

Primer on the Rheumatic Diseases

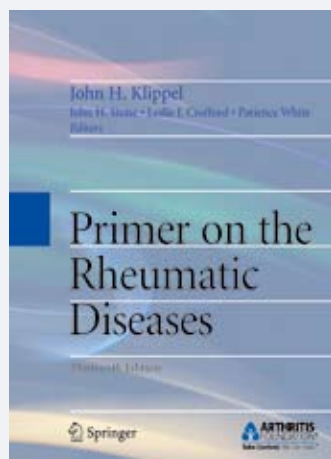
Edited by: John H. Klippel, John H. Stone, Leslie J. Crofford,
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Primer on the Rheumatic Diseases

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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

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- Individual chapters (and more than doubling of the allotted text) to the metabolic and inflammatory myopathies, once included in the same chapter.
- Reorganization of the vasculitis section along more rational and all-inclusive lines, with a chapter entitled "ANCA-Associated Vasculitis" that addresses together Wegener's granulomatosis, microscopic polyangiitis, and the Churg-Strauss syndrome, disorders with striking similarities but important contrasts.
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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 pathology. 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–.5g 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 dermatitis–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

CHAPTER 25 • LESS COMMON ARTHROPATHIES 493

ASSOCIATIONS WITH NEUTROPHILIC DERMATOSES.

SWEET'S SYNDROME	PYODERMA GANGRENOSUM (PG)	RHEUMATOID NEUTROPHILIC DERMATITIS	BOWEL-ASSOCIATED DERMATITIS ARTHRITIS SYNDROME	NEUTROPHILIC DERMATITIS 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

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monocyte colony-stimulating factor imatinib, minocycline, hydrocortisone. For acute disease, a dnisone tapered over 2 weeks is current disease without an 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.

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

FEATURE	WEGENER'S GRANULOMATOSIS
ANCA positivity	80%–90%
ANCA antigen specificity	PR3 > MPO
Fundamental histology	Leukocytoclastic vasculitis; necrotic granulomatous inflammation (rare in renal biopsy specimens)
Ear/nose/throat	Nasal septal perforation; saddle-nose deformity; conductive or sensorineural hearing loss; subglottic stenosis
Eye	Orbital pseudotumor, scleritis (rhegmatoid), scleromalacia perforans, episcleritis, uveitis
Lung	Nodules, infiltrates, or cavity lesions; alveolar hemorrhage
Kidney	Segmental necrotizing glomerulonephritis; rare granulomatous features
Heart	Occasional valvular lesions
Peripheral nerve	Vasculitic neuropathy (10%)
Eosinophilia	Mild eosinophilia occasionally

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

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.



FIGURE 25E-1
Sweet's syndrome.

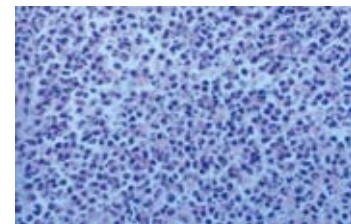


FIGURE 25E-2
Histopathological findings in Sweet's syndrome.



FIGURE 21C-1
Saddle-nose deformity in Wegener's granulomatosis.

Color figures depict
cutaneous findings and
histopathology

Details musculoskeletal
signs and symptoms

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