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Primer on the Rheumatic Diseases

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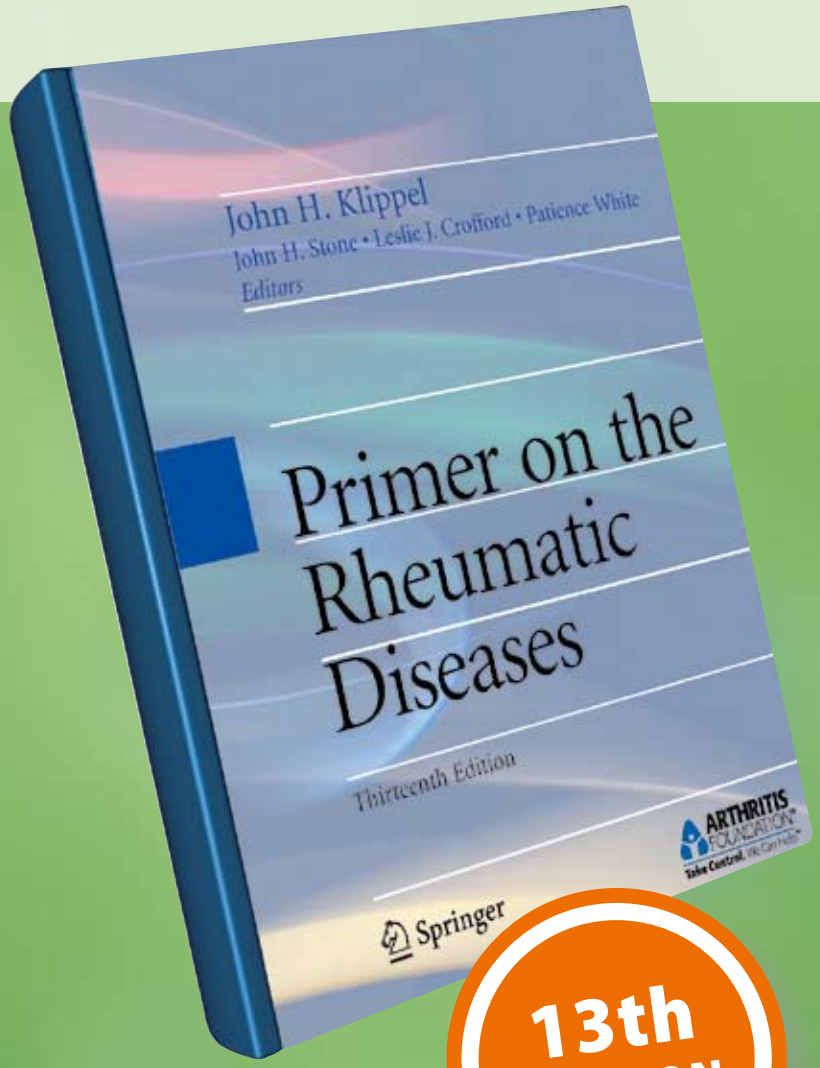

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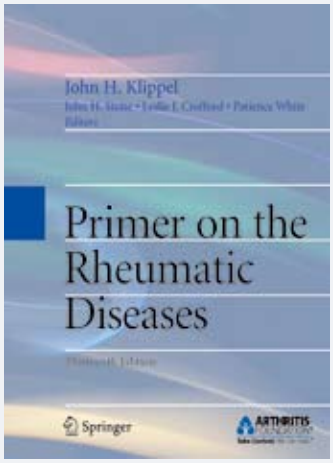

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Primer on the Rheumatic Diseases

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Primer on the Rheumatic Diseases is one of the most prestigious and comprehensive texts on arthritis and related diseases, including osteoarthritis, rheumatoid arthritis, osteoporosis, lupus and more than 100 others. It offers medical students and physicians a concise description of the current science, diagnosis, clinical consequences, and principles of management. New and expanded chapters heighten the translational nature of this edition. Students, trainees, and practicing clinicians all need a standard textbook that can change with the times and reflect recent strides taken in understanding and treating rheumatic disease. The Primer fills that need.

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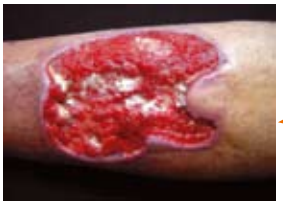

FIGURE 25E-3

Pyoderma gangrenosum in a patient without an associated disease.

Patients with PG also demonstrate pathergy. Thus, this condition has been reported following a variety of surgical procedures, for example, thoracotomy or fasciotomy. The systemic associations vary depending on the type of PG. Classical disease and peristomal PG are associated more frequently with inflammatory bowel disease and/or arthritis. Careful evaluation for inflammatory bowel disease is warranted in cases of peristomal PG, even when the stoma was created for other reasons (e.g., following cancer surgery). In contrast, atypical pyoderma gangrenosum is found more frequently in the setting of myelocytic leukemia or preleukemic conditions.

The diagnosis of PG is one of exclusion. Although biopsies should be performed to exclude other conditions, PG does not have a distinctive histopathology. Because of the importance of excluding disease mimickers—particularly infections—biopsy is almost always performed as part of the evaluation, despite the possibility that the ulcer will extend through pathergy. Culture of the lesions following skin biopsy is essential. Infectious mimickers are not common but include deep fungal infections; for example, blastomycosis, sporotrichosis, histoplasmosis, and coccidioidomycosis; as well as nocardiosis, tuberculosis, atypical mycobacteria; and herpes simplex virus. Following diagnosis, appropriate studies should be undertaken to exclude inflammatory bowel disease, rheumatoid arthritis (RA), systemic vasculitis, paraproteinemia, and other hematologic disorders. As with Sweet's syndrome, neutrophilic infiltration of organs other than the skin may sometimes occur in PG.

For cases of PG associated with an underlying disease (e.g., inflammatory bowel disease or RA), treatment of the primary condition often leads to improvement in PG. Prednisone (1 mg/kg/day) is generally the first line of therapy for idiopathic PG. Infliximab (3–5 mg/kg every 6 weeks following two initial doses 2 weeks apart) is also an effective therapy for PG, even in the absence of inflammatory bowel disease. Other therapies employed in PG include dapsone [100–200 mg/day (assuming normal levels of glucose-6-phosphate dehydrogenase; G6-PD)], thalidomide (100 mg/day), cyclosporine (3 mg/kg/day), azathioprine [2 mg/kg/day, assuming normal levels of thiopurine methyltransferase; (TPMT)], and mycophenolate mofetil (1.0– .5 g b.i.d.).

Neutrophilic dermatosis of the dorsal hands (NDDH; 4), considered by some to be a separate disease entity, is regarded more commonly as a variant of either Sweet's syndrome and atypical PG. NDDH is associated with the same underlying conditions as Sweet's syndrome and atypical PG, and the management considerations are identical.

Rheumatoid neutrophilic dermatosis, an unusual complication of RA, is characterized by symmetrical erythematous papules and plaques on the dorsal hands, elbows, and extensor surfaces of the forearms (5). Patients generally have active and often severe RA, but the condition has been reported in at least two patients with seronegative RA. In terms of histopathology, rheumatoid neutrophilic dermatosis resembles Sweet's syndrome. Treatments that have been suggested include glucocorticoids, dapsone (100–200 mg/day), and colchicine (0.6 mg b.i.d.); however, spontaneous resolution has been reported to occur.

Bowel-associated dermatosis–arthritis syndrome was first recognized in the 1970s following gastric bypass surgery for morbid obesity. Fortunately, because of major alterations in surgical technique, this syndrome is

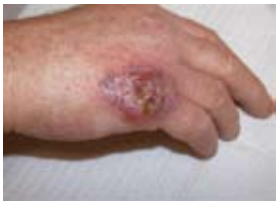

FIGURE 25E-4

Atypical pyoderma gangrenosum, also known as neutrophilic dermatosis of the dorsal hands.

Expanded chapter on the cutaneous manifestations of disease

IATIONS WITH NEUTROPHILIC DERMATOSES.

SWEET'S SYNDROME	PYODERMA GANGRENOSUM (PG)	RHEUMATOID NEUTROPHILIC DERMATOSIS	BOWEL-ASSOCIATED DERMATOSIS ARTHRITIS SYNDROME	NEUTROPHILIC DERMATOSIS OF THE DORSAL HANDS
Some	20%–25%	No	Yes	Occasional
Occasional	10% for superficial forms, less for classical PG	Yes, occasionally seronegative	No, but joint disease may simulate RA	Occasional
25%–30%	15% for the superficial forms	No	No	15%
Rare	Rare	No	No	Rare
Possible	No	Possible	No	Possible
Occasionally	No	No	No	Possible
Occasionally	No	No	No	No

-monocyte colony-stimulating
ulocyte colony stimulating factor
imatinib, minocycline, hydra-
aceptives. For acute disease, a
ednisone tapered over 2 weeks is
current disease without an asso-
corticoid-sparing agents such as
immunosuppressive agents, and tumor necrosis factor alpha (TNF-alpha) antagonists are often used. In patients with acute, idiopathic disease, the prognosis is generally good; many have only episode. However, the course of patients with underlying leukemia or myelodysplasia follows that of the associated disease. Absent disease remission or a cure, recurrences are common.

Pyoderma gangrenosum (PG) is a form of ulcerative skin disease. There are at least four clinical variants of PG: classical, atypical, peristomal, and mucosal (3). The classical lesion is a rapidly progressing, painful ulcer, most often on the leg, with a violaceous, undermined (overhanging) border (Figure 25E-3). Atypical PG occurs as a more superficial lesion, often on the dorsal hands (Figure 25E-4), extensor forearms, or face. The border of atypical PG may appear bullous, leading to clinical confusion with Sweet's syndrome. Peristomal PG occurs as a deep ulcer near the site of a stoma, usually created after gastrointestinal or genitourinary surgery. Finally, mucosal PG is associated with ulcerations that can resemble simple aphthae or vegetative lesions. Mucosal PG must be differentiated from Behçet's disease.

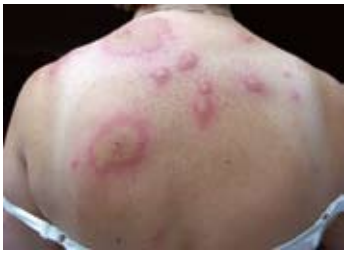

FIGURE 25E-1

Sweet's syndrome.

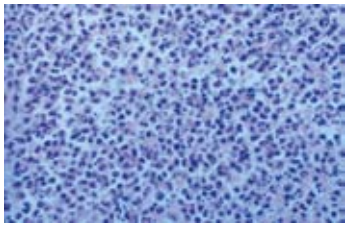

FIGURE 25E-2

Histopathological findings in Sweet's syndrome.

Color figures depict cutaneous findings and histopathology

TABLE 21C-3. CLINICAL FEATURES OF THE PRIMAR
VASCULITIDES.

FEATURE	WEGENER'S GRANULOMATOSIS
ANCA positivity	80%–90%
ANCA antigen specificity	PR3 > MPO
Fundamental histology	Leukocytoclastic vasculitis; necrot granulomatous inflammation (r in renal biopsy specimens)
Ear/nose/throat	Nasal septal perforation; saddle-n deformity; conductive or senso hearing loss; subglottic stenosis
Eye	Orbital pseudotumor, scleritis (risk scleromalacia perforans), episcl uveitis
Lung	Nodules, infiltrates, or cavitary les alveolar hemorrhage
Kidney	Segmental necrotizing glomerulo rare granulomatous features
Heart	Occasional valvular lesions
Peripheral nerve	Vasculitic neuropathy (10%)
Eosinophilia	Mild eosinophilia occasionally None All

SOURCE: Reproduced with permission from Seo P, Stone JH. The antineutrophil cytoplasmic antibody-associated vasculitides. Am J Med 2004;117:39–50.
ABBREVIATIONS: ANCA, antineutrophil cytoplasmic antibody; MPO, myeloperoxidase; PR3, proteinase 3.

CLINICAL FEATURES

There is substantial overlap in many of the clinical features of the AAVs. In some cases, distinguishing among two or more of these diseases on the basis of clinical features alone is difficult (Table 21C-3).

Upper Respiratory Tract and Ears

Although patients with the CSS or MPA may experience substantial ear, nose, or sinus disease, this pattern of involvement is most characteristic of WG. More than 90% of patients with WG eventually develop upper airway or ear abnormalities. The nasal symptoms of WG include nasal pain and stuffiness, rhinitis, epistaxis, and brown or bloody crusts. Nasal inflammation may lead to septal erosions, septal perforation, or, in many cases, nasal bridge collapse—the "saddle-nose deformity" (Figure 21C-1). The distinction between active WG in the sinuses and secondary infections in the sinuses may be challenging (see Nonmedical Interventions section).

In 60% to 70% of patients with the CSS, allergic rhinitis is the earliest disease manifestation, typically appearing years before the development of full-blown

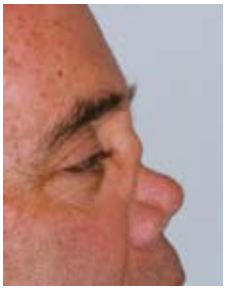

FIGURE 21C-1

Saddle-nose deformity in Wegener's granulomatosis.

Details musculoskeletal signs and symptoms

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From the Foreword

The 13th edition of the **Primer on the Rheumatic Diseases** *is an extraordinary handbook for clinical care. The Primer will educate trainees, update established clinicians, and help health care providers from all walks of the profession provide better care for patients with arthritis and rheumatic diseases. I congratulate the editors on their superb work. In addition, the multiple contributors — many of whom are members of the American College of Rheumatology — should be thanked for their scholarly contributions to the Primer.* ► **Michael E. Weinblatt**, MD, Professor of Medicine, Harvard Medical School, Brigham and Women's Hospital, Boston, MA, USA

About the Editors

John H. Klippel, M.D. is the President and Chief Executive Officer of the Arthritis Foundation. He previously served as a Senior Investigator in the Arthritis and Rheumatism Branch (NIH) (1976-1987), Clinical Director of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) (1987-1999), and Medical Director of the Arthritis Foundation (1999-2003). He is a diplomat of the American Board of Internal Medicine and a fellow of the American College of Physicians and the American College of Rheumatology. His honors and awards include the Surgeon General's Exemplary Service Award, Distinguished Clinical Teacher Award (NIH Clinical Center), Directors Award (NIH Clinical Center) and the Burroughs-Wellcome Visiting Professor Award from the Royal Society of Medicine in London.
He received a bachelor's degree from Bowling Green State University and a doctor of medicine degree from the University of Cincinnati College of Medicine. He completed his residency in internal medicine at Yale-New Haven Hospital and his fellowship in rheumatology at the National Institutes of Health and the University of California at San Diego.

John H. Stone, M.D., M.P.H., co-founded and directed the Vasculitis Center at Johns Hopkins University. Dr. Stone attended Harvard Medical School before training in internal medicine at Johns Hopkins and performing his rheumatology fellowship at the University of California-San Francisco. While on the faculty at Johns Hopkins, Dr. Stone served as the Principal Investigator for first randomized clinical trial in Wegener's granulomatosis in the U.S. and organized the Rituximab in ANCA-Associated Vasculitis trial. From 2002 to 2006, Dr. Stone served as the Deputy Director for Clinical Research at the Johns Hopkins Bayview Medical Center. He was named a Hugh and Renna Cosner Scholar in the Cosner Program on Translational Research (2005). Dr. Stone became Deputy Editor for Rheumatology at UpToDate in 2006 and is an Associate Physician at the Massachusetts General Hospital.

Leslie J. Crofford, M.D. is an active member of the American College of Rheumatology, serving previously as a member of the Committee on Research and Chair of the Committee on Journal Publications. She is currently Vice-President of the American College of Rheumatology Research and Education Foundation and sits on the Executive Committee of the College. Dr. Crofford was elected to the American Board of Internal Medicine for Rheumatology in 2002 and is currently serving her second term. She is on the Board of Trustees of the Ohio River Valley Chapter of the Arthritis Foundation and has served on the Medical and Scientific Committee of the National Arthritis Foundation. Dr. Crofford is active as a clinical rheumatologist and has been named as one of America's Top Doctors.

Patience White, M.D. is the chief public health officer of the Arthritis Foundation. In addition to her work there, she is a professor of medicine and pediatrics at the George Washington University School of Medicine and Health Sciences and teaches a Health Policy seminar for Stanford University at the Stanford in Washington campus in Washington DC.

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