

CONTENTS

Preface xi
John H. Stone

Vasculitis: A Collection of Pearls and Myths 691
John H. Stone

Since Kussmaul and Maier described the index case of vasculitis in 1866, the field has seen many changes. What was considered for decades to be only a single disorder is now known to consist of at least 15 to 20 distinct conditions. Important strides have been made in unraveling the pathophysiology of some individual forms of vasculitis, but many mysteries remain. Over time, numerous myths and occasional pearls have arisen from the care of patients with these disorders. This collection of pearls and myths gathers lessons about the status of clinical care of vasculitis patients in the year 2008.

Targeting B Lymphocytes as Therapy for ANCA-Associated Vasculitis 741
Jason M. Golbin and Ulrich Specks

This article focuses on the initial results achieved with the more selective immunosuppressive approach of B-lymphocyte depletion in patients who fail cyclophosphamide or have contraindications for its use in the treatment of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV). This novel approach has sparked hope for patients and physicians in their search for effective, well-tolerated therapy for AAV. B-cell depletion is now undergoing rigorous investigation in randomized clinical trials.

Airway Involvement in Wegener's Granulomatosis

755

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Wegener's granulomatosis is characterized by necrotizing granulomatous inflammation and necrotizing vasculitis affecting predominantly small arteries, arterioles, capillaries, and venules. In contrast to the well-described pulmonary parenchymal involvement of Wegener's granulomatosis, the lower airway (tracheobronchial) disease manifestations are less well recognized by clinicians. Consequently, mild disease of the airways is easily missed. There is a relative paucity of published information on various tracheobronchial manifestations of Wegener's granulomatosis. This article provides a comprehensive review of the diagnosis and management of the infraglottic tracheobronchial disease manifestations.

Takayasu Arteritis: What is the Long-Term Prognosis?

777

Kathleen Maksimowicz-McKinnon and Gary S. Hoffman

Takayasu arteritis (TA) is a form of idiopathic large vessel vasculitis that predominantly affects women of reproductive age. Although TA is a rare disease, the interpretation of longitudinal data from several countries provides new insights into the clinical course and outcomes in TA across different racial and ethnic groups. Contemporary studies belie prior perceptions of TA as a disease with a self-limited, benign course. We now recognize this disease as one that often relapses, leaves the patient chronically dependent on glucocorticoids for disease control, and frequently leads to disability. Limited data suggest that the targeted inhibition of tumor necrosis factor (TNF) might be an effective therapy for TA.

Pyoderma Gangrenosum: An Update

787

Jeffrey P. Callen and J. Mark Jackson

Pyoderma gangrenosum (PG) can be differentiated into classic and atypical forms. The classic form is characterized by ulcers and the atypical form by deep erosions with bullous blue-gray margins. Pathergy, the development of cutaneous lesions at sites of trauma, is a common feature of both forms of PG. Approximately 50% of patients who have PG have underlying systemic diseases, most commonly inflammatory bowel disease, myeloproliferative disorders, and various forms of inflammatory arthritis. The diagnosis of PG is one of exclusion. The management of this disorder begins with treatment of any underlying disease and local or systemic glucocorticoids or immunomodulating therapies.

Retroperitoneal Fibrosis: Evolving Concepts

803

Augusto Vaglio, Alessandra Palmisano, Domenico Corradi,
Carlo Salvarani, and Carlo Buzio

Retroperitoneal fibrosis (RPF) is a rare fibro-inflammatory condition that is idiopathic in most cases, but may be secondary to various

causes. Although the cause and pathogenesis of the idiopathic form are unknown, immunogenetic factors and immunopathologic/autoimmune mechanisms are probably involved. Idiopathic RPF usually develops around the abdominal aorta and iliac arteries but in some cases may also involve the thoracic aorta and the origin of its major branches, with a pattern similar to that of other forms of large-vessel vasculitis. In addition, the disease is frequently associated with autoimmune conditions affecting other organs. Glucocorticoids alone or in combination with immunosuppressive agents are usually effective treatment options, but the disease frequently has a chronic relapsing course.

Five Clinical Conundrums in the Management of Giant Cell Arteritis

819

Maria C. Cid, Ana García-Martínez, Ester Lozano,
Georgina Espígol-Frigolé, and José Hernández-Rodríguez

Clinicians who treat patients with giant cell arteritis (GCA) face many unresolved challenges. Visual loss still occurs in 15% to 20% of patients despite the availability of therapy for the disease that is generally effective. Aneurysm formation and large vessel stenosis are increasingly recognized complications. Substantial iatrogenic morbidity stems from glucocorticoid therapy, and recent trials have failed to identify an efficient steroid sparing agent. In this review, the authors address five major clinical conundrums in the management of GCA.

Scleritis and Peripheral Ulcerative Keratitis

835

Anat Galor and Jennifer E. Thorne

Scleritis and peripheral ulcerative keratitis (PUK) can present as isolated conditions or as part of a systemic inflammatory or infectious disorder. Both are serious ocular conditions that can result in vision loss and require early diagnosis and treatment. Nearly two thirds of patients with non-infectious scleritis require systemic glucocorticoid therapy and one fourth need a glucocorticoid-sparing agent as well. Essentially all patients with non-infectious PUK require systemic glucocorticoids. A detailed clinical history, thorough physical examination, and thoughtful laboratory evaluations are important in the exclusion of underlying disorders and extraocular involvement.

Cogan's Syndrome: An Audiovestibular, Ocular, and Systemic Autoimmune Disease

855

Mehrdad Mazlumzadeh and Eric L. Matteson

Cogan's syndrome is an immune-mediated systemic disorder characterized by ocular and audiovestibular inflammation. Although interstitial keratitis in the absence of an infectious cause is the classic form of eye involvement, other nonspecific inflammatory eye

diseases can also occur, often making prompt diagnosis of the underlying disease challenging. The audiovestibular disease is difficult to treat and can lead to profound hearing loss. A poor outcome, especially complete hearing loss, can sometimes be prevented through a timely recognition and initiation of glucocorticoid therapy at the onset of the disease.

Index

875