

CLINICAL, HISTOLOGICAL AND IMMUNOLOGICAL CHARACTERISTICS OF NEUMANN-TYPE PEMPHIGUS VEGETANS IN A DOG

Marianne Heimann⁰, Luc Beco¹, Michel Petein⁰, Vincent Rademaeker² Thierry Olivry³.

⁰Institut de Pathologie et de Génétique, Loverval, Belgium

¹Cabinet Vétérinaire Reine Astrid, Spa, Belgium;

²Cabinet Vétérinaire, Montzen, Belgium

³Department of Clinical Sciences, College of Veterinary Medicine, NC State University, Raleigh, North Carolina, USA

Pemphigus vegetans is a very rare cutaneous autoimmune blistering acantholytic disease of humans that combines features of both pemphigus foliaceus and mucosal lesions of pemphigus vulgaris. We report the clinical, histopathological and immunological findings in a dog whose lesions resembled those of pemphigus vegetans of humans

SIGNALMENT 4 year old male greater Swiss mountain dog

HISTORY

Three months duration of mildly pruritic dermatitis that was characterized by crusts distributed over the right axilla and inguinal regions. Over the two following months, the dermatitis progressed and involved the trunk and ear pinnae. Anal, prepuce and oral mucosae developed erosive lesions.

CLINICAL FINDINGS

The dog is in good general condition with a mild hyperthermia 39,8°C.

DERMATOLOGICAL FINDINGS

The dermatological lesions were generalized but more severe over the head, the ear pinna, the ventral part of the thorax, the groin and the axillae. Lesions consisted of diffuse generalized erythema, scaling, follicular casts, hyperkeratotic papules and verrucous plaques. Papules, crusts, erosions and ulcerations covered the ear pinnae (Fig 1). The ventral aspect of the feet was erythematous. A mild purulent exudate was observed in the ear canals. Erosions and ulcerations were observed on the anal mucosae, the prepuce and the hard palate (Figs 2, 3, 4). Small vesicles were seen on the lips.



Fig 1: Ear pinna



Fig 2: Anal mucosae



Fig 4: hard palate



Fig 3: prepuce (see also the cutaneous verrucous plaques, arrows)

HISTOPATHOLOGY

Histopathology of cutaneous lesions consisted of a mixed pattern of superficial and deep intra-epidermal neutrophilic and eosinophilic pustules with isolated and clustered acantholytic keratinocytes (Figs 5, 6). Additionally, suprabasal epidermal clefts leaving rounded basal keratinocytes at the bottom of the vesicles were discovered (Figs 7, 8). These dual acantholytic changes were observed also within hair follicle epithelium (Fig 6). Dermal inflammation was mixed and perivascular.

IMMUNOLOGICAL FINDING

Direct immunofluorescence:

Direct immunofluorescence revealed intercellular IgG at all epidermal layers (strongest in the stratum spinosum) multifocal intracellular keratinocyte fluorescence and multifocal basement membrane fluorescence. Specific IgA, IgM and C3 deposits were not observed

Indirect immunofluorescence :

This method revealed a very high titer of IgG binding to keratinocyte membranes at all levels of several stratified epithelial substrates. Titers varied from 1:2,500 to 1:10,000 depending upon the substrate used.

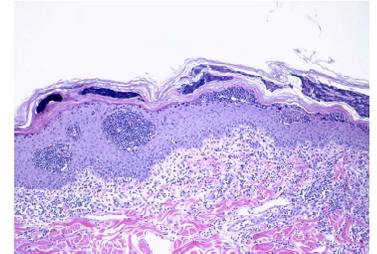


Fig.5: deep and superficial Acantholytic pustules (HE x 100)

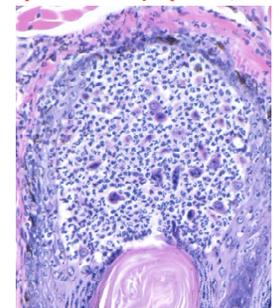


Fig.6: Acantholytic pustule (HE x 400)

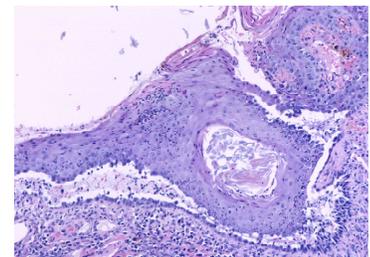


Fig7 : Suprabasilar cleft (HE x 250)

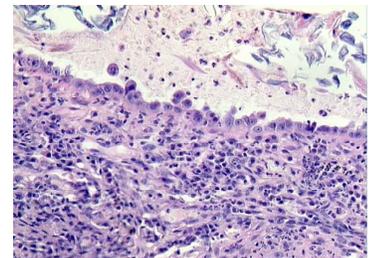


Fig. 8: Suprabasilar cleft (HE x 630)

DISCUSSION :

The disease described herein met all criteria used for the diagnosis of pemphigus vegetans (Neumann-type) in humans: 1) **Clinical Signs**: presence of oral non infectious erosions, cutaneous flaccid blisters that erode easily with progressive intractable erosions and crusts; 2) **Histopathology**: suprabasal clefts and intraepidermal pustules with acantholysis; 3) **Immunofluorescence**: presence of skin-fixed and circulating IgG auto-antibodies that recognize epidermal keratinocyte membranes.

In conclusion, canine pemphigus vegetans (Neumann-type) is a rare variant of pemphigus that can be added to the four other variants identified in this species: pemphigus vulgaris (1975), pemphigus foliaceus (1977), pemphigus erythematosus (1980) and paraneoplastic pemphigus (1998).