

# Cutaneous mucinosis and mastocytosis in a shar-pei

Alfonso López, Donna Spracklin, Sandra McConkey, Paul Hanna

**Abstract** — A 7-year-old shar-pei was presented because of a recurrent dermatologic condition. Skin biopsies revealed an idiopathic (primary) cutaneous mucinosis that initially responded to corticosteroids. The condition reappeared 2 years later and subsequent biopsies revealed a mast cell tumor in some of the skin sites previously diagnosed with mucinosis.

**Résumé** — Mucinose cutanée et mastocytose chez un Shar-pei. Un Shar-pei de 7 ans a été présenté suite à une affection dermatologique récurrente. Des biopsies de la peau ont révélé une mucinose cutanée idiopathique (primaire) qui au départ répondait aux corticostéroïdes. L'affection est réapparue 2 ans plus tard et des biopsies subséquentes ont révélés une tumeur mastocytaire dans certains des sites cutanés où on avait auparavant diagnostiqué une mucinose.

(Traduit par docteur André Blouin)

Can Vet J 1999; 40: 881–883

A 7-year-old, spayed, female shar-pei was presented to the veterinarian for recurrence of a dermatologic condition. Three years previously, this dog had numerous soft bullae, some of which were ulcerated, on both shoulders, axillae, and the left hock. Idiopathic mucinosis was diagnosed from biopsies taken from affected skin. Treatment at that time included intermittent low doses of methylprednisolone acetate (Medrol, Pharmacia & Upjohn, Don Mills, Ontario), 0.3 mg/kg body weight (BW), PO, q24h, and cephalexin (Novo-Lexin, Novopharm, Toronto, Ontario), 26 mg/kg BW, PO, q12h, which temporarily controlled the problem. The cutaneous condition reappeared 2 y later, and it involved more than 30% of the skin surface and was particularly severe over the ventral abdomen and thighs. Both hind limbs, from the hock to the hip, were grossly swollen, while the forelimbs and thorax were spared. Physical examination revealed marked thickening of the skin with varying degrees of alopecia, hyperpigmentation, thinning of the epidermis, and focal ulceration (Figure 1). Palpation of affected areas revealed multifocal (0.5–5.0 cm) subcutaneous swellings, due to soft fluctuating fluid, which under slight pressure oozed mucinous fluid onto the skin surface.

Skin scrapings for mites were negative and bacterial culture from an ulcerated nodular lesion yielded coagulase-negative *Staphylococcus* and *Enterococcus*. Routine hematological examination and serum chemistry analysis yielded normal values, except for a mildly decreased thyroxine (T4) concentration of 13.8 nmol/L (reference 15 to 52 nmol/L). Treatment with thyroid replacement hormone and an appropriate antibiotic for several months failed to improve the skin condition.

Department of Pathology and Microbiology, Atlantic Veterinary College, University of Prince Edward Island, 550 University Avenue, Charlottetown, Prince Edward Island C1A 4P3 (López, McConkey, Hanna) and Fundy Veterinarians, Truro, Nova Scotia (Spracklin).

Address correspondence and reprint requests to Dr. Alfonso López; e-mail: lopez@upej.ca.

Unstained impression smears and additional skin biopsies fixed in formalin were submitted to the diagnostic laboratory at the Atlantic Veterinary College.

On microscopic examination, the impression smears contained moderate numbers of erythrocytes and nucleated cells streaming in a background of fine pink stippling, which sometimes appeared fibrillar. The nucleated cells were comprised of approximately 20% eosinophils, 40% nongranulated spindle cells, and 40% well-granulated mast cells (Figure 2). Low numbers of neutrophils were also seen. The mast cells showed marked anisocytosis (15 to 40  $\mu\text{m}$  in diameter) and were occasionally binucleated. Nuclei were round or oval with granular chromatin and 1 to 4 nucleoli. The spindle cells were approximately 20 to 60  $\mu\text{m}$  in length. These nongranulated cells had single, central, round nuclei; granular chromatin; and, sometimes, a single nucleolus. The smears were compatible with a mast cell tumor and the pink background, although nonspecific, was suggestive of mucinous material, consistent with cutaneous mucinosis.

At the time when 6 biopsies taken from thighs, abdomen, flank, and shoulder were trimmed, skin samples were found to be covered with abundant mucinous material. Microscopically, the epidermis was moderately hyperplastic with focal areas of ulceration surrounded by neutrophils. The dermal architecture was conspicuously disrupted due to severe separation of collagen fibers by abundant pale basophilic, mucinoid, alcian blue-positive ground substance. The dermis was also diffusely infiltrated by some eosinophils and large numbers of round cells with morphologic features consistent with mast cells. Toluidine blue-staining revealed faintly positive metachromatic granules in the cytoplasm of these cells. Based on the cytological and histological findings, we added cutaneous mastocytosis to the previous diagnosis of cutaneous mucinosis.

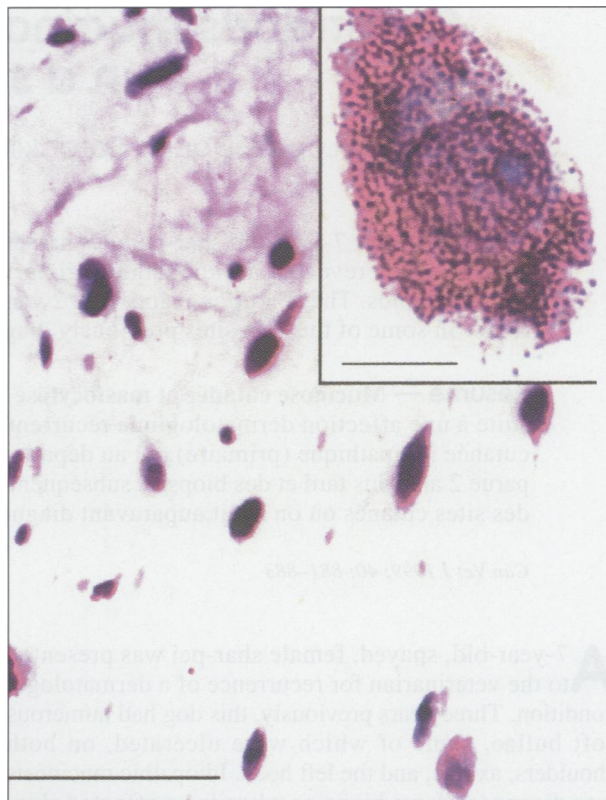
The dog was treated with immunosuppressive doses of methylprednisolone acetate (Medrol, Pharmacia & Upjohn), 1 mg/kg BW, PO, q12h, and azathioprine (Immunan, GlaxoWellcome, Mississauga, Ontario), 2 mg/kg BW, PO, s.i.d. Response to treatment was



**Figure 1.** A 7-year-old, spayed female shar-pei with cutaneous mucinosis and mastocytosis. Note severe, nodular swelling of the skin with diffuse alopecia and locally extensive areas of ulceration.

slow and gradual; after several months, the subcutaneous swelling was reduced by 90% and all the ulcers had healed, but the epidermis remained hairless. Four weeks later, while still receiving prednisone, 0.5 mg/kg BW, PO, q48h, and azathioprine, 2 mg/kg BW, PO, q48h, a firm ulcerated nodular mass erupted on the left thigh, and the left inguinal lymph node became noticeably enlarged. The dosage of azathioprine was increased to 2 mg/kg BW daily; however, the ulcerated areas on the lateral thigh continued to worsen, involving a surface area of more than 20 cm in diameter. The tumoral mass was now 8 cm in diameter and had a large necrotic core. The owners declined further treatment and requested euthanasia.

Postmortem examination revealed enlarged inguinal lymph nodes, but no evidence of tumoral metastasis to the internal organs. Fixed samples of skin, lymph nodes, lung, liver, kidneys, spleen, adrenal glands, and intestine were submitted for histopathological examination. Microscopic findings corroborated the previous diagnosis of cutaneous mucinosis and cutaneous mastocytosis. Except for the skin and inguinal lymph nodes, no other organs or tissues showed microscopic evidence of neoplastic mast cells. The inguinal lymph nodes were massively infiltrated with neoplastic mast cells and also contained focal areas of necrosis. Affected skin showed



**Figure 2.** Impression smear taken from affected skin (Wright-Giemsa stain). Note a homogenous background of mucin containing many round cells; bar = 100  $\mu$ m. Insert: mast cell with abundant cytoplasmic granules; bar = 10  $\mu$ m.

marked dermal expansion and effacement associated with edema, mucin, and large numbers of mast cells with fewer eosinophils. The mast cells had varying amounts of poorly delineated and well-granulated cytoplasm, in which the granules stained metachromatically with toluidine blue. The nuclei were round or oval with granular chromatin and had single or multiple nucleoli ranging from 5 to 12  $\mu$ m in diameter; mitotic figures were frequently observed (present in most random high power fields). Microscopic features were consistent with a grade 3 mast cell tumor.

Primary cutaneous mucinosis resulting from an excessive accumulation of mucinous material in the dermis is a rare degenerative skin condition in dogs, except for the shar-pei (1,2). Cutaneous mucinosis is clinically characterized by thickening of the skin, which, in severe cases, may cause gross deformity of affected tissues (1,3). It is also common for dermal mucin to form epidermal vesicles that eventually rupture and ooze viscous material onto the surface of the skin. The clinical diagnosis of cutaneous mucinosis can be confirmed easily by skin biopsies, which typically show a pale, basophilic, alcian blue-positive ground substance displacing collagen fibers and causing a variable degree of dermal atrophy (1,3). Cutaneous mucinosis is so common in the shar-pei that some authors consider the condition normal in this breed (1,4). In other canine breeds, cutaneous mucinosis is rare and generally occurs secondary to diseases such

as hypothyroidism (myxedema), lupus erythematosus, or neoplasia (1,5,6).

Mastocytosis is a generic term used to describe an assorted group of reactive or neoplastic diseases characterized by an abnormal accumulation of mast cells in tissues (1,3,7). Reactive mastocytosis occurs in response to an inflammatory reaction, such as parasitism, while neoplastic mastocytosis does not have a specific etiology and constitutes one of the most common cutaneous neoplasms of dogs and cats (1,8,9). According to the distribution of mast cells, mastocytoses are classified as systemic when there are many organs involved, or localized when the infiltration of mast cells is restricted to a particular tissue, such as the skin in cutaneous mastocytosis (3,7,10).

In the case reported here, gross and microscopic examination proved that the mast cell infiltration exclusively involved the skin and regional lymph nodes, thus falling into the category of cutaneous mastocytosis. Similar changes are referred to in humans as "diffuse cutaneous mastocytosis" (11).

It has been reported that the extremities and the inguinal and perineal areas are preferred sites for malignant mast cell tumors in some breed of dogs, including the shar-pei (12). Our findings are consistent with this report, in which the inguinal lymph nodes were the only tissues showing evidence of metastasis. The microscopic findings seen in the different skin biopsies taken from this dog suggest that cutaneous mucinosis was the first dermatologic problem, which was followed 2 y later by the eruption of a locally invasive mast cell tumor.

What made this case unusual was the fact that cutaneous mastocytosis appeared in the skin sites previously diagnosed as affected by idiopathic dermal mucinosis. Interestingly, a similar case was reported in a shar-pei in which swelling and deformity of the distal limbs, presumed to be the result of cutaneous mucinosis, failed to respond to corticosteroid therapy (13).

In conclusion, cases of cutaneous mucinosis in shar-pei dogs unresponsive to treatment should be investigated for a coexisting cutaneous mastocytosis. Cytological or microscopic examination of biopsy material can be readily used to confirm the diagnosis (1,3).

### Acknowledgments

The authors thank David Sims, Carolyn Lariviere, and Julio Martínez for assistance with the digital photography.

CV


### References

1. Yager JA, Wilcock BP. Surgical Pathology of the Dog and Cat. Dermatopathology and Skin Tumors. London: Mosby, 1994: 30-38.
2. Bombhard von D, Kraft W. Idiopathic mucinosis cutis in Chinese Shar pei dogs: Epidemiology, clinical features, histopathologic findings and treatment. Tierärztl Prax Ausg K Klientiere Heimtiere 1998; 26: 189-196.

3. Gross TL, Ihrke PJ, Walder EJ. Veterinary dermatopathology: A macroscopic and microscopic evaluation of canine and feline skin diseases. Toronto: Mosby, 1992: 228-229.
4. Dunstan RW, Rosser EJ, Jr. Newly recognized and emerging genodermatoses in domestic animals. Curr Probl Dermatol 1987; 17: 216-235.
5. Doliger S, Delverdier M, More J, Longeart L, Regnier A, Magnol JP. Histochemical study of cutaneous mucins in hypothyroid dogs. Vet Pathol 1995; 32: 628-634.
6. Dillberger JE, Altman NH. Focal mucinosis in dogs: seven cases and review of cutaneous mucinosis of man and animals. Vet Pathol 1986; 23: 132-139.
7. O'Keefe DA, Couto CG, Burke-Schwartz C, Jacobs RM. Systemic mastocytosis in 16 dogs. J Vet Intern Med 1987; 1: 75-80.
8. Davis BJ, Page R, Sannes PL, Meuten DJ. Cutaneous mastocytosis in a dog. Vet Pathol 1992; 29: 363-365.
9. Moulton JE. Tumors of the urinary system. In: Moulton JE, ed. Tumors of Domestic Animals, 2nd ed. Berkeley: Univ California Pr, 1978: 288-308.
10. Valent P. Biology, classification and treatment of human mastocytosis. Wien Klin Wochenschr 1996; 108: 385-397.
11. Mackey S, Pride HB, Tyler WB. Diffuse cutaneous mastocytosis. Treatment with oral psoralen plus UV-A. Arch Dermatol 1996; 132: 1429-1430.
12. Miller DM. The occurrence of mast cell tumors in young Shar-peis. J Vet Diagn Invest 1995; 7: 360-363.
13. Madewell BR, Akita GY, Vogel P. Cutaneous mastocytosis and mucinosis with gross deformity in a Shar-pei dog. Vet Dermatol 1992; 3: 171-175.

## LEBA III

**THE ONLY PRODUCT PROVEN TO CLEAN TEETH WITHOUT BRUSHING**



- Not a toothpaste
- Easier than brushing
- Herbal product, causes no enamel damage and no side effects
- Ideal for home dental program
- Excellent for maintenance after a periodontal cleaning

**LEBALAB INC.** Available to your veterinarian from:  
Midwest Vet. Drug Co-Op, WDDC,  
CDMV and Vet Purchasing

Suite 261 2-1200 London Rd. Sarnia, Ontario  
Tel: (519) 542-3165 Fax: (519) 542-5196