

## Insight Into Pemphigus –An Autoimmune Skin Disease

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### Definition

Pemphigus is an autoimmune dermatosis. It is characterized by the formation of autoantibodies against the keratinocyte membrane that loosens intercellular adhesions between keratinocytes, leading to an intraepidermal pustule or vesicle formation.

### Types

- (i) Pemphigus foliaceus (most common)
- (ii) Pemphigus erythematosus
- (iii) Pemphigus vulgaris (more serious)
- (iv) Pemphigus vegetans (only in dogs)
- (v) Paraneoplastic pemphigus.

### Clinical Findings

#### Pemphigus foliaceus

- The primary lesion in canine pemphigus is a pustule. These pustules are replaced by yellow to brown crusts. Lesions may begin around the nasal planum and eyes, dorsal muzzle, lip margins and pinnae. Nasal depigmentation can be seen. The distribution of pustules in pemphigus foliaceus is either truncal or non-truncal (in limbs).
- The less common findings are pruritus, mucosal & oral lesions, paronychia, and hyperkeratotic & fissured foot pads. Severe cases may develop bacterial cellulitis. Alopecia and seborrhoea are seen in more chronic cases.

#### Pemphigus foliaceus



### **Pemphigus erythematosis**

- Pemphigus erythematosis is characterized by an erythematous pustular dermatitis of the face and ears.

### **Pemphigus vulgaris**

- It is a more serious disease than pemphigus foliaceus, as the lesions develop deeper in the epidermis and leave erosions and ulcerations. Vesicular/bullous, erosive to ulcerative lesions are seen primarily in the oral cavity and mucocutaneous junctions.

### **Diagnosis**

Diagnosis is based on clinical history, physical examination, findings, cytology, and histopathology. Adjunctive tests include direct and indirect immunofluorescence testing as well as immunoperoxidase staining.

#### **1. Cytology**

Cytology of an intact pustule is obtained by puncturing the pustule, expressing its contents onto a glass slide and staining the sample. Cytology reveals non-degenerative neutrophils and eosinophils. Large numbers, rafts or clusters of acantholytic keratinocytes are very suggestive of pemphigus.

#### **2. Biopsy**

The most important diagnostic tool is the skin biopsy. Tissue immunohistochemistry can be performed on the skin biopsy sample.

### **Management And Therapeutics**

Pemphigus is treated with either glucocorticoids alone or in combination with other immunosuppressive drugs. Often, a combination of drugs gives a better result than glucocorticoids alone.

#### **Glucocorticoids**

- Mild skin lesions are treated with topical glucocorticoids such as betamethasone or fluocinolone until lesions resolve.
- In moderate cases prednisone or prednisolone at an immunosuppressive (induction) dose of (up to) 2.2 to 4.4 mg/kg/day in dogs and 4.4 to 6.6 mg/kg/day in cats. In most cases, disease remission occurs in two to four weeks. Thereafter maintained a target dose of 1.1 mg/kg every other day in the dog and 2.2 mg/kg every other day in the cat is achieved.
- Pulse therapy is indicated in severe cases. In this methylprednisolone acetate @ 11 mg/kg is given in 5% Dextrose is administered over a period of 1 to 3 hours daily for one to three days. This is followed by prednisolone at an immunosuppressive dose and is tapered to a maintenance dose.



### Immunosuppressive drugs

Commonly used immunosuppressive drugs are **azathioprine** and **chlorambucil**.

### Miscellaneous drugs

- **Gold salts:** gold salts (chrysotherapy) can be used as a follow up therapy, after steroids have been tapered and discontinued. In dogs and cats, **urothioglucose** is given at 1 mg/kg weekly IM and at increasing intervals. The oral dose of **auranofin** is 0.1-0.2 mg/kg sid. Adverse side effects include hepatotoxicity, bone marrow suppression, and proteinuria.
- **Dapsone** and **sulfasalazine** has been tried with variable success. **Mycophenolate mofetil:** Mycophenolate mofetil @ 22 to 39 mg/kg q8h gave 50% success in treating pemphigus foliaceus in dogs.

### Reference

1. BSAVA Manual of Small Animal Dermatology, 2nd edition, 2003, edited by Aiden P.Foster and Carol S. Foil, Published by British Small Animal Veterinary Association.

