

Craniomandibular osteopathy in West Highland white terrier – a case report



Rajka Turner, Berislav Radišić and Marija Lipar*

Introduction

Craniomandibular osteopathy (CMO) in dogs is an uncommon non-neoplastic proliferative bone disease that somewhat resembles infantile cortical hyperostosis (ICH) and Paget's bony disease in humans (Padgett and Mostosky, 1986). It is most frequently diagnosed in West Highland white terriers (Montgomery, 2003), Scottish terriers, cairn terriers (Riser and Newton, 1985; Johnson, 2010) and Boston terriers (Piermattei et al., 2006; Johnson, 2010). However, it has also been reported in Labrador retrievers (Montgomery, 2003), Great Danes (Piermattei et al., 2006; Johnson, 2010), Shetland sheep dogs (Taylor et al., 1995), boxers (Shultz, 1978), Doberman Pinschers (Huchowsky, 2002), Pyrenean mountain dogs (Franch et al., 1998), Pitt Bull terriers (Thompson et al., 2011) and bullmastiffs (Huchowsky, 2002). It primarily affects the mandible and tympanic bullae, though it can also occasionally affect other skull bones, such as the frontal (Dennis et al., 1993), parietal (Riser and Newton, 1985; Dennis et al., 1993; Weisbrode, 2007) and nasal bones (Taylor et al., 1995). In some cases, the bones of the appendicular skeleton may also be involved (Riser and Newton, 1985; Padgett and Mostosky,

1986; Montgomery, 2003). The disorder is usually resolved spontaneously at the age of 11 to 13 months (Riser and Newton, 1985) and symptomatic treatment is designed to alleviate pain and discomfort, while also providing nutritional support. Radiography (Riser and Newton, 1985; Huchowsky, 2002; Piermattei et al., 2006) and magnetic resonance imaging (MRI) (Matiasovic et al., 2016) are the most valuable diagnostic procedures for CMO, since the results of the serum chemistry and haematology profile are considered unspecific for the disorder (Riser and Newton, 1985; Huchowsky, 2000).

Additional diagnostic procedures, such as bone biopsy, may also be used in more atypical cases of CMO to confirm the diagnosis (Johnson, 2010). Our goal is to remind practitioners of the existence of this rare disease in both terrier and non-terrier breeds, as it may be easily misdiagnosed.

Case presentation

An 8-month old, intact, male West Highland white terrier with a history of intermandibular swelling of a 3-week duration was referred to the surgery

Rajka TURNER, DVM, Best friends group, Cambridgeshire, UK, (Corresponding author, e-mail: turner.rajka@gmail.com), Zagreb, Croatia; Berislav RADIŠIĆ, DVM, PhD, Full Professor, Marija LIPAR, DVM, PhD, Scientific Advisor, Faculty of Veterinary Medicine University of Zagreb, Croatia

clinic. The vaccination record was up to date. Temperature, respiratory and heart rate were within the reference range. A painful swelling, measuring approximately 1 cm in diameter, was palpable between the *rami mandibulae*. During physical examination, the animal was reluctant to open the mouth and showed obvious signs of discomfort on passive jaw manipulation. The blood test showed no detectable abnormality in the complete blood cell count (CBC), whereas the serum chemistry profile showed slightly elevated alkaline phosphatase (AP 287 U/L; reference range 23–212 U/L).

Obtaining skull radiographs and collecting tissue samples for the pathohistological diagnosis (PHD) required the use of general anaesthesia.

Fifteen minutes prior to the induction of general anaesthesia (GA), the animal was sedated with 0.05 µg/kg IM Medetomidine. Following induction with 4 mg/kg IV Propofol,

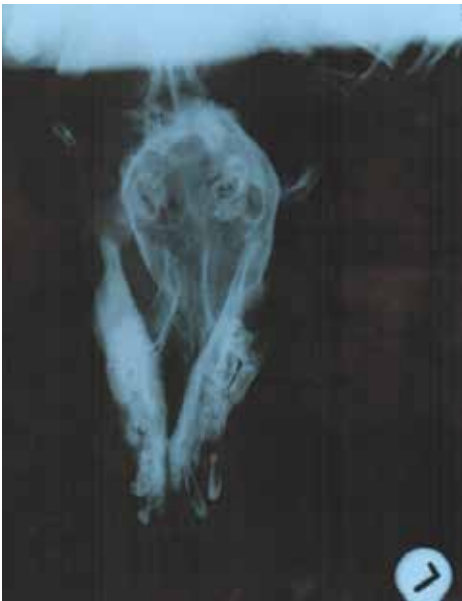


Figure 1. Ventrodorsal radiograph of the skull demonstrating new bone formation on both rami mandibulae with no interference of tympanic bullae and temporomandibular joints

the dog was intubated and anaesthesia was maintained with 1.5% Isoflurane in 100% oxygen. Fifteen minutes before GA induction, Methadone at a dosage of 0.25 mg/kg IM was administered for improving surgical analgesia. The procedure took 20 minutes to complete, after which anaesthesia was discontinued and recovery followed within 10 minutes. Cefuroxime 20 mg/kg IV was administered as a prophylactic antimicrobial therapy. Perioperative fluid management protocol included IV administration of 0.9% sodium chloride.

The ventro-dorsal skull radiograph revealed characteristically bilateral and slightly asymmetric bone proliferations located on both *rami mandibulae*, with unaffected temporomandibular joints and tympanic bullae (Figure 1).

A tissue sample for PHD was obtained from the middle third of the left *ramus mandibulae*, a few centimetres distal to the crown of the fourth premolar tooth. PHD demonstrated bone remodelling and inflammation in the periosteal region and overlying soft tissue. The islets of newly formed cartilage tissue in the periosteal region were enclosed with a considerable amount of red blood cells and neutrophils, with clusters of morphologically characteristic osteoclasts and fewer osteoblasts. Newly deposited coarse bone fibres that stained unevenly dark were arranged in an interlacing pattern, accompanied by an infiltration of inflammatory cells, predominantly neutrophils. At the periphery, a large bundle of mature collagen remaining from lamellar bone destruction was visible, with an agglomeration of degenerated neutrophils attached to its side.

The results of the physical examination, radiography and PHD were most consistent with the diagnosis of CMO.

Symptomatic treatment included orally administered Methylprednisolone

5 mg/kg SID and Ranitidine 0.1 mg/kg BID to be given during the first two weeks. Following that time, treatment with Methylprednisolone in a dosage of 1 mg/kg PO SID was continued for an additional two weeks.

At four weeks after admission, the owner informed us that the animal was pain free and had no observable difficulties while feeding. Five months after discontinuation of the Methylprednisolone therapy, the animal appeared to be completely recovered.

Discussion

CMO is a result of abnormal bone proliferation of mandibular bone, tympanic bullae and occasionally other bones of the skull. The maxillary, frontal, lacrimal and parietal bones are more frequently affected in small breeds, whereas in large breeds the lesions are usually confined to the mandibular bone (Huchowsky, 2002). The thickening of the calvarial wall, which in pathomorphological features strongly resembles CMO, was described under the term "*calvarial hyperostotic syndrome*" and observed in bullmastiffs (Pastor et al., 2000; McConnell et al., 2006; Weisbrode, 2007) and in a Pitt Bull terrier, where the mandible was also involved along with calvarium bony proliferation (Thompson et al., 2011). Involvement of the auditory canal (Huchowsky, 2002), and retrobulbar bone proliferation resulting in exophthalmos (Dennis et al., 1993) have also been reported. With respect to its physical appearance and clinical symptoms, this rarely occurring disease can mimic a wide variety of disorders and that should be considered during the differential diagnosis, including masticatory muscle myositis, osteomyelitis, hypertrophic osteodystrophy (HOD), bone neoplasia, traumatic periostitis (Huchowsky, 2002)

and secondary hyperparathyroidism (Wiggs and Lobprise, 1997).

The main characteristics of CMO are osteoclastic resorption of the pre-existing lamellar bone and its replacement with coarse trabecular new bone tissue expanding beyond normal boundaries (Thornburg, 1997; Johnson, 2010). The massive infiltration of inflammatory cells is commonly observed during the process of osteoclastic destruction of the lamellar bone and in adjacent areas, as the overlying muscle fibres and connective tissue have been destroyed at the rough margins of the newly proliferated bone (Riser and Newton, 1985; Thornburg, 1997). Between the newly grown coarse woven bone parts, the normal bone marrow is usually replaced by highly vascular, fibrous *stroma* (Riser and Newton, 1985; Taylor et al., 1995). A mosaic pattern of thin irregular basophilic cement lines, marking the borders where each new layer has formed, can be seen in new primitive bone (Weisbrode, 2007; Johnson, 2010), leaving the impression as if the new woven bone had been plastered upon old bone (Riser and Newton, 1985). The growth of abnormal bone subsides at 7 to 8 months of age and, in some cases, may regress completely when dogs are 11 to 13 months old (Riser and Newton, 1985; Piermattei et al., 2006). Disease self-limitation and recovery usually follows the completion of regular endochondral ossification and growth plate closure (Riser and Newton, 1985; Montgomery 2003; Piermattei et al., 2006). Although the primary condition does not seem to be fatal, euthanasia has been performed in several cases of temporomandibular joint fusion and excessive tympanic bullae proliferation due to severe pain and malnourishment arising from the inability to eat (Huchowsky, 2002; Montgomery, 2003; Piermattei et al., 2006).

The etiology of CMO is still not fully known, although an autosomal

recessive mode of inheritance had been demonstrated in West Highland white terrier (Padget and Mostosky, 1986; Weisbrode, 2007). An inapparent infection with canine distemper virus has also been suggested as a possible initiating factor for the disease (Huchowsky 2002; Piermattei et al., 2006).

The fundamental diagnostic tool for CMO is radiographic examination. Radiographic signs include usually bilateral symmetric or asymmetric cotton-wool-like radiopaque irregular bone proliferations in affected skull areas (Johnson, 2010). Often, bone cortices and the medullary cavity cannot be seen due to superimposed newly proliferated dense bone tissue (Riser and Newton, 1985). Overlying soft tissue may appear thickened, and diffuse, patchy bone radiolucency may be seen occasionally, indicating bone resorption in adjacent areas (Taylor et al., 1995). With the completion of the growth process, bone proliferations become radiographically static and tend to become reduced, while the irregular edges of newly proliferated bone usually become smooth (Riser and Newton, 1985; Piermattei et al., 2006). Although slowly regressing, the radiographic abnormality or impaired function may remain, particularly if the temporomandibular joints and tympanic bullae have been involved extensively, resulting in permanent jaw movement restriction (Wiggs and Lobprise, 1997; Johnson, 2010). Bone biopsy may be useful in atypical cases of CMO, as in rarely affected breeds, especially if the lesions are unilateral and confined to the mandible.

In most occasions, the complete blood count and serum chemistry profile are within the normal range (Huchowsky, 2002). Nevertheless, there might be an elevation in serum concentrations of alkaline phosphatase (AP) and phosphorous, though these could be interpreted as normal findings

in growing dogs, since in young healthy animals, these values often exceed the adult reference range. Although uncommonly found, hypoalbuminemia might be present in animals suffering from malnutrition over an extended period of time (Taylor et al., 1995).

Treatment is symptomatic and aims to alleviate pain and provide nutritional support for the starving patient. Nonsteroidal anti-inflammatory drugs (NSAIDs) are frequently used as well as corticosteroids, although not at the same time as these drugs combined together can cause serious gastro-intestinal injury and ulcerations. Patients can benefit from their analgesic effects anti-inflammatory actions, but it has been acknowledged that these drugs do not influence the size of the bone proliferation (Riser and Newton, 1985). Broad spectrum antibiotics are usually given with corticosteroids to prevent possible secondary bacterial infections arising from glucocorticoid immunosuppressive action.

Surgical attempts to remove bone proliferations or to increase range of motion of temporomandibular joint have been unsuccessful (Riser and Newton, 1985). In one documented case, the surgical excision of the exostoses resulted in their regrowth within 3 weeks (Piermattei et al., 2006).

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Abstract

The case report describes craniomandibular osteopathy observed in an 8-month old West Highland white terrier. The animal was presented with a history of painful intramandibular swelling lasting three weeks.

Although most frequently observed in that particular breed, the condition is generally rare and therefore likely to be unrecognized by clinicians. The diagnosis was based on clinical and radiological findings with bone biopsy used as a supplemental diagnostic method. Symptomatic treatment with anti-inflammatory drugs was administered to alleviate pain and discomfort during the bone proliferation phase. In this case, the prognosis was favourable due to the unaffected temporomandibular joint and tympanic bullae, making special nutritional therapy protocol unnecessary. The condition usually appears in dogs aged 4 to 8 months, and is characterized by non-neoplastic bone remodelling and its irregular thickening with inflammation. Predominantly affecting the mandible and occasionally other bones of the skull, the disorder commonly results in painful and difficult mastication, leading to nutritional impairment and dehydration of the animal. The disease is mostly self-limiting and recovery coincides with the completion of the growth process and epiphyseal plate closure at 11 to 13 months of age. However, in some cases, radiographic abnormality or impaired function of the tympanic bullae or temporomandibular joint may remain, resulting in permanent restriction of jaw movement.

Key words: *mandible, osteopathy, bone proliferation, inflammation, epiphyseal plate closure*

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Kraniomandibularna osteopatija u zapadno-škotskog bijelog terijera – prikaz slučaja

Rajka TURNER, dr. med. vet., Best friends group Cambridgeshire, Velika Britanija; dr. sc. Berislav RADIŠIĆ, dr. med. vet., redoviti profesor, dr. sc. Marija LIPAR, dr. med. vet., znanstvena savjetnica, Veterinarski fakultet Sveučilišta u Zagrebu, Hrvatska

Prikaz slučaja opisuje kraniomandibularnu osteopatiju koja se pojavila u 8 mjeseci starog zapadno-škotskog bijelog terijera. Životinja je upućena u kiruršku ambulantu zbog bolne otekline koja se tri tjedna prije pojavila u intermandibularnom prostoru. Iako najčešće dijagnosticirana u navedene pasmine, kraniomandibularna osteopatija je općenito rijedak poremećaj te postoji velika vjerojatnost da je zbog toga kliničari neće prepoznati. Dijagnoza poremećaja se temeljila na nalazima rendgenografije dok je biopsija kosti služila kao dopunska metoda za potvrdu dijagnoze. Simptomatsko liječenje bilo je usmjereno olakšavanju bolnosti tijekom proliferativne faze bolesti. U ovom slučaju prognoza je bila povoljna, jer promjene nisu zahvaćale temporomandibularni zglob niti timpanične bule, zbog čega nije bilo potrebno primjenjivati poseban protokol

za prehranu pacijenta. Poremećaj se najčešće javlja u 4 do 8 mjeseci starih pasa i karakterizira ga neneoplastično remodeliranje kosti s nepravilnim koštanim bujanjima i upalom. Proliferacija koštanog tkiva najčešće zahvaća mandibulu, a ponekad i druge kosti lubanje te uglavnom rezultira bolnošću prilikom uzimanja hrane i žvakanja, zbog čega pacijent postaje pothranjen i dehidriran. Bolnost spontano posustaje te oporavak koincidira sa završetkom rasta i zatvaranjem koštanih zona rasta u dobi od 11 do 13 mjeseci. Međutim, ponekad radiološki vidljive lezije i narušena funkcija temporomandibularnog zgloba može ostati, što rezultira trajno smanjenim opsegom pokretljivosti zgloba.

Ključne riječi: *mandibula, osteopatija, koštana proliferacija, upala, epifizno zatvaranje kosti*