Seminars in Diagnostic Pathology 34 (2017) 273–284

Contents lists available at ScienceDirect

Seminars in Diagnostic Pathology

journal homepage: www.elsevier.com/locate/serndb

Disorders characterized by predominant or exclusive dermal inflammation

Mark R. Wick

Section of Dermatopathology, Division of Surgical Pathology & Cytopathology, University of Virginia Medical Center, Room 3020 University of Virginia Hospital, 1215 Lee Street, Charlottesville, VA 22908-0214, USA

ARTICLE INFO

Keywords:
Dermal inflammatory diseases
Gyrate erythemas
Viral exanthems
Pigmented purpura
Polymorphous light eruption
Tumid lupus
Cutaneous lymphoid hyperplasia
Sweet syndrome
Pyoderma gangrenosum
Hidradenitis suppurativa
Autoinflammatory dermatitis
Arthropod bite reactions
Parasitic infestations
Bullous pemphigoid
Wells syndrome
Hypereosinophilic syndrome
Churg-Strauss disease
Drug eruptions
Chronic urticaria
Urticarial vasculitis

ABSTRACT

Some cutaneous inflammatory disorders are typified by a predominant or exclusive localization in the dermis. They can be further subdivided by the principal cell types into lymphocytic, neutrophilic, and eosinophilic infiltrates, and mixtures of them are also seen in a proportion of cases. This review considers such conditions. Included among the lymphoid lesions are viral exanthems, pigmented purpuras, gyrate erythemas, polymorphous light eruption, lupus tumidus, and cutaneous lymphoid hyperplasia. Neutrophilic infiltrates are represented by infections, Sweet syndrome, pyoderma gangrenosum, and hidradenitis suppurativa, as well as a group of so-called "autoinflammatory" dermatitides comprising polymorphonuclear leukocytes. Eosinophil-dominated lesions include arthropod bite reactions, cutaneous parasitic infestations, the urticarial phase of bullous pemphigoid, Wells syndrome (eosinophilic cellulitis), hypereosinophilic syndrome, and Churg-Strauss disease. In other conditions, eosinophils are admixed with neutrophils in the corium, with or without small-vessel vasculitis. Exemplary disorders with those patterns include drug eruptions, chronic idiopathic urticaria, urticarial vasculitis, granuloma faciale, and Schnitzler syndrome (chronic urticarial with a monoclonal gammopathy).

Several dermatitides are typified by inflammatory cell infiltrates in the corium, with limited or no involvement of the epidermis. These disorders have a variety of etiologies, and the nature of the leukocytes that mediate them is also heterogeneous. Despite that seeming complexity, a systematic approach can be taken to the diagnosis of predominantly-dermal inflammatory skin conditions. This review endeavors to present the essentials of that process.

Dermal inflammation dominated by lymphoid cells

Chronic inflammatory disorders in the dermis may comprise relatively modest numbers of lymphoid cells, or, in counterpoint, they may efface almost the entire corium. Even when only perivascular aggregates of lymphocytes are present, their sizes and densities may lead the histological differential diagnosis in dissimilar directions. In addition, potential associations with pigment deposits, erythrocyte extravasation, zones of edema, or stromal mucinosis are likewise important (Fig. 1).

Scant perivascular dermal lymphoid infiltrates

Disorders featuring the presence of scanty perivascular dermal lymphoid infiltrates (PDLI) may simply represent very early phases of dermatitides that are not yet fully developed, such as incipient granuloma annulare.[1] That eventuality can be addressed by obtaining biopsies from lesions at clinically-different stages of evolution, to determine whether they have dissimilar microscopic appearances. In circumstances where PDLIs are consistently unimpressive in size or density in several samples, it may be assumed that this image does indeed accurately reflect the disease in question.

The next point of attention in such cases is whether the lymphoid infiltrates are associated with extravasated erythrocytes or deposits of hemosiderin. If so, the likely diagnosis is that of a pigmented purpuric eruption (PPE) (Fig. 2). Synonyms include "progressive pigmentary dermatosis," "purpura pigmentosa chronica," and "progressive pigmenting purpura." Eponymous examples of these conditions are Schamberg's disease, Majocchi's disease, Gougerot-Blum syndrome, and Ducas & Kapetanakis purpura.[2,3] They all feature inflammatory damage to small dermal blood vessels, which become structurally-incompetent and spill erythrocytes into the surrounding corium. Over time, that process results in hemosiderin

E-mail address: mrwick1@usa.net

http://dx.doi.org/10.1053/j.semdp.2016.12.002
0740-2570/

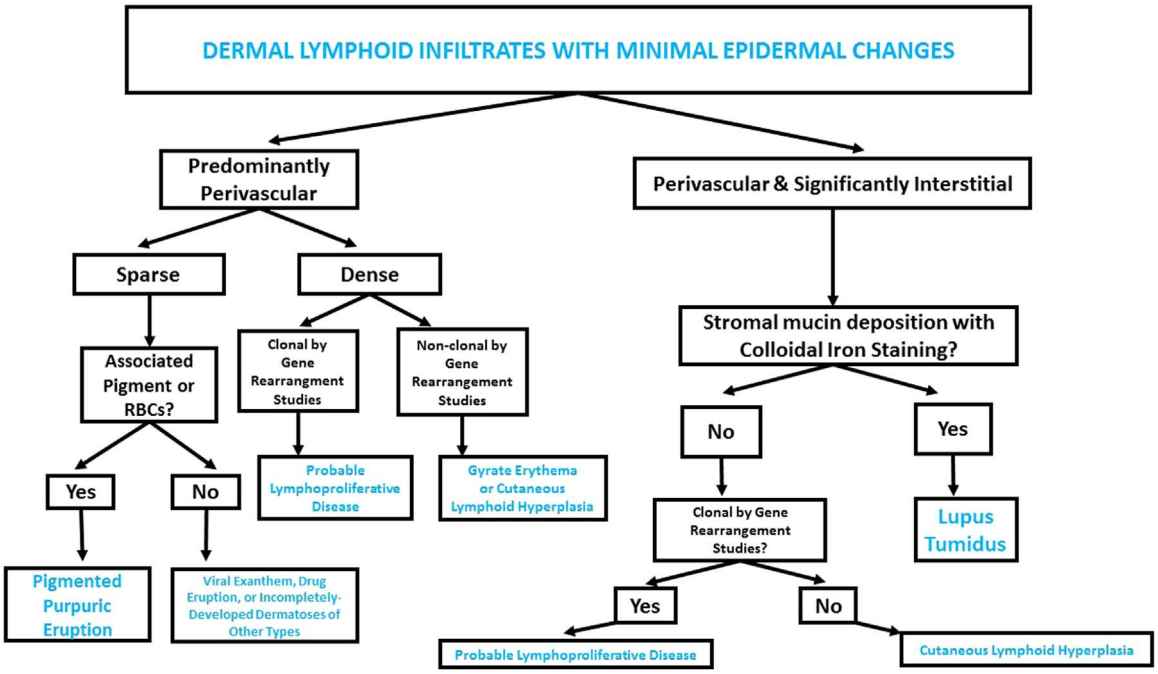

Fig. 1. Algorithm for histopathologic diagnosis of dermal infiltrates dominated by lymphoid cells.

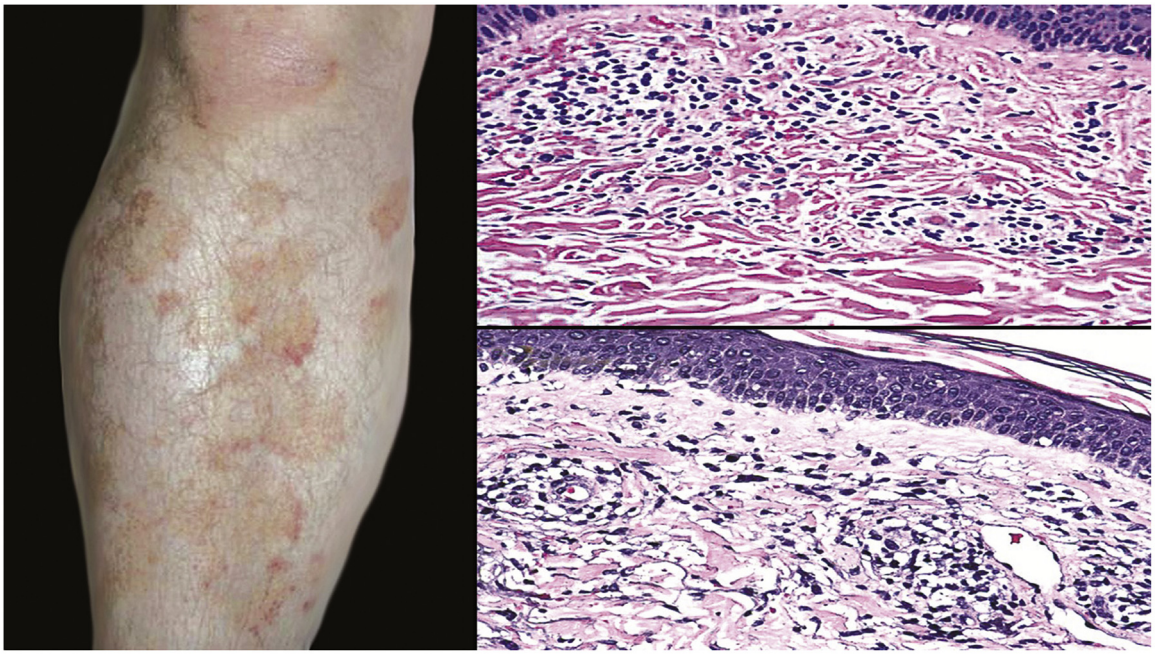

Fig. 2. Pigmented purpuric eruption. Coalescent macules and papules with a "cayenne pepper" appearance are present on the legs (left). Microscopically, a scant perivascular dermal infiltrate of lymphocytes is seen, together with scattered erythrocytes (right panels).

deposition. Clinically, PPEs manifest with painless, non-blanchable, reddish-, cayenne pepper-, or copper- colored macules, which often coalesce to form patches. The legs are the favored location for such lesions, and adults are usually affected.[2]

In the absence of associated erythrocytes or hemosiderin, two other diagnostic possibilities become principal considerations. Those are viral exanthems and lymphocyte-mediated cutaneous drug reactions.

Many types of viruses can produce skin lesions. In addition to those in the *Herpes* family, others include poxviruses, papoviruses, reoviruses, orthomyxoviruses, togaviruses, paramyxoviruses, flaviviruses, parvoviruses, hepadnaviruses, lentiviruses (such as human immunodeficiency virus), and other retroviruses. The most common —but the most nondescript—viral exanthem is a diffuse, morbilliform, maculopapular eruption (Figs. 3 and 4), similar to that seen in rubeola (measles). It can be a manifestation of infection with many, if not most, of the viruses just mentioned above.[4–11] Histologically, non-herpetic, non-poxvirus-related viral skin eruptions usually produce only an unimpressive dermal perivascular chronic inflammatory infiltrate, with minimal, if any, abnormalities in the epidermis. The pathologic findings typically do not allow for a precise identification of the causal organism; indeed, of themselves, they do not even lead to a definitive conclusion that the illness in question is infectious. That decision must rest on clinical correlation.[9]