4-year-old collie mix female dog with bilateral, non-symmetric alopecia, scarring and hypopigmentation on the dorsum of the nose. Which of the following is the most likely disease?

1. Systemic lupus erythematosus
2. Thermal burn
3. Pemphigus foliaceus
4. Erythema multiforme
5. Ischemic dermatopathy CORRECT

**Signalment and history:** A 4-year-old collie mix female dog was examined for 2.5-year duration of a bilateral, non-symmetric alopecic lesion with scarring and hypopigmentation on the dorsal skin of the nose (Figure 1). The lesion involves the nasal planum where it is associated with multifocal erosions; exfoliative and erosive lesions are also present on the footpad and cicatritial and alopecic lesions are present on the tip of the ears. The dogs is also affected by an eosinophilic colitis and it is under a hypoallergenic diet.

**Histopathologic description:** The histopathological examination of the biopsy from the dorsum of the nose revealed the presence of a severe and diffuse follicular atrophy with a lesser involvement of the sebaceous glands. Dermal collagen appears to be rarefied and pale eosinophilic and homogeneous (Figure 2 and 3). Multifocal degeneration of basal keratinocytes is also observed, sometimes responsible for artefactual detachment at epidermal dermoepidermal junction. In the basal cell layer are multifocal apoptotic cells and the dermo-epidermal interface is obscured by a cell-poor infiltrate consisting of lymphocytes, histiocytes and rare mast cells, plasma cells and neutrophils. Small caliber vessels in the superficial dermis sometimes have hypereosynophilic walls without endothelial cell lining (Figure 4). In the superficial dermis pigment incontinence is observed in association with a lymphocytic and histiocytic mural folliculitis. In some areas there is a periadnexal lymphocytic infiltrate with multifocal deep extension and involvement of muscle fibers causing myositis (Figure 5).

**Morphologic diagnosis:** "Cell-poor" lymphocytic interface dermatitis and folliculitis with follicular atrophy, vasculopathy (cell poor vasculitis) and myositis.

**Name the condition:** Ischemic dermatopathy – Dermatomyositis (d.d. Dermatomyositis-like disease).

**Comment:** The histopathological lesions in the biopsy examined are suggestive of an ischemic dermatopathy which, for the breed reported (collie mixed) and for the early age onset, are compatible with a canine familial dermamyositis (dd ischemic dermatopathy with early onset and no breed predisposition, dermamyositis-like disease). Canine familial dermamyositis is an uncommon hereditary disease of collies, Shetland sheepdogs, Beauceron shepherds, Belgian Tervurens and Portuguese water dogs. A similar disease, called dermamyositis-like disease, has been described in the Pembroke Welsh corgi, Lakeland terrier, chow chow, Jack Russell terrier, German shepherd dog, Kuvasz, Rottweiler and in mongrels and has been recognized in other purebred dogs. The disease is characterized by skin lesions and generalized myositis, with early onset between 7 weeks and 6 months of age. The skin lesions are located on the face, ears, lips, tail tip, and bony prominences of the distal legs. In the early phase, there is erythema, scaling, crusting, and ulceration. Chronically affected dogs develop alopecia and scarring, with hyper- or hypopigmentation. Myositis usually affects the masticatory muscles and muscles of the extremities below the elbow and stifle. Although muscle lesions are rare, when present, they can be the most severe clinical presentation of dermamyositis. Severely affected dogs may have generalized myositis and develop megasophasus. The cause of the disease is poorly understood. Elevated serum complement levels and class G immunoglobulins have been demonstrated, therefore a genetically determined immune-mediated pathogenesis is suspected. Inheritance studies of dermamyositis in the collie suggest that it may be an autosomal dominant trait with incomplete penetrance. In Shetland Sheepdog, the locus affecting the dermamyositis phenotype seems to be located near marker FH3570 in chromosome 35. A gene expression profile study investigating affected and normal skin of dogs with dermamyositis demonstrated...
a differential regulation of 285 genes and an absence of disease-specific autoantibodies in dermatomyositis affected dogs. A very recent study, through whole genome resequencing, identified primary candidate polymorphisms in conserved regions of PAN2 and MAP3K7CL on chromosomes 10 and 31, respectively, in Collies and Shetland Sheepdogs. These two loci, in association with MHC haplotypes, were associated with an increased risk of disease development. Histological examination demonstrates a cell-poor lymphocytic or lympho-histiocytic interface dermatitis and folliculitis with hydropic degeneration and apoptosis of basal keratinocytes. The dermis is homogenous and pale and hair follicles are atrophic. Blood vessel show features of cell-poor vasculopathy. All these changes were grouped together by Gross TL et al. under the term ‘ischaemic dermatopathy’. This histological pattern is common to several different clinical pictures: 1. canine familial dermatomyositis; 2. juvenile-onset ischemic dermatopathy with no breed predilection (dermatomyositis-like disease); 3. postrabies vaccination panniculitis; 4. generalized vaccine-induced ischaemic dermatopathy; and 5. adult-onset (nonvaccine-induced) generalized ischaemic dermatopathy (“generalized idiopathic ischemic dermatopathy”). The present case and the concurrent presence of histologic evidence of myositis, in association with the breed (collie mix), is suggestive of a canine familial dermatomyositis. However, as information on the hereditability and familiarity of skin lesions in this animal are unknown, a dermatomyositis-like disease has to be considered as a differential diagnosis.

References:


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Figure legend:

**Figure 1:** 4-year-old collie mix female dog. Bilateral, non-symmetric alopecia with scarring and hypopigmentation on the dorsal skin of the nose with involvement and multifocal erosions of the nasal planum.

**Figure 2:** 4-year-old collie mix female dog. Diffuse and moderate to severe follicular atrophy with homogeneization and pallor of dermal collagen.
Figure 3. 4-year-old collie mix female dog. Sever follicular atrophy with increased prominence of perifollicular connective tissue and multifocal periadnexal lymphocytic infiltration.

Figure 4. 4-year-old collie mix female dog. Obscuration of the dermal-epidermal junction by a cell-poor lymphocytic infiltration, with basal cell degeneration and apoptosis. There is loss of endothelial cells and mummification of vessel walls in superficial dermal vessels (insert).
Figure 5. 4-year-old collie mix female dog. In the deep portion of the biopsy there are perimysial lymphocytic and histiocytic infiltrations associated with myocyte fiber degeneration and phagocytosis.