these clinical conditions remains unknown. In spite of their relatively low prevalence, the systemic vasculitides have been the object of recent, intensive, basic, and clinical studies. For this reason, it can be safely stated that this group of diseases is one of the most rapidly progressing areas of clinical medicine, as evidenced by the dramatic achievements in terms of clinical remission and overall prognostic improvement.

The pathophysiology of the vasculitides is multifactorial and thus in most cases poorly defined. Among the many potential influences on disease expression, sex, ethnicity, and genetic as well as environmental factors are likely to play a role. In addition, the vascular damage characteristic of the systemic vasculitides may be the result of autoimmune responses, such as antineutrophil cytoplasmic autoantibodies, anti-endothelial cell autoantibodies, immune complex deposition, an immune response to foreign antigens or infectious agents, and T-lymphocyte responses with granuloma formation.

The general aims of this book are:

(i) To provide an in-depth update of the major pathogenetic, genetic, and clinical advances in the field encompassing the vasculitides, including, for each condition, a summary of the most cogent information scattered in the medical literature but not always readily retrievable
(ii) To describe not only conventional treatments, but also the more recently developed and tested drugs as well as efforts at patient-tailored therapies, especially for patients with refractory and relapsing disease
(iii) To point out future directions of research that, while challenging, are likely to be profitable in terms of improved diagnosis and therapy
If this book serves as a stimulating resource for basic and clinical researchers and specialists in related disciplines, as well as practicing physicians and advanced medical students interested in this fascinating branch of medicine, our efforts as editors will have been fully rewarded. The lion’s share of the merit should, however, be given to the international contributors who accepted our invitation to collaborate in this project and who, in doing so, were able to impart the knowledge gained during their multiyear experience in this field.

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