Hereditary Pyogranuloma and Vasculitis of the Nasal Plane in Scottish Terriers: Two New Cases from Argentina and the United States

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Abstract: Hereditary pyogranuloma and vasculitis of the nasal plane was diagnosed in two young Scottish terriers: one from Argentina and one from the United States. To the authors’ knowledge, this is the first report of this condition occurring outside of Denmark. In one dog, there was a good response to the concurrent oral administration of prednisolone and cyclosporine.

Key words: nasal plane, pyogranuloma and vasculitis, Scottish terrier

Introduction

There are few inflammatory dermatoses that can be isolated to the nasal plane (planum nasale) which include discoid lupus erythematosus, pemphigus erythematosus, pemphigus foliaceus, pemphigus vulgaris, adverse cutaneous drug reaction, hereditary nasal parakeratosis (Labrador retrievers), and dermal arteritis of the nasal philtrum (predominantly in St. Bernards). In 1991, a unique, presumably hereditary pyogranulomatous and vasculitic disorder of the nasal plane, nostrils, and nasal mucosa was reported in five young Scottish terriers in Denmark. To the authors’ knowledge, this condition has not been reported since the original description.

The purpose of this article is to describe two more cases of this apparently very rare genodermatosis of Scottish terriers seen in both North and South America.
Case Reports

Case 1

A two-month-old, male Scottish terrier was presented to the School of Veterinary Medicine at the University of Buenos Aires (UBA) for an ulcerative condition involving the nasal plane. According to the owner, the condition was first noticed 15 days earlier. The puppy initially exhibited bilateral mucoid nasal discharge, sneezing, and two small ulcers on the nasal plane. The attending veterinarian prescribed a 10-day course of cephalaxin, which was of no benefit. The ulcerative process continued to worsen.

On presentation at the UBA, the puppy was in good health except for the muzzle. The ulcerative process now extended from the rostral, lateral, and ventral aspects of the nasal plane to the upper lips (Figs. 1 and 2). The normal anatomy in these areas was effaced, including partial destruction of alar cartilage and nasal septum. The ulcerative surface was oozing a serosanguineous material and multifocally crusted. The condition was apparently nonpruritic and nonpainful.

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Routine hemogram and serum biochemistry results were unremarkable except for a slight leukocytosis (15,000/mm$^3$; normal range 6,000 to 10,000/mm$^3$; 90% neutrophils). Cytological findings in a direct smear included numerous neutrophils and fewer macrophages, fibrocytes, and erythrocytes. Microorganisms were not seen. Routine radiographs of the nasal cavity revealed no significant findings. Biopsy specimens were submitted for histopathologic evaluation. As microorganisms were not seen in cytological preparations, the puppy was sent home on: (1) daily cleaning of the ulcerated area with a 5% chlorhexidine solution, and (2) prednisolone administered orally at 0.5 mg/kg once daily.

Histopathological examination revealed diffuse pyogranulomatous inflammation with surface ulceration and necrosis. Numerous blood vessels within the pyogranulomatous reaction exhibited leukocytoclastic vasculitis (Fig. 3). Special stains were negative for the presence of microorganisms.
The final clinicopathological diagnosis was hereditary pyogranuloma and leukocytoclastic vasculitis of the nasal plane of Scottish terriers. The dog was re-examined after seven days of treatment, and the ulcerated area seemed to be less inflamed and less exudative. The dog was discharged on cyclosporine (5 mg/kg given orally, once daily), but was lost to follow-up.

Case 2
A 14-month-old, spayed female Scottish terrier was presented to the local veterinarian for an ulcerative condition of the nasal plane. According to the owner, the condition was first noticed when the dog was about 3 months old. The initial observations included bilateral nasal discharge, sneezing, and focal ulceration of the nasal plane. The condition had been slowly progressive despite courses of antibiotic therapy.

Examination by the local veterinarian revealed no abnormalities except for the nasal region. The rostral and ventral aspects of the nasal plane were completely ulcerated, with partial destruction of alar cartilage and nasal septum (Fig. 4). The condition was apparently nonpruritic and nonpainful. Results of routine hemogram, serum biochemistry, and urinalysis were unremarkable. Bacterial culture revealed a coagulase-positive Staphylococcus sp. and Streptococcus suis susceptible in vitro to numerous antibiotics. Fungal culture was negative.

Histopathological evaluation of biopsy specimens sent to the Diagnostic Laboratory at the College of Veterinary Medicine at Cornell University revealed pyogranulomatous inflammation and leukocytoclastic vasculitis (Figs. 5 and 6). No microorganisms were seen.

The final clinicopathological diagnosis was hereditary pyogranuloma and leukocytoclastic vasculitis of the nasal plane of Scottish terriers. Based on the results of culture and susceptibility testing, the dog was treated with cephalexin which resulted in minor improvement. One of the authors (DWS) discussed the histopathological findings and clinical diagnosis with the local veterinarian. Treatment options discussed included prednisolone, pentoxifylline, and cyclosporine. The dog was treated with prednisolone (2 mg/kg orally, every 24 hours) and cyclosporine (5 mg/kg orally, every 24 hours, with food).
Re-examination by the local veterinarian after one month of treatment revealed almost complete resolution of the ulceration. Depigmentation and scarring were prominent. For the next month, the cyclosporine was continued while the prednisolone dosage was gradually tapered. The dog is currently receiving only cyclosporine every 48 hours and is in remission.

Discussion

Hereditary pyogranuloma and vasculitis of the nasal plane is a distinctive disorder described only in young Scottish terriers. The condition usually commences with bilateral serous-to-mucoid nasal discharge in 3- to 6-week-old puppies. In the minority of cases, clinical signs are initially noted at 5 months of age. Sneezing, focal ulcers on the nasal plane, or both of these may also be present. Most affected dogs are otherwise healthy, although short periods of fever and depression have been reported. The sex was recorded for only four affected dogs: three females and one male.

As the disease progresses, ulcers enlarge and coalesce. The rostral and ventral aspects of the nasal plane, nasal philtrum, and nostrils are most severely affected. Ulcers are well-demarcated and have raised borders. Severely affected dogs suffer partial loss of alar cartilage and nasal septum. The ulcerative process can extend to the upper lips. Perhaps surprisingly, the condition does not appear to be painful or pruritic.

The etiopathogenesis of this syndrome is unknown. The exclusive occurrence in Scottish terriers, many of which were related, and the onset of clinical signs in very young dogs suggest a hereditary basis. In the original report, 50% of the puppies in two consecutive litters from the same sire and dam were affected, suggesting an autosomal dominant inheritance pattern. Detailed information on the siblings and parents of Case 1 reported herein was not available. For Case 2, the sire and dam were normal, and had not produced other litters. All 5 siblings of the affected dog were normal. The onset of clinical signs has not been associated with prior vaccination or drug administration, and infectious agents are not visualized in tissue specimens. The sterile pyogranulomatous inflammation and leukocytoclastic vasculitis are consistent with an immune-mediated pathogenesis.

Histopathological findings are characterized by a unique combination of pyogranulomatous inflammation and leukocytoclastic vasculitis. Histological examination of the nasal tissues of three 3-week-old puppies that were euthanized due to nasal discharge revealed that the characteristic histopathologic changes were already present in the nasal submucosa. The condition is apparently restricted to the nasal plane, nostrils, and nasal mucosa, as: (1) affected dogs typically show no signs of systemic involvement, and (2) no other tissues were involved in the one dog that was necropsied.

Therapeutic information on this syndrome is meager. Broad-spectrum antibiotics —whether administered empirically, or on the basis of culture and susceptibility testing— have not been effective. One dog from the original report received immunosuppressive doses of prednisone (2 mg/kg given orally, once daily) for 24 days, and the lesions resolved. Because of the vasculitic nature of the disease and a desire to reduce or eliminate the need for prednisone in a young dog, dapsone (1 mg/kg, given orally, every 8 hours) was added to the treatment protocol. Four weeks later, as the prednisone dose was being tapered, the condition relapsed and the dog was euthanized.

One of our dogs was treated with a combination of prednisolone and cyclosporine. This protocol was chosen because of the known immunomodulatory properties of these two agents. The condition was almost completely resolved after 30 days of treatment, and the dog is currently in remission on alternate-day cyclosporine.

Based on the short-term success with immunomodulatory treatments in only two dogs, it is possible that long-term control can be achieved. The topical administration of tacrolimus —another potent immunomodulatory compound— may also have a place in the treatment of this condition.

When the breeder of the dogs in the original report of this condition stopped using the involved sire and dams, no further cases were seen in the subsequent two years in Denmark. Indeed, to the authors’ knowledge, no further cases of this condition had been reported until 2008, when the two dogs reported herein were diagnosed in Argentina and the United States. It is clear that the genetic potential for this condition is still present in the Scottish terrier breed and far beyond the boundaries of Denmark.

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References