



# Arizona Veterinary Specialists' News



LEADERS IN SPECIALTY CARE

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## The Pemphigus Complex: A Brief Review

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The pemphigus complex is a group of uncommon autoimmune skin diseases that affect both cats and dogs alike. These diseases are typically identified by acantholysis, or a breakdown in the bonds connecting the epidermis. This results in the presence of acantholytic keratinocytes, which are round darkly staining “fried egg”-like cells. The more common of the subsets of this complex is pemphigus foliaceus, however this group of autoimmune disease also includes variants such as pemphigus vulgaris, paraneoplastic pemphigus, and pemphigus erythematosus to name a few. This will be a brief review of the subsets of the pemphigus complex seen by veterinary dermatologists.

### Pemphigus Foliaceus

Pemphigus foliaceus (PF), canine in particular, is the most commonly identified subset of the pemphigus complex. This disease can develop at any age, but is more commonly seen in middle-aged to older dogs. Pemphigus foliaceus is typically idiopathic in nature, however possible association with drug administration has been established. The disease can resolve once the drug has been withdrawn, or persist permanently.

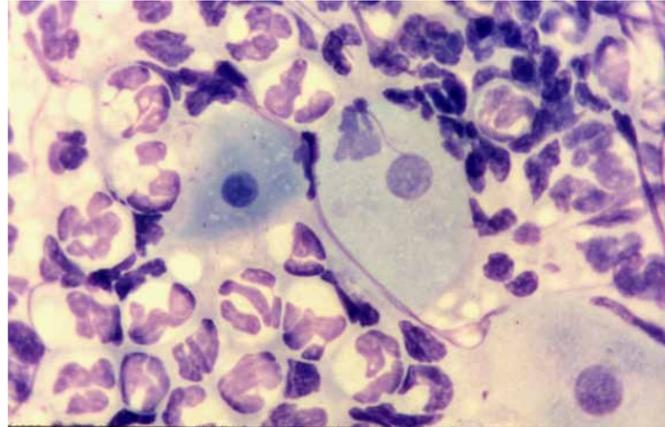
PF is defined as a pustular disease, with pustules to erosions developing typically over the head, face, and ears. These lesions are typically bilaterally symmetric, and if chronic can present as widespread areas of serous crusting or post-inflammatory alopecia (Figure 1) The footpads are often also affected, some with identifiable pustules and others with fissures and crusting along the paw pads themselves. Rapid development of lesions or slow, chronic changes can be seen.



**Figure 1.** Large “bullous” pustules spanning more than one follicle caused by pemphigus foliaceus.

Diagnosis is ideally made with biopsy of intact pustules, which histopathologically is suggestive of pemphigus foliaceus if it contains acantholytic keratinocytes and neutrophils residing in subcorneal (meaning within the stratum corneum) pustules. These cells are typically also seen on cytologic evaluation of pustules (Figure 2), but

care must be made to exclude other causes of acantholysis (certain fungal infections, deep staphylococcal infections) prior to a diagnosis of pemphigus foliaceus. A detailed history of drug administration should be taken at time of appointment, to identify any potential causative link. Prognosis is typically good pending response to appropriate therapy.



**Figure 2.** Note three large basophilic staining acantholytic keratinocytes amongst neutrophils. Note lack of microorganisms such as coccoid bacteria.

### Pemphigus Vulgaris

Pemphigus vulgaris, while more severe than pemphigus foliaceus, is a more rare manifestation of the pemphigus complex. Slow or rapid onset of lesions can be seen, typically located around the mouth and mucocutaneous junctions. Intact pustules/bulla can be seen at these locations but more often they are ruptured, leaving behind large erosions along the mucocutaneous junctions.

As lesions in pemphigus vulgaris are deeper within the skin, cytology is rarely helpful as acantholytic keratinocytes are rarely seen. Ideally referral to a veterinary dermatologist for biopsy is recommended, as ideal locations for biopsy for a greater chance of obtaining a diagnosis are not easy to identify. Biopsy results in this disease typically show a suprabasilar intraepidermal cleft, namely detachment of the cells within a certain

layer of the epidermis. Prognosis is unfortunately relatively poor in these cases.

### Paraneoplastic Pemphigus

This extremely rare variant of the pemphigus complex described in a few published case reports. In these cases, underlying neoplasia (one a thymic mass, another a splenic mass) was identified after the patients were non-responsive to immunosuppressive therapy. The theory for this variant of pemphigus is that underlying neoplasia induces circulating autoantigens that are the hallmarks of pemphigus. These cases typically respond very poorly to medical management.

### Pemphigus Erythematosus

Pemphigus erythematosus is a variant of the pemphigus complex considered a combination between two dermatologic autoimmune diseases: pemphigus foliaceus and discoid lupus erythematosus. The most common presentation of this variant of pemphigus is symmetrical crusting lesions along the dorsal muzzle, face, and ears. This can lead to erosions and ulcerations of the nasal planum, with loss of pigment (Figure 3). Referral to a veterinary dermatologist for biopsy is recommended, as histopathology of these lesions show evidence of both pemphigus foliaceus and discoid lupus erythematosus.

Luckily, this variant of pemphigus is typically not aggressive, and responds well to the appropriate long-term immunosuppressive therapy for control of pemphigus as well as environmental management for decreased sun exposure.



**Figure 3.** Note crusting of planum nasale and periocular skin due to pemphigus erythematosus.

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