

Craniomandibular Osteopathy in a West Highland White Terrier

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ABSTRACT

Background: Craniomandibular osteopathy is a bone disorder that occurs in dogs but is not very commonly reported. It is characterized by a non-neoplastic bilaterally symmetrical irregular proliferation of skull or long bones. Immature dogs are most commonly reported with age ranging from three to eight months. The aim of this paper is to report a case of CMO in a West Highland White Terrier.

Case: A 5-month-old entire West Highland white terrier was referred to the Veterinary Hospital with sudden history of severe pain and mandibular swelling, dysphagia, lethargy and weight loss. On physical examination the dog was quiet, alert and responsive. There was thickening of the mandibular bodies with pain at palpation and when opening the mouth. Complete blood count was within normal range. Simple radiographs demonstrated discrete and active new bone formation on the ventral aspect of the mandibular body. Carprofen, dypirone and tramadol were prescribed twice daily for 10 days. 30 days later new radiographs showed more extensive areas of periosteal proliferation advancing to the temporomandibular joint and progression of the looseness of the characteristic compact appearance of lamellar bone of the mandible. The dog was more lethargic, not eating well and in pain. Medication was again prescribed but the treatment was unsuccessful. The dog returned for consultation with severe pain, dysphagia and dramatic weight loss. The dog's owner opted then for euthanasia. Necropsy was performed and histopathological exam revealed irregular and plentiful radiating bony trabeculae composed mostly of woven bone, and the trabecular surfaces displayed scalloping and large number of osteoclasts and osteoblasts, confirming the diagnosis.

Discussion: Craniomandibular osteopathy has been reported in West Highland and Scottish Terriers, and also in Boston Terrier, Cairn Terrier, Shetland Sheepdog, Labrador Retriever, Danish, English Bulldog, Doberman Pinscher, Irish Setter and Boxer. It is also known as 'lion jaw' or 'Westie disease' because the most commonly affected breed is the West Highland White Terrier. An autosomal recessive mode of inheritance has been suggested for terriers breeds. Other theories for the occurrence of CMO in other breeds have been suggested and involve other factors such as bacterial (*Escherichia coli*) or viral (canine distemper virus) infections. The main clinical signs are basically enlarged and painful jaw, drooling, and intermittent fever sometimes as well as lethargy, as observed in this case. Primarily the bones of the skull are affected although there are reports of this condition on the metaphyses of long bones even leading to angular deformities. The dog on this case didn't have any long bones alterations. In severe cases the disease may progress to ankylosis of the temporomandibular joint, which seemed to be occurring as the disease progressed to this joint. Diagnosis is based on clinical, radiographic and histopathological examination and was correctly made and confirmed by necropsy. The prognosis for the disease is usually good, normally clinical signs resolve when the animal reaches about 1 year of age, unless there is involvement of the temporomandibular joint because it will likely lead to ankylosis. Early diagnosis is very important for pain control and supportive and symptomatic medical management of craniomandibular osteopathy, a condition that has no cure so far. Anti-inflammatory drugs are the treatment of choice and were used in this case. Unfortunately this patient didn't respond well to medical management and progressed to a condition incompatible with life due to dysphagia and severe pain, so euthanasia was performed.

Keywords: osteoproliferative disorder, skull, mandible, canine, animal.

INTRODUCTION

Craniomandibular osteopathy (CMO) is an infrequent bone disorder that occurs in dogs [5,12]. It was first reported in West Highland White Terriers in 1958 [6]. Several breeds can be affected, but the most common are West Highland White and Scottish terriers [3,4,12]. It is characterized by a non-neoplastic bilaterally symmetrical irregular proliferation of bone of endochondral origin as the ramus of the mandible, tympanic bullae and occipital bonebut has also been described in long bones [12]. It has been suggested comparison between CMO and infantile cortical hyperostosis (Caffey's disease) in human patients and familial infantile cortical hyperostosis in rhesus monkeys [1,10]. The condition is an autosomal recessive trait [7]. Immature dogs are most commonly reported with age ranging from three to eight months [12]. The aim of this paper is to report a case of CMO in a West Highland White Terrier.

CASE

A 5-month-old entire West Highland White Terrier was referred to the Veterinary Hospital with sudden history of severe pain and mandibular swelling (Figure 1), dysphagia, lethargy and weight loss. On physical examination the dog was quiet, alert and responsive. There was thickening of the mandibular bodies with pain at palpation and when opening the mouth with enlarged submandibular lymph nodes. Complete blood count was within normal range. Simple radiographs demonstrated discrete and active new bone formation on the ventral aspect of the mandibular body (Figure 2A). Carprofen¹ (Rymadil® - 2.2 mg/kg BID), dypirone² (Analgex® V - 25 mg/kg TID) and tramadol³ (Tramal® - 4 mg/kg TID) all orally were prescribed for 10 days. 30 days later new radiographs showed more extensive areas of periosteal proliferation advancing to the temporomandibular joint (TMJ) and progression of the looseness of the characteristic compact appearance of lamellar bone of the mandible (Figure 2B). The dog was more lethargic, not eating well and apparently with increased pain. Medication was again prescribed but the treatment was unsuccessful. The dog returned for consultation with severe pain, dysphagia and dramatic weight loss. The dog's owner opted then for euthanasia. Necropsy was performed and histopathological exam confirmed the diagnosis.



Figure 1. A 5-month-old West Highland White Terrier with mandibular swelling and drooling on first consultation.

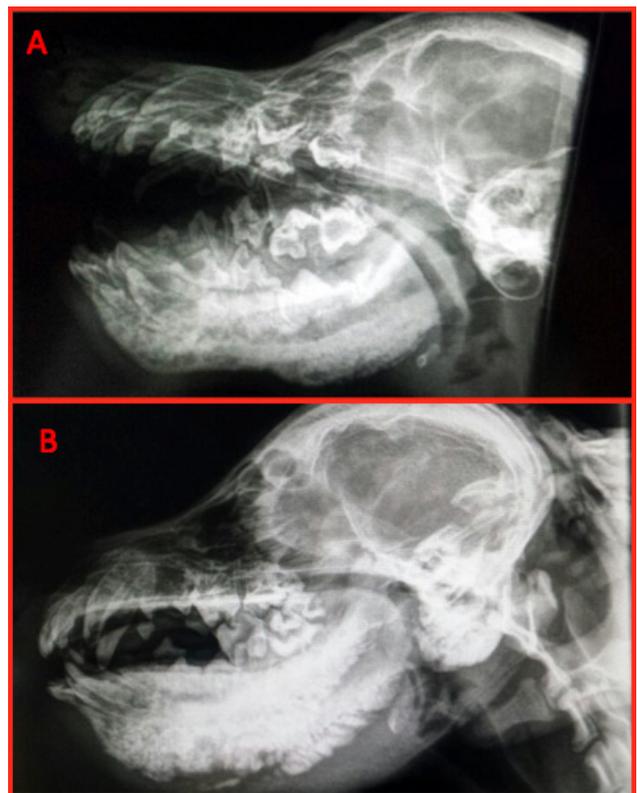


Figure 2. Skull radiographs on lateral oblique projection. At the moment of diagnosis showing active and discrete new bone formation on the mandibular body (A). Within 30 days of evolution - notice periosteal proliferation advancing to the TMJ (B).

DISCUSSION

Craniomandibular osteopathy has been reported in West Highland and Scottish Terriers, and also in Boston Terrier, Cairn Terrier, Shetland Sheepdog, Labrador Retriever, Danish, English Bulldog, Doberman Pinscher, Irish Setter, Boxer, Bullmastiff and others [3,4,9]. It is also known as 'lion jaw' or 'Westie disease' because the most commonly affected breed is the West Highland White Terrier [12]. An autosomal recessive mode of inheritance has been suggested for terriers breeds [7]. Other theories for the occurrence of CMO in other breeds have been suggested and involve other factors such as bacterial (*Escherichia coli*) or viral (canine distemper virus) infections [5,7]. The main clinical signs are basically enlarged and painful jaw, drooling, and intermittent fever sometimes as well as lethargy, as observed in this case [12]. Exophthalmos and strabismus have been reported [2].

Primarily the bones of the skull are affected, although there are reports of this condition on the metaphyses of long bones even leading to angular limb deformities as carpal deformity [8]. The dog on this case didn't have any long bones alterations. In severe cases the disease may progress to ankylosis of the temporomandibular joint, which seemed to be occurring as the disease progressed to this joint [8,12]. Diagnosis is based on clinical, radiographic and histopathological

examination and was correctly made and confirmed by necropsy and histopathological analysis [12]. The differential diagnoses are osteosarcoma, hyperparathyroidism or normal callus formation [7].

The prognosis for the disease is usually good if there is no involvement of the temporomandibular joint, normally clinical signs resolve when the animal reaches about 1 year of age, but if the TMJ is involved there will likely be a progression towards ankylosis [12]. Early diagnosis is very important for pain control and supportive and symptomatic medical management of CMO, a condition that has no cure so far [8]. Anti-inflammatory drugs are the treatment of choice and non-steroids anti-inflammatory drugs (NSAIDs) were used in this case although some authors recommend the use of steroidal ones [3]. Unfortunately this patient didn't respond well to medical management and progressed to a condition incompatible with life due to dysphagia and severe pain, so euthanasia was performed.

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Declaration of interest. The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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