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Animal Model: The Mode of Inheritance of Craniomandibular Osteopathy in West Highland White Terrier Dogs

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Craniomandibular osteopathy is a disease of several breeds of dogs, principally West Highland White and Scottish terriers. It is characterized by a non-neoplastic proliferation of bone on the ramus of the mandible and/or the tympanic bulla. The disease in various respects resembles Paget's disease and infantile cortical hyperostosis of humans. A retrospective pedigree analysis of a kindred of West Highland White terriers was performed to determine if the trait was inherited and to determine mode of inheritance. This study indicated that in West Highland White terriers, the condition is an autosomal recessive trait.

Key words: autosomal recessive trait, animal model (canine), non-neoplastic proliferation of bone, Paget's disease, infantile cortical hyperostosis, craniomandibular osteopathy.

INTRODUCTION

Craniomandibular osteopathy (CMO) or osteoarthropathy, also known as lion jaw, westy jaw, or scotty jaw, is a disorder of dogs producing a bilateral irregular dense osseous proliferation on the mandible and/or tympanic bulla. Occasionally other areas of the cranium, the radius, and ulna may also be involved. It most commonly affects West Highland White and Scottish terriers but has also been reported in Cairn and Boston terriers, Great Danes, Doberman Pinschers, and in a Labrador Retriever [Riser et al, 1967; Pool and Leighton, 1969; Watson et al, 1973; DeVries and Van De Watering, 1973]. The condition has been reported in the United Stated, Canada and Europe [Riser et al, 1967; Watson et al, 1973; DeVries and Van Watering, 1973]. The disease is a non-neoplastic proliferative disorder of bone with an inflammatory component early in the course of the disease. The condition is most often recognized in 4-7 month-old dogs with signs of discomfort while chewing or eating. Experienced breeders and veterinarians may recognize the disorder at a younger age by clinical signs and/or palpation. Radiographic demonstration of the characteristic bony lesions is the most common method of confirming the diagnosis.

The disease is clearly familial and is generally accepted as being inherited, but a specific mode of inheritance has not been demonstrated [Riser et al, 1967; Pool and Leighton, 1969]. DeVries and Van De Watering {1973] published a pedigree of Scottish terriers showing that all affected dogs in this kindred were sired by the same male, indicating a strong hereditary predisposition.

MATERIALS AND METHODS

A retrospective pedigree analysis of a kindred of West Highland White terrier dogs was undertaken to determine a possible hereditary pattern of CMO. The pedigree is presented in Figure 1. The retrospective analysis (Table I) suggested that the trait was inherited.

Putative carriers in Figure 1 were identified based on the production of at least one affected offspring; all litters produced in subsequent matings by the individuals were

investigated in order to determine if additional affected offspring were produced. One prospective cross of an affected male to an affected female was made to provide additional evidence for the simple autosomal recessive hypothesis (Fig. 1).

Initial identification of affected dogs in this kindred was based on the appearance of clinical signs in at least one pup in the litter and a complete physical examination of all littermates including palpation of the mandible, the angle of the mandible, and the area of the tympanic bullae. Radiographs were used to confirm the presence or absence of CMO. No cases were found in which dogs were identified radiographically as having CMO that were not identified as having the disorder by palpation during physical examination. In our experience, palpation during physical examination is an effective and accurate method of determining the presence or absence of this trait and for several litters, along with the appearance or absence of clinical signs, was the basis for diagnosis.

RESULTS

In most cases of CMO observed in these dogs, the lesions were symmetrical and bilateral and most were diagnosed at age 4-5 months. Although CMO has been reported to be self-limiting, at least in some instances, all affected dogs in this study were treated with corticosteroids and none required euthanasia.

In the pedigree shown (Fig. 1), there was a total of 126 offspring of which 24 had CMO and 102 were phenotypically normal: 19% of the dogs examined in this lineage were affected with CMO. Equal numbers of males and females were affected. No evidence suggesting dominant inheritance was present. Several pups were born dead or died perinatally and we could not assess their CMO status nor could we determine the number or sex of these animals since they were disposed of by the owners. Table I shows those litters having at least one affected offspring among them. If this is an autosomal recessive trait, one would expect 25% of the offspring (18.75) to be affected. We observed 25%, a total of 19 affected dogs in the 18 litters studied. In a prospective mating of two affected dogs, all four of the offspring (two of each sex) had CMO and in one mating of an affected dog to a putative carrier one of two puppies (50%) was affected as would be expected with an autosomal recessive trait. A chi-square statistical analysis for a recessively inherited trait of the number of dogs expected and observed to have CMO showed the numbers were not significantly different at the 0.05 level.

The pedigree and data are consistent with the inheritance of this disorder as an autosomal recessive trait.

DISCUSSION

Craniomandibular osteopathy is a disease of dogs, principally West Highland White and Scottish terriers. Radiographically, it is manifested by an irregular dense bony proliferation on the ramus of the mandible and/or tympanic bulla, which is readily apparent when compared to films of normal dogs. The histopathology of the disorder has been described by Riser et al [1967], DeVries and Van De Watering [1973], Thornburg [1979], and Alexander [1983]. There is an early inflammatory infiltrate with deposition of woven bone at the periphery of the normal bone occurring in a "haphazard" fashion. The new coarse tubercular bone displays a "mosaic pattern of irregular cement lines indicating the sporadic and rapid deposit and resorption of the abnormal bone" [Riser et al, 1967]. With corticosteroid treatment (or sometimes spontaneously) remodeling occurs, the new bone is removed, and the jaw line returns to a normal or near normal appearance.

Several reports [Riser, 1966: Jubb and Kennedy, 1970: Pool and Leighton, 1969] stressed the resemblance of this disease histologically to Paget's disease of humans. Others suggest a closer resemblance to infantile cortical hyperostosis (Caffey-Silverman syndrome) based upon the distribution of the lesions, the clinical course of the disease, and response to treatment [Riser et al, 1967; Thornburg, 1979]. The etiology of Paget's disease is unknown, although there is growing evidence that it is due to a viral infection [Isselbacher et al, 1980]. Infantile cortical hyperostosis in humans in an autosomal dominant trait [Gerrard et al. 1961]. In some respects, CMO resembles both of these disorders but is identical to neither. Further, based on the evidence presented here, the mode of inheritance of CMO appears to be an autosomal recessive trait.

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Fig. 1. Pedigree of a family of West Highland White terriers with CMO showing 24 affected dogs among 126 total offspring.

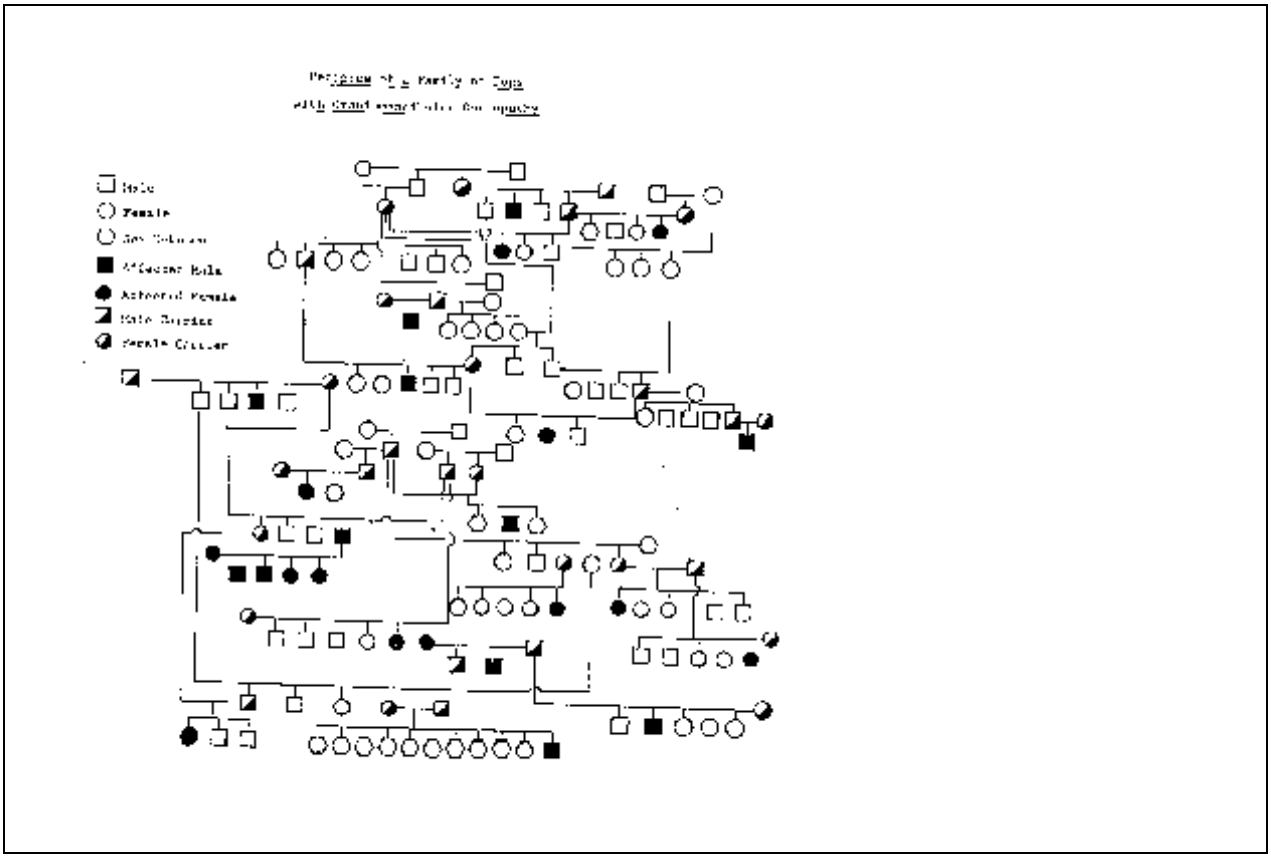

TABLE I. The Number of CMO puppies Expected and Observed in 18 Littes Having at Least One Affected Dog in Each Litter

Size of litter	No. of such litters	Total dogs	Expected No. of affected dogs: Uncorrected per litter	Corrected per litter	Uncorrected in all such litters	Corrected in all such litters	Affected observed
1	2	2	0.25	1	0.5	2	
2	1*	2	0.5	1.143	0.5	1.143	1
3	4	12	0.75	1.292	3	5.188	4
4	4*	16	1	1.463	4	5.852	4
5	4	20	1.25	1.640	5	6.564	
6	2	12	1.5	1.825	3	3.653	
11	1	11	2.75	2.871	2.75	2.871	1
Total	18	75			18.75	27.264	19

*Prospective mating of affected to affected dogs was excluded from the table as well as the mating of the affected to carrier dog.