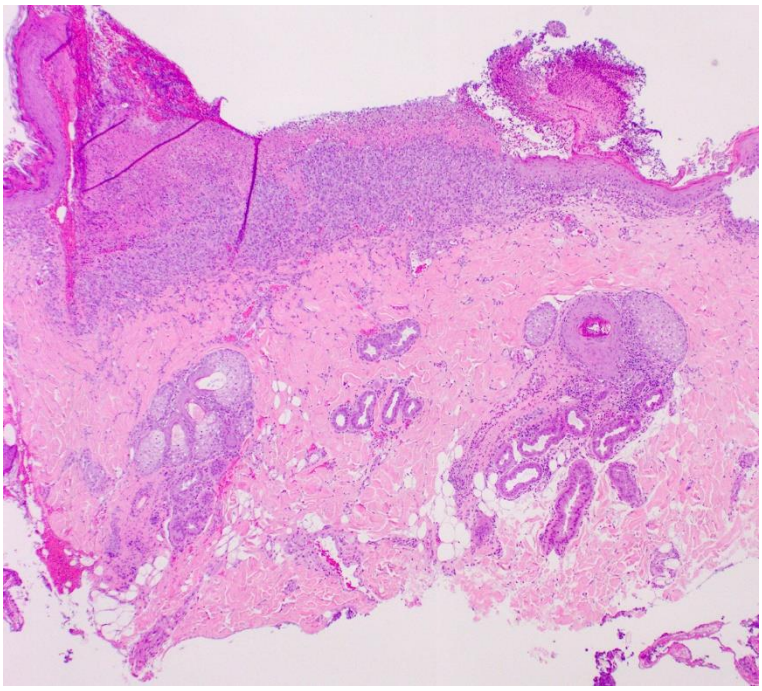
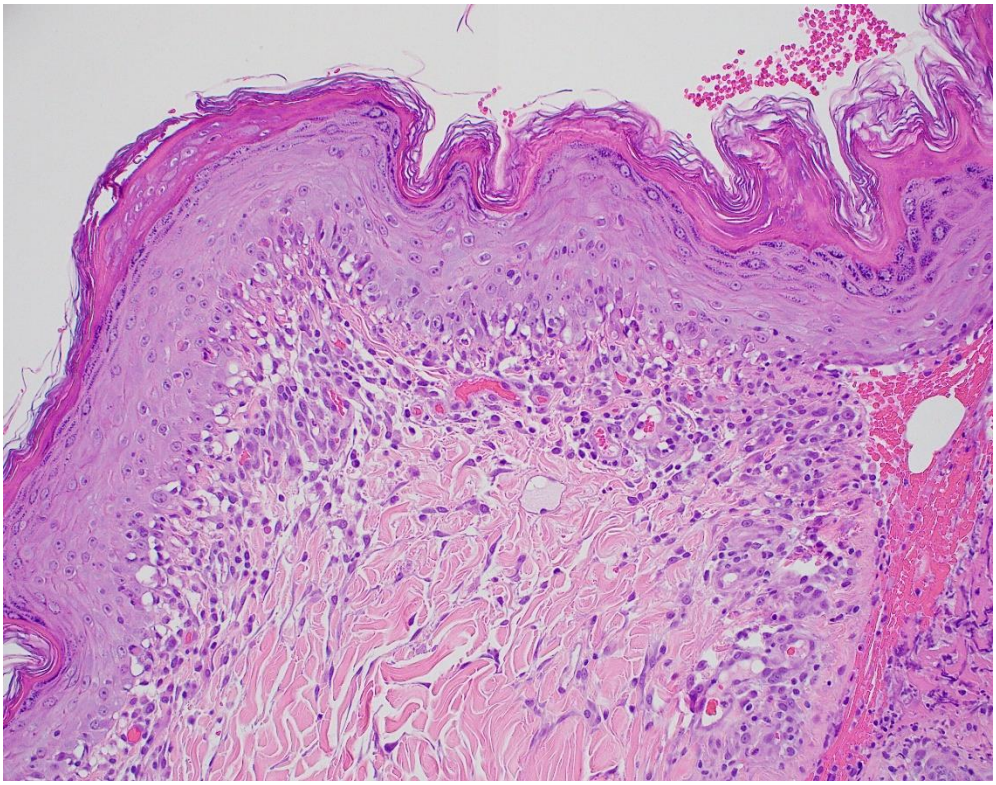
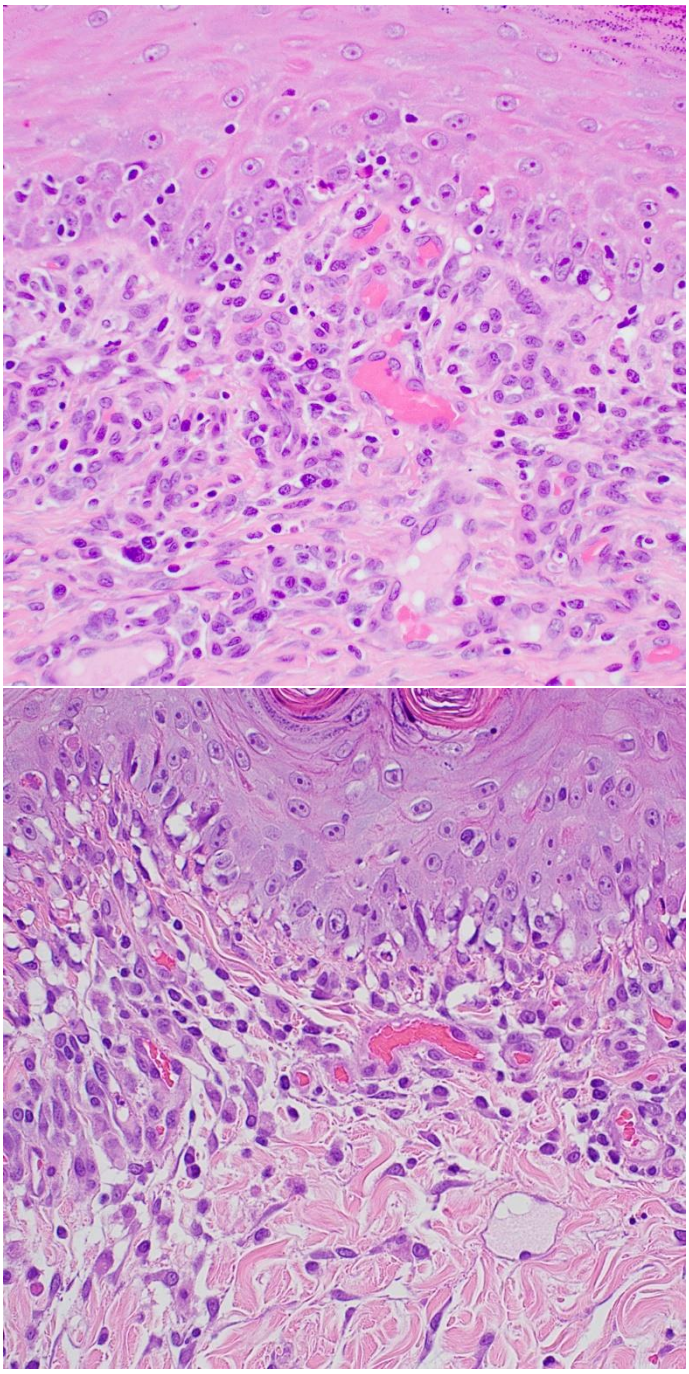
10-year-old male castrated Shetland Sheepdog with erosive to ulcerative and crusting lesions on the ventral abdomen, periocular and perioral skin.

Clinical history: The patient was presented for a second opinion following three months of progressive pyoderma and has been treated with cephalexin, prednisone and oclacitinib. A partial response was achieved initially, but upon discontinuation of medications lesions rapidly progressed. The patient was also being treated with L-thyroxine by the referring veterinarian (dose unknown). Punch biopsies were obtained from the ventral abdomen.







Figures 1-4

What is the most likely diagnosis?

A. Systemic lupus erythematosus
B. Vesicular cutaneous lupus erythematosus
C. Erythema multiforme
D. Bullous pemphigoid

Answer:

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Signalment: 10-year-old male castrated Shetland Sheepdog with erosive to ulcerative and crusting lesions on the ventral abdomen, periocular and perioral skin.

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Histopathologic Description:

Two biopsy samples from the caudal abdomen are similar. The epidermis is hyperplastic and acanthotic, subtended by a lichenoid-interface infiltrate of small lymphocytes admixed with fewer plasma cells. There is frequent basal cell apoptosis associated with lymphocyte exocytosis. Transepidermal apoptosis is not identified. There is vacuolation of the stratum basale with dermoepidermal separation at the edge of the ulcers and the edge of the biopsy samples. Centrally, ulcers are covered by a thick exudative crust of fibrin, serum, and cell debris. The crusts contains numerous colonies of cocci. There is also a mild perivascular infiltrate of lymphocytes, plasma cells and neutrophils. Apocrine glands are also inflamed.

Morphologic Diagnosis: LYMPHOPLASMACYTIC LICHENOID INTERFACE DERMATITIS WITH DERMOEPIDERMAL CLEFTS, ULCERS, AND CRUSTS CONTAINING NUMEROUS COCCI

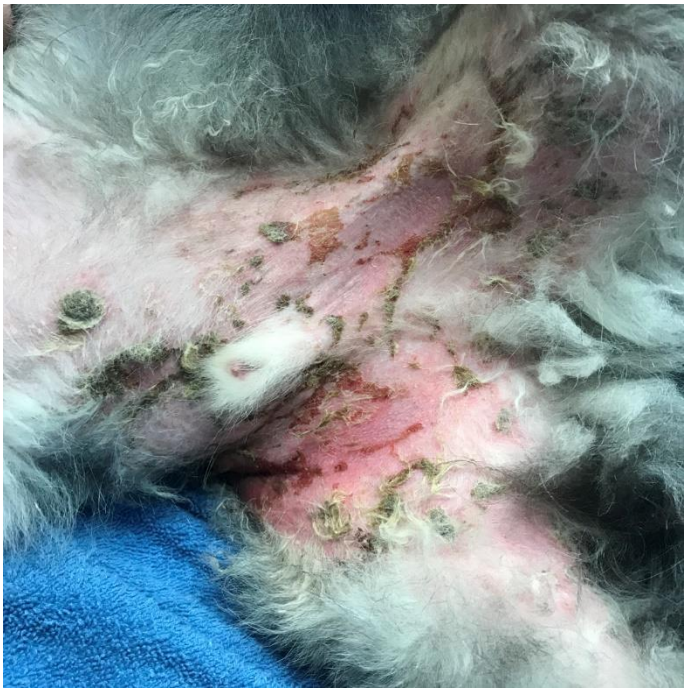

Figure 5. Lesions at presentation: Erosions, ulcers and crusts along the ventrum and inguinal region. Some are annular to polycyclic to serpiginous

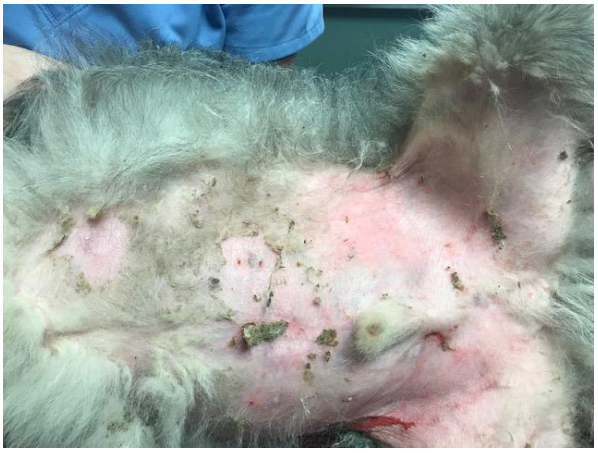

Figure 6 and 7: Following 21 days of treatment with immunosuppressive dose of prednisone (2mg/kg/day) and culture directed antimicrobial therapy erosions and ulcers are drier, less red and improved. Topical tacrolimus was applied to a remaining erosion on the right fold. Lesions continued to resolve and patient did well for 3 months of follow up at which time the patient died and a pulmonary neoplasm was identified with post-mortem radiographs.

Comment:

This is a case of vesicular cutaneous lupus erythematosus (VCLE) of Shetland sheepdogs and collies. There can be close histopathologic overlap with an erythema-multiforme reaction pattern, which is

considered as a possible, but less likely differential. In VCLE, cytotoxic interface dermatitis tends to be more targeted at the basal cell layer. The signalment and clinical history are also supportive. Often, at the time of biopsy, vesicles have ruptured leading to ulcers, as in this case.

VCLE is auto-immune disease, with some homology to subacute cutaneous lupus erythematosus in humans. (1, 2) Adult dogs are affected, predominately in the summer, with lesions distributed over more sparsely haired areas of the body. Mucous membranes may be rarely involved. (3)

Immunopathologic characterization of the infiltrate has confirmed the presence of CD8 and CD4 positive T-cells and CD1+ antigen presenting dendritic cells. Apoptosis of basal keratinocytes has also been confirmed by TUNEL staining. Jackson et al found 9/11 dogs had positive IgG autoantibodies to ENA (extractable nuclear antigens) via ELISA and Immunoblotting (1).

A proposed pathogenesis includes UV radiation induced apoptosis of keratinocytes that express nuclear proteins on the cell surface. Auto-antibodies that develop as a consequence of this expression mediate further apoptosis by antibody dependent cytotoxicity. Circulating antibasement membrane IgG, IgA, and IgM and serum IgG antinuclear antibodies (1:64) have not been detected. Therapeutic options include sun avoidance, immunomodulatory drugs including glucocorticoids, nicotinamide-tetracycline and calcineurin inhibitors. (3-6) Response is variable, with some dogs achieving complete or long term remission, however some dogs have been euthanized as a consequence of their disease in initial case series. (7, 8)

Case provided by: Dr. Charles Bradley, University of Pennsylvania, School of Veterinary Medicine, PA USA and Dr. Joseph Mastroianni, Wallenpaupack Veterinary Clinic, Hawley, PA, USA.

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