



Latin Comparative Pathology Group

The Latin Subdivision of the CL Davis Foundation

Diagnostic Exercise

Case #: 30 Month: March Year: 2013

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History: 3 year old male llama with one year history of progressive, mildly pruritic, seborrheic skin problems. The skin condition progressed from seborrheic crusty dermatitis to crusty alopecic occasional ulcerated dermatitis. The animal was found dead in the stall. The postmortem examination revealed generalized skin lesions and severe fibrinous peritonitis.



Fig 1- Skin- Head

There is severe periocular and patchy alopecia, with multifocal crusts and few small ulcers. The changes are generalized.



Fig 2- Skin inguinal region

Severe diffuse crusty dermatitis with ulceration

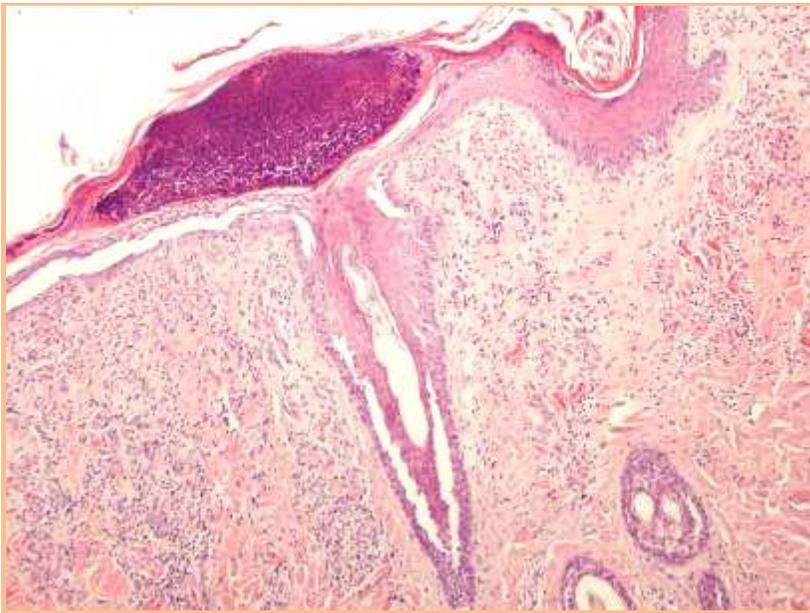


Fig 3- Skin 10x- H&E

Focal epidermal pustular formation. The pustule is filled with degenerated neutrophils. There is suprabasilar acantholysis with vesicle formation.

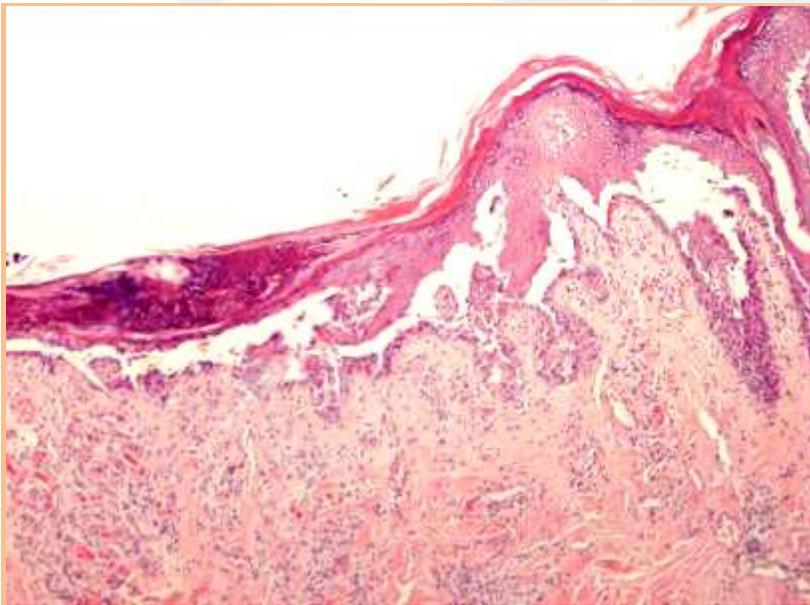


Fig 4a Skin 10x H&E

Occasionally beneath the pustules there are small vesicles at the suprabasal level. In addition there is a superficial perivascular and perifollicular dermatitis.

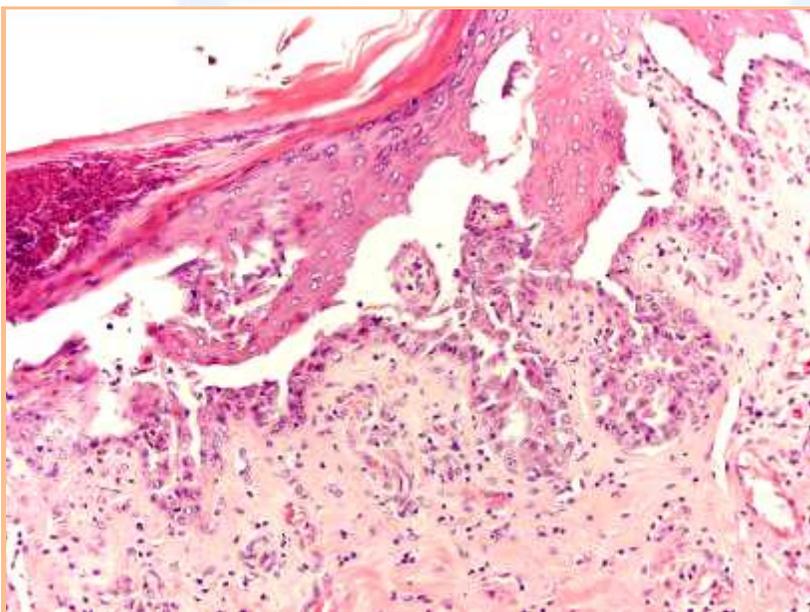


Fig 4b skin 20x H&E

Close magnification of vesicle and bulla formation with epidermal separation at the suprabasal level. The typical lesion of Pemphigus vulgaris is detected at higher magnification. There are intracorneal pustules, with or without suprabasal vesicles. The vesicles extend into the follicular epithelium.

Follow-up questions: *Description, Morphologic diagnosis, Differential diagnosis, possible pathogenesis.*

Morphologic Diagnosis

Haired Skin: Multifocal severe pustular and suprabasilar vesicular dermatitis with occasional ulceration and moderate superficial perivascular dermatitis and intralesional acantholytic cells.

Differential Diagnosis:

The lesions are very suggestive of an immune mediated dermatopathy compatible with Pemphigus vulgaris.

Possible pathogenesis

Autoantibodies against Desmoglein 3 (DSg3)

Discussion

Pemphigus vulgaris (PV) is an immune-mediated dermatosis that has been reported rarely in dogs and cats. In dogs, PV evolves from mucosal to mucocutaneous presentation. PV is a rare disease in horses. The characteristic lesion is the presence of intraepidermal suprabasilar vesicles in the epidermis due to autoantibodies against Desmoglein 3 (DSg3). This immune-mediated response presumably leads to the suprabasilar acantholysis responsible for the vesicle and bulla formation. Special stains with direct immunofluorescent antibodies were performed using anti-goat IgG and anti-llama IgG. Both antibodies were positive at the level of the intercellular space in the epidermis and the vesicles. Dsg 3 is part of the Cadherin group of transmembrane desmosomal adhesion molecules of keratinocytes found at the suprabasilar level. Pemphigus foliaceus (PF) is the most common form of pemphigus and is the most common immune-mediated dermatosis in dogs, cats and horses. PF is an autoimmune dermatopathy, targeting desmoglein 1, which is another adhesion molecule.

References and recommended literature.

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Christiano, a. M., & uitto, j. (1996). Molecular complexity of the cutaneous basement membrane zone. *Experimental dermatology*, 5(1), 1-11.

Olivry, t. (2006). A review of autoimmune skin diseases in domestic animals: i-superficial pemphigus. *Veterinary dermatology*, 17(5), 291-305.

<http://onlinelibrary.wiley.com/doi/10.1111/j.1365-3164.2006.00540.x/pdf>

Please send your comments/questions to the whole LCPG list by hitting "reply to all".

A final document containing this material with answers and a brief discussion will be posted on the C. L. Davis website by the end of the current month (http://www.cldavis.org/lcpg_english.html).