Acquired cutaneous perforating disorders: Clues to diagnosis by silhouette

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ABSTRACT
Inflammatory dermatoses are interpreted by pattern recognition. Primary perforating disorders including reactive perforating collagenosis, elastosis perforans serpiginosa, perforating folliculitis, and Kyrle’s disease show clinical and histopathological overlap. This article highlights the importance of architectural based analysis by “silhouette” observed under scanning magnification for diagnosis of primary perforating disorders.

Key words: Cutaneous, perforating disorders, silhouette

INTRODUCTION
Dermatology is an intrinsically visual speciality. Historically, drawings have been used to illustrate both clinical and histopathological aspects of skin lesions.[1] Pattern recognition is a vital skill for histopathological diagnosis of inflammatory dermatoses. This approach was well illustrated in the textbook written by Dr. Bernard Ackerman that is used universally by all practicing dermatopathologists.[2]

“Silhouette” of the skin lesion, observed under scanning magnification, provides an overview of the histopathology and facilitates distinction of closely related conditions. This approach is exemplified in evaluation of cutaneous perforating disorders, the focus of this article.

Perforating disorders may be divided into primary and secondary disorders. The key histopathologic feature of the primary type is perforation of the dermo-epidermal junction with transepidermal elimination. The primary type may be acquired or familial. Both acquired and inherited forms show similar histopathology. Distinction is based on clinical presentation. Inherited form presents in childhood. Acquired forms occur in adults over 18 years of age and are mostly seen in setting of diabetes, chronic renal failure, and hyperuricemia.[3]

In secondary type, the perforation and transepidermal elimination of collagen/elastic fibers and/or other material (amyloid, calcium, and fungal hyphae) occur as a secondary event to an underlying pathology such as granuloma annulare, necrobiosis lipodica, pseudoxanthoma elasticum, calcinosis cutis, healing wounds, and keratoacanthoma.[3,4]

Perforating disorders are characterized by umbilicated papulo-nodular lesions with keratotic plug.[3] To take advantage of the architectural silhouette, the biopsy must include the entire lesion centered over the keratotic plug and surrounding epidermis, best achieved by selecting a small size lesion.

The primary perforating disorders, namely, reactive perforating collagenosis (RPC), elastosis perforans...
serpiginosa (EPS), perforating folliculitis (PF), and Kyrle’s disease (KD) will be included in this article. The transepidermal elimination observed in the primary disorders relates to collagen, elastic fibers and keratinous debris.

**REACTIVE PERFORATING COLLAGENOSIS**

The silhouette is saucer shaped [Figure 1a]. A shallow crateriform epidermal invagination, filled with basophilic debris, neutrophils, parakeratotic keratin, and collagen fibers, is seen [Figure 1b]. Surrounding epidermis is acanthotic. Epidermal base shows variable features ranging from full-thickness necrosis to being partially damaged. Mild perivascular inflammation is seen in the underlying dermis. Diagnostic clue is to locate transgressing collagen fibers. Such fibers can be identified at several spots. Extruding collagen fibers stand out by being vertically oriented at the entry point along the base of the invagination and appearing as a “rocket” streaking through the basophilic keratotic plug. A careful search may show fibers interposed between the keratinocytes at the junction between the plug and intact epidermis [Figure 1c and d]. Collagen fibers appear pink with H and E and blue with Masson’s trichrome stain. Keep in mind that collagen fibers traversing through invagination may lose their staining quality [Figure 1d].

**ELASTOSIS PERFORANS SERPIGINOSA**

The silhouette is serpiginous [Figure 2a]. Irregular epidermal hyperplasia forms a curvilinear channel with overlying keratinous plug and neutrophils [Figure 2b]. A bulbous invagination at the base is filled with basophilic debris and coarse clumped thickened eosinophilic elastic fibers [Figure 2c]. The perforating channel can be hyperplastic epidermis, follicular, or sometimes acrosyringeal. Foreign-body giant cell reaction is sometimes noted in adjacent dermis. Elastic stains such as Verhoeff-Van Gieson [Figure 2d] and Orcein show increased coarse elastic fibers in the papillary dermis. Elastic fibers appear pale or eosinophilic as they are eliminated. The best stain for dermal elastic fibers in this condition is Giemsa. Altered fibers in EPS may resemble hyphae of Mucor or Rhizopus [Figure 2c]. The lesion heals with a dermal scar that is devoid of elastic fibers.

Papular lesions with central scale of EPS are arranged in annular or serpiginous pattern. Clinical differential diagnosis could include other annular lesions such as granuloma annulare (which may also show incidental perforation), tinea corporis, porokeratosis, and sarcoidosis. All these conditions show distinctive pathology.

**PERFORATING FOLLICULITIS**

The silhouette is angulated vase [Figure 3a]. The follicular infundibulum/isthmus is dilated and is filled with necrotic debris, ortho- and parakeratotic keratin, and inflammatory cells [Figure 3b]. The perforation appears along the thinned lateral wall of follicular epithelium [Figure 3c]. Adjoining dermis shows...
Degenerative changes and sometimes, eosinophilic collagen and/or elastic fibers are seen entering the perforation. Hair shaft may be seen within the follicle as curled up or trapped within the debris.\textsuperscript{[4]} Disruption or break of the follicular epithelium along lateral border should be noted. Sometimes, one observes thinning of the epithelial wall, which is the likely site of perforation, surrounded by inflammatory infiltrate, a mix of neutrophils and granulomatous infiltrate. It is possible that the initial sections will not reveal the follicular pathology in its entirety. In the absence of follicle in the section, the clue to presence of follicular pathology will be a localized area of inflammation in the dermis.

Differential diagnosis includes keratosis pilaris and various folliculitides.\textsuperscript{[4]} Keratosis pilaris does show dilated follicular infundibula, but perforation is absent. Ruptured folliculitis shows extensive damage to the follicular epithelium and is much more inflammatory.

**Kyrle’s Disease**

The silhouette is flask shaped [Figure 4a]. On a historical note, the first case was described by Dr. Kyrle in a diabetic woman, illustrated by a fine histopathologic sketch in the original paper published in 1916.\textsuperscript{[4]} The round flask-shaped invagination is filled with parakeratotic plug and basophilic necrotic debris [Figure 4b]. Invagination either involves follicle or is epidermal. As the lesion evolves, the basal layer becomes attenuated and usually breaks at one or more points thus exposing the keratinous material to the dermis.\textsuperscript{[4]} Serial sections are helpful in demonstrating the perforation. However, perforation does not always occur and is not an absolute requirement for the diagnosis. Surrounding epidermis is acanthotic. Inflammatory infiltrate of neutrophils at the point of perforation and perivascular lymphocytic infiltrate in the vicinity are seen. An occasional elastic fiber is seen only exceptionally and not as a routine.\textsuperscript{[4]}

Clinically, most lesions of KD are follicular based. Thus, the closest differential diagnosis is PF. Histologically, follicular infundibulum containing curled up hair shaft with a perforation along the lateral wall favors PF.\textsuperscript{[4]}

**CONCLUSION**

The perforating disorders have overlapping clinical and histological features. Furthermore, transepidermal elimination of collagen and/or elastic fibers is common to all perforating disorders. Therefore, silhouette-based pattern is a useful clue for distinction.

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