A 3-year-old female Akita Inu with acute onset of pruritic erosions, ulcerations and crusts on the nasal planum, around the eyes and on the vulva. The dog has eye-problems since two years. Which of the following is the most likely diagnosis?

A. Atopic dermatitis  
B. Uveodermatologic syndrome (Vogt-Koyanagi-Harada-like syndrome)  
C. Mucocutaneous lupus  
D. Epidermolysis bullosa acquisita

Signalment and history: A 3-year-old, female intact, completely white, Akita Inu was referred for acute onset of pruritic skin lesions. The dog has suffered already during the last two years from generalized pruritus, but without clearly noticeable skin lesions. In addition the dog has also a long history of conjunctivitis, affecting both eyes, and chronic uveitis and glaucoma in the left eye. Recently the owner reported difficulties in seeing. The dog has been seen during this 2-years-period by two different ophthalmologists and the given possible causes for the eye problems were infectious or autoimmune. One week prior to the visit of a dermatologist the owner noted progressing skin lesions associated with pruritus. At the time of the dermatological consultation the dog was on oclacitinib 0.5 mg/kg orally once daily to manage the pruritus and the suspected autoimmune skin disease, and dorzolamide hydrochloride-timolol maleate ophthalmic solution twice daily in both eyes. The dog has been regularly vaccinated and never travelled abroad. The general examination of the dog revealed difficulties in seeing, otherwise the general condition was within normal limits. The dermatological exam revealed erosions, ulcerations and crusts on the nasal planum and around the eyes. Crusts were also present on the lips and around the paw pads. In addition ulcerations and crusts were seen in the perivulvar area (Figures 1, 2 and 3). Cytology from the ulcerative lesions showed neutrophils, macrophages and cocci. The differential diagnoses were uveodermatologic syndrome (Vogt-Koyanagi-Harada-like syndrome), discoid lupus erythematosus, systemic lupus erythematosus, pemphigus foliaceus, pemphigus erythematosus and epitheliotropic lymphoma. During the sedation hematology and serum biochemistry profile were tested and were unremarkable. Several 6mm punch biopsies were taken from the lesion on the nasal planum, the paw pads, the perilabial and the perivulvar skin.

Histopathologic description:  
In all biopsies (Figures 4, 5, 6 and 7) there is lichenoid infiltrate immediately subjacent to the epidermis. This infiltrate is severe in the biopsies from the nose and the foot pad and mild to moderate in the biopsies from the vulva and the lip. Multifocally the dermo-epidermal junction is obscured and the inflammatory cells are present also in the basal cell layer. Rarely apoptotic cells are present in the basal cell layer. Inflammatory cells consist of large, often pale macrophages and fewer neutrophils, lymphocytes, and plasma cells. Macrophages occasionally contain very fine, dust-like, granular melanin pigment. There is mild to moderate dermal edema. Biopsies from the nose, the vulva and the lip are covered multifocally by serocellular crusts which contain occasional bacterial colonies. In the areas of crusting transepidermal exocytosis of neutrophils is present. In addition, ulceration is seen in the biopsy of the nasal planum.
Morphologic diagnosis: severe, diffuse histiocytic and lymphocytic, lichenoid interface-dermatitis with pigmentary incontinence

Name the condition: uveodermatologic syndrome (Vogt-Koyanagi-Harada-like syndrome)

Follow up: While waiting for the results of the skin biopsies the dog received prednisolone 1.5 mg/kg orally once a day and chlorambucil 0.15 mg/kg orally once a day. After two weeks of administration the skin lesions improved significantly. Repeated hematology and serum biochemistry remained unremarkable. The recheck by the ophthalmologist showed an improvement of the eye problems.

Comment: Uveodermatologic syndrome (Vogt–Koyanagi–Harada-like syndrome) is a rare canine disease of probable autoimmune etiology. It is characterized by concurrent granulomatous uveitis and depigmenting skin disease. The Vogt-Koyanagi-Harada syndrome is a human autoimmune disease, that can affect pigmented tissues of several organs. The pathomechanism of these two diseases has been postulated to be similar and based on cell-mediated hypersensitivity and antibodies directed against melanin and related proteins. In humans, there is a meningoencephalitic phase that precedes uveitis and deafness. Only in the end the patient develops skin lesions. In dogs, ophthalmic disease and dermatologic disease both commonly occur, although skin lesions usually occur later. Interestingly, clinical signs compatible with a meningoencephalitic phase are only rarely seen in dogs. Also in the presented case ocular problems developed earlier than the skin lesions. Interestingly, for both ophthalmologists, who saw the dog, the ocular signs were not definitively convincing to give a diagnosis of uveodermatologic syndrome and to start the correct treatment. One possible reason for this might be that the dog presented only with pruritus but no other obvious skin lesions. It is well possible that the initial signs of VKH skin lesions, namely the depigmentation were not obvious in this dog since the dog is completely white. Interestingly, histology revealed nevertheless in some biopsies pigmentary incontinence. As a conclusion an uveodermatologic syndrome should be considered in dogs with eye problems even if the dogs do not present with typical skin lesions especially if the dog belongs to a predisposed breed, like the Akita Inu.

References:


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FIGURES

Figure 1.