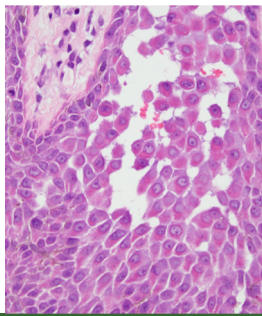


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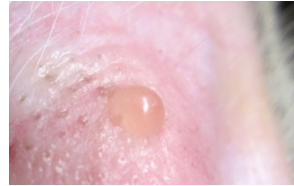
## Introduction

- Autoimmune pustular skin disease characterized by acantholysis: loss of adhesion between keratinocytes



## Pemphigus Introduction

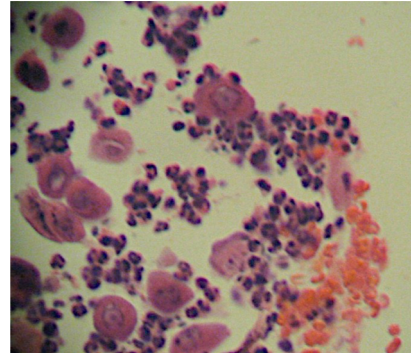
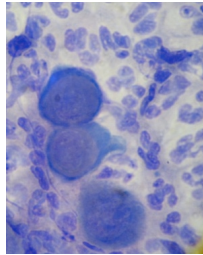
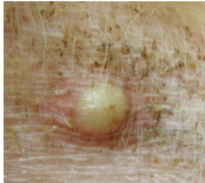
- Pemphigus: Greek word “blister”
- Superficial:
  - **pemphigus foliaceus (PF)**
  - pemphigus erythematosus (PE)?
- Deeper:
  - Pemphigus vulgaris (PV)
  - Paraneoplastic pemphigus (PNP)
  - Pemphigus vegetans (PVeg)



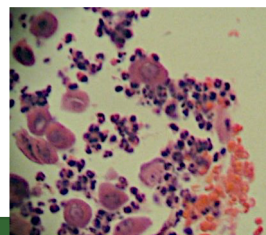
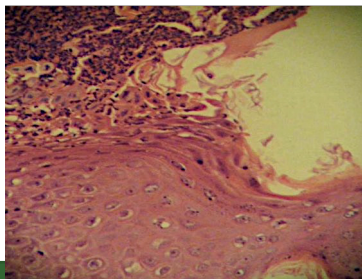
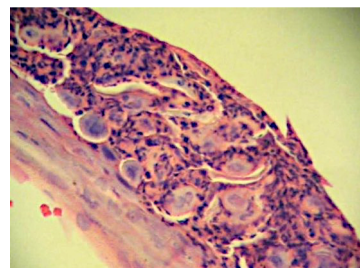
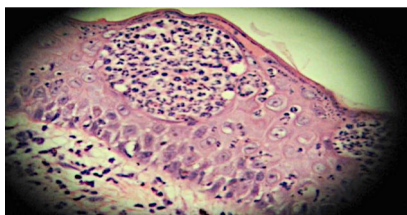
## Pemphigus foliaceus

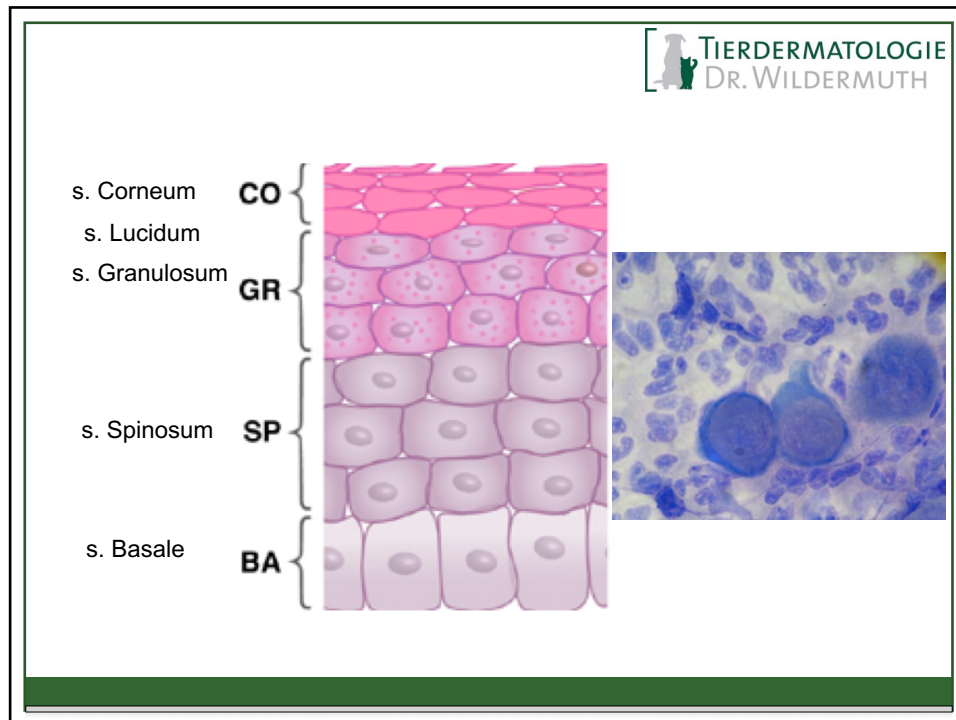
- Most common autoimmune disease: dogs & cats
  - Chow Chow, Akita
- Lesions:
  - Pustule -> Crust with underlying ulceration -> Alopecia
  - May have fever, cellulitis (limb edema), lethargy, pruritus
- Location:
  - Face dominant +/- paw pads
    - Periocular, bridge of the nose, pinnae
  - Trunk dominant +/- paw pads
  - Combination

Classic Lesion = Pustule  
Classic Cell = acantholytic cell



## Pemphigus foliaceus





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## Destruction of Desmosomes

- **Exact** mechanism unclear
- **Classical theory: Steric hindrance**
  - Antibodies bind desmosomal proteins and physically disrupt the desmosome



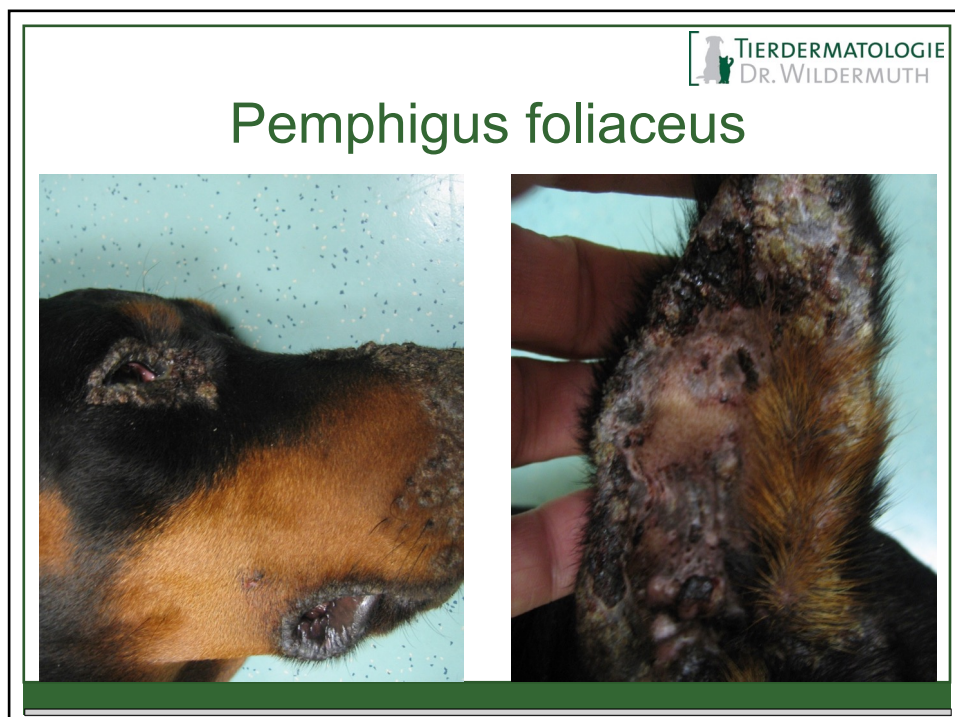
## Pathogenesis



- **Pemphigus foliaceus**
  - Major autoantigen:  
Desmocollin 1 (Dsc1)
    - P. Bizikova, et. al, 2012
  - Minor autoantigen:  
Desmoglein 1 (Dsg1)
- **Pemphigus vulgaris**
  - Major autoantigen:  
Desmoglein 3 (Dsg3)

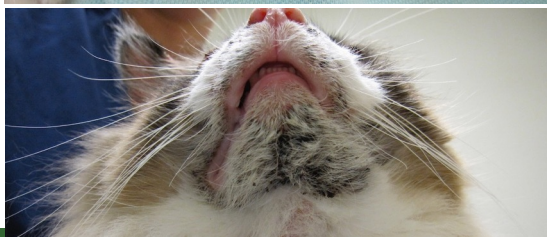
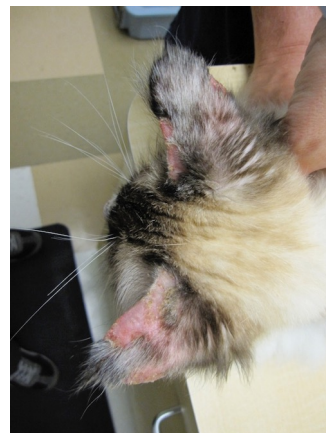
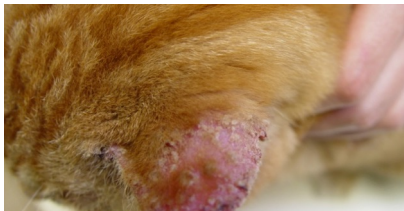








## Pemphigus foliaceus







## Drug Induced PF



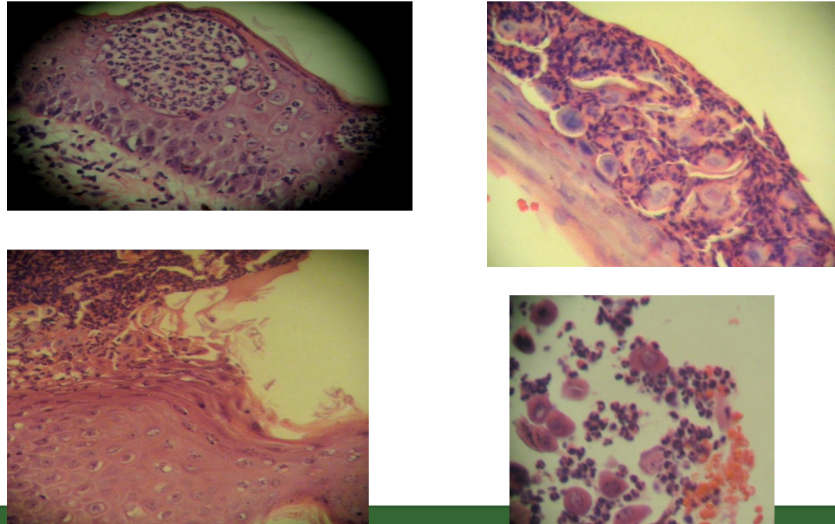
Trimethoprim-sulfa induced PF

## Metaflumazone/amitraz topical induced PF

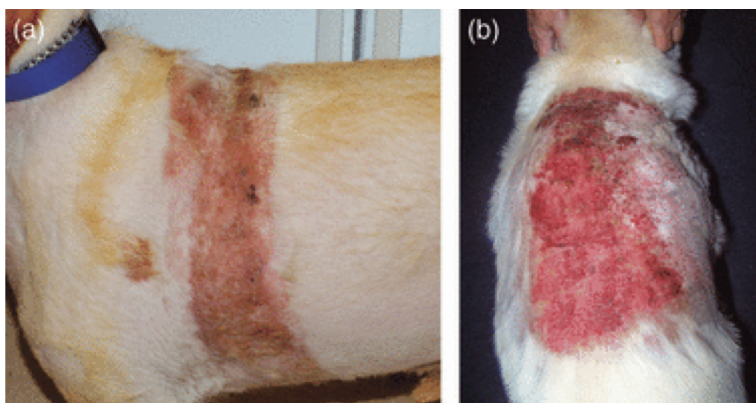


Rosenkrantz


## Pemphigus like drug reaction




## Metaflumazone/amitraz topical induced PF




Oberkirchner 2011



## Canine Contact Metaflumazone/amitraz -triggered PF



Oberkirchner 2011



## Pemphigus erythematosus (PE)

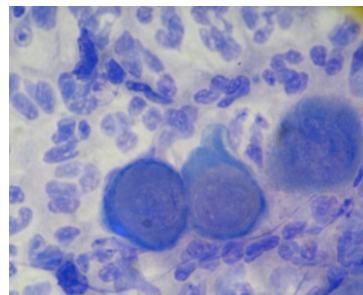
- Variant of PF limited to the face
  - Controversial
  - Does not generalize
- Similar to PF & DLE histologically:
  - Epidermal Pustules but also interface dermatitis
- Collies & German Shepherds predisposed
- Photoaggravation factor?

## Pemphigus erythematosus?



## Diagnosis: Pemphigus foliaceus

- Clinical Symptoms
- Cytology: acantholytic cells
  - +/- Pyoderma (cocci, neut.)
- Negative Fungal Culture
- BIOPSY!

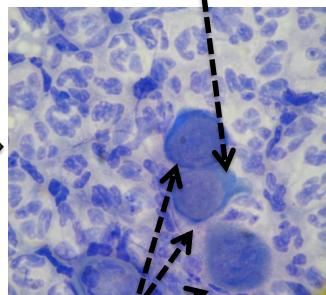
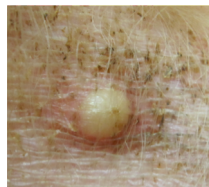
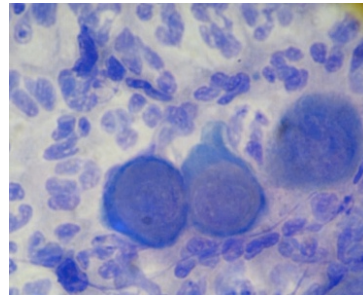




## Cytology: Acantholytic Cell

### • Pemphigus Foliaceus

- Dermatophyte:
  - Trichophyton
- Pyoderma
  - Bullous Impetigo
  - Exfoliative Superficial Pyoderma



Cytology:  
Acantholytic cells

Biopsy +  
Fungal  
culture





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Biopsy +  
Fungal  
culture


Dermatophytosis:  
Trichophyton

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## Pemphigus Foliaceus Diagnosis

- Histological presence of acantholysis
  - Most common - corneal, granular or upper spinous cell layers
  - Eosinophils may be abundant


CO  
GR  
SP  
BA

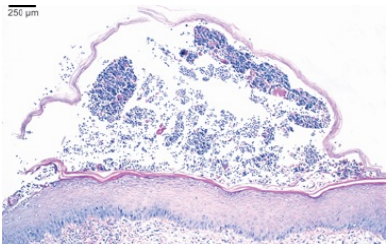
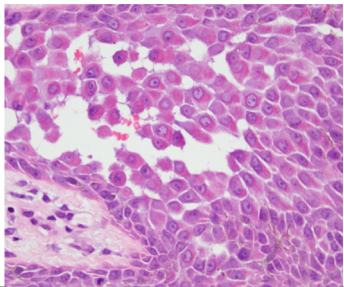


## Pemphigus Foliaceus Diagnosis



Optimal biopsy is an  
Intact pustule



Pemphigus foliaceus



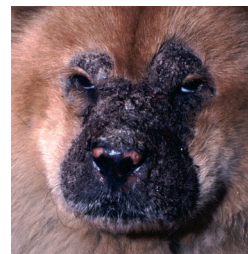
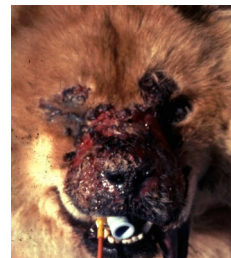



## Blood Panel

- CBC
  - Moderate to marked leukocytosis
  - Neutrophilia
  - Mild non-regenerative anemia
- Chemistry Panel
  - Mild hypoalbuminemia
  - Mild elevations in globulins

## Prognosis

- Variable
  - Most often chronic, long-term therapy
  - Dependent on:
    - Response to medication(s)
    - Number and severity of side-effects of medication(s)
    - Costs
- Müller 2004
  - 97 dogs: 74% survival rate
  - Average time improvement 6 weeks
  - 16 euthanized



## Therapy



- Systemic Therapy often needed
  - 1. Glucocorticoids orally: 1mg/kg twice daily for 5-10 days, then slowly taper – one “strong” day, one “taper” day, alternating
  - Goal: days without steroids: minimize chronic side effects
  - Special considerations with steroids:
    - Larger dogs: PU/PD, Muscle loss
    - Older dogs: muscle loss is more problematic
    - Puppies: PU/PD Polyphagia already a problem
    - Don't combine with NSAIDS

## Pemphigus foliaceus



Besitzer: Karius		Tiername: ...			
Medikament: Dexamethason (Prednisolon) 20mg					
Tag	Datum	Tabletten		gegeben	Bemerkungen (z.B. erbrechen)
		morgens	abends		
1	7.4	1			
2	8.4	1			
3	9.4	1/2			
4	10.4	1			
5	11.	1/2			
6	12	1			
7	13	<del>1/2</del> 1/2			
8	14	1			
9	15	1/2			
10	16	1			
11	17	0			
12	18	1			
13	19	0			
14	20	1			
15	21	0			
16	22	1			
17	23	0			
18	24	1			
19	25	0			
20	26	1/2			
21	27	0			
22	28	1/2			
23	29	0			
24	30.	1/2			

## Therapy



- 2. Azathioprine (AZA)
  - Most common & useful second drug for PF (with steroids)
    - 2mg/kg once daily
  - Special considerations with azathioprine:
    - Very good to reduce steroids: larger dogs
      - Daily side-effects are rare
    - Bone marrow suppression, Hepatitis
      - Drug is cheap
      - Bloodchecks: after 1 week, every 2 weeks for 3 months, then 4 times per year: not cheap

## Therapy



- 2 or 3 Option? Ciclosporin
  - 5mg/kg once daily – 10mg/kg once daily
  - Not as effective as steroids or AZA
    - 50% of the time effective?
  - Very expensive
  - Vomiting, Diarrhea, Lethargy possible
    - Chronic: occasional Hypertrichosis, gingival Hyperplasia
- Apoquel?
  - 0.5mg/kg twice daily – can try it; my experience not helpful
  - Report effective in cat



## Therapy

- 2 or 3 Option: Mycophenolate Mofetil (MMF)
  - lymphocytotoxic immunosuppressive agent
    - Some dermatologists instead of AZA as first drug with steroids
  - Dose: 10mg/kg twice daily
    - **Absorption is variable**
  - A retrospective study of adverse effects of mycophenolate mofetil administration to dogs with immune-mediated disease JVIM 2021 Fukushima et al
    - GI 24.4% [31/127], neutropenia 4% [3/76], anemia 4% [1/25], thrombocytopenia 4.0% [1/25]

## Topical Therapy



## Topical Therapy

- Triamcinolon 0,1% twice daily, then taper
- Tacrolimus 0,1% twice daily, then taper
- Hydrocortison 0,584 mg/ml (Cortavance)
  
- Pyoderma and PF
  - Be aware of it (cytology) & treat
  - Can occurs right away OR later if Cushingoid!

## Questions?

