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CASE REPORT

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Calvarial hyperostosis syndrome in a Dalmatian dog

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Key Clinical Message

Calvarial hyperostosis syndrome is an uncommon and self-limiting disease affecting juvenile dogs. Only symptomatic treatment has been described, and diagnosis is based on clinical findings, imaging, and disease progression.

Abstract

This is the first reported case of calvarial hyperostosis syndrome in a Dalmatian dog. It is an uncommon osteoproliferative disease with diagnosis frequently based on clinical signs, imaging findings, and disease progression, with only symptomatic treatment described. Case describes a 5-month-old Dalmatian dog presented with a facial mass and difficulty eating. After imaging, mass was observed to be osteoproliferative, nonaggressive, and without affection of the temporomandibular joints and mandibles. Histology revealed an osseous–cartilaginous and proliferative lesion, together with scant amount of neutrophiles. Clinical improvement was observed after symptomatic treatment, and moderate lesion regression was observed in a CT reevaluation 6 months later.

KEYWORDS

calvarial, Dalmatian, hyperostosis, osteoproliferative, self-limiting

1 | INTRODUCTION

Calvarial hyperostosis syndrome (CHS) is a poorly described osteoproliferative disease characterized by nonneoplastic and asymmetric proliferation of the flat bones of the skull. Clinically and histologically, it resembles craniomandibular osteopathy (CMO) in dogs or infantile cortical hyperostosis (ICH) in humans.[1–6] Distinction between CHS and CMO is mostly based on localization, imaging findings, and mild histological differences. CHS was first reported in Bullmastiffs,[2,3] and in other breeds such as Pit Bull Terrier, American Pit Bull Terrier, English Springer Spaniel, and Weimaraner.[1,4–6] The clinical presentation usually consists of painful inflammation of the skull bones (usually flat bones such as parietal), lack of involvement of the temporomandibular joint and mandibles, together with possible associated lymphadenopathy, eosinophilia, and fever.[1,2] It is a self-limiting disease that affects young patients from a few weeks of age, without evidence of sexual predisposition. As it is a self-limiting disease, there is no specific treatment for it, with only analgesia being provided if needed.[1,3,4]

Here we report the clinical, radiological, and histopathological features of CHS in a 5-month-old Dalmatian dog, being the first case described in this breed.

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2 | CASE HISTORY

A 5-month-old male Dalmatian dog presented to the Fundació Hospital Clinic Veterinari (Universitat Autònoma de Barcelona) with a 2-week history of apathy and difficulty eating and drinking. The dog had presented 2 weeks prior to another veterinary practice, where a nodule/mass in the right mandibular lymph node region and otitis externa in the right ear were detected. Antibiotic (amoxicillin-clavulanic acid 20 mg/kg BID orally for 10 days), anti-inflammatory (meloxicam 0.1 mg/kg SID orally for 7 days) and cleaning drops were prescribed. During the treatment, the owners observed improvement, with no clear pain signs or difficulty eating, although 5 days after discontinuing it, clinical signs got worse, and the dog was brought to our hospital. The physical examination revealed right-sided atrophy of the masseter and temporal muscles, severe pain when opening the mouth and the presence of a hard nodule at the base of the right ear of about 5 cm in diameter. The lymph nodes were normal at palpation. No other abnormalities were present during physical exam. A complete blood count (CBC) and biochemistry were performed, with only mild monocitosis [1.81 k/µl (0.16–1,12 K/µl)] present. After initial consultation, the initial differential diagnoses included masticatory myositis, changes due to prior trauma or foreign body, or other causes.

Based on the findings, radiographs were performed under sedation, for which a combination of dexmedetomidine (3 µg/kg), methadone (0.2 mg/kg), ketamine (1 mg/kg), alfaxalone (1 mg/kg), and midazolam (0.2 mg/kg) intramuscularly was administered, together with propofol (2 mg/kg) intravenously. When sedated, no difficulties were noted while opening the mouth, with a normal range of movement of the mandibles. Skull radiographs were taken in dorsoventral (DV) and oblique (right dorsal-left ventral) projections. The radiographs revealed a round osteoproliferative mass with slightly irregular yet well-defined margins (4.6 cm high × 4.7 cm long × 6.2 cm wide), located ventral to the right tympanic bulla and in relation with the temporomandibular joint. The mass presented heterogeneous mineral opacity with no evidence of bone lysis in the adjacent structures. The ventral wall of the right bulla was thickened (6.3 mm) and the external ear canal could not be well visualized, while it was well aerated on the left side (Figure 1). Under the same anesthetic episode, fine needle aspiration cytologic samples from both the mandibular lymph nodes and the mass were obtained. Lymph node cytology revealed the presence of a heterogeneous population of lymphocytes together with a moderate amount of plasma cells, compatible with a reactive lymph node. The mass cytology was considered nondiagnostic as very few cells were obtained.

After the radiographs, a presumptive diagnosis of atypical craniomandibular osteopathy (CMO) was established, as it was not a breed with a described predisposition and there was only unilateral involvement. Even so, other possible etiologies (e.g., osteomyelitis associated to foreign body, trauma, and neoplastic process) were not ruled out and treatment with NSAIDs (meloxicam 0.1 mg/kg SID orally for 7 days, with subsequent reduction to 0.05 mg/kg SID orally for 7 days more) and a soft diet were started. Monthly follow-ups were suggested as well as performing advanced imaging techniques (computed tomography, CT) and biopsy in the event of unfavorable clinical evolution. When the anti-inflammatory treatment was stopped after 15 days, the patient showed worsening of the clinical signs together with a mild lameness in a hind limb, so CT scan and biopsies were planned.

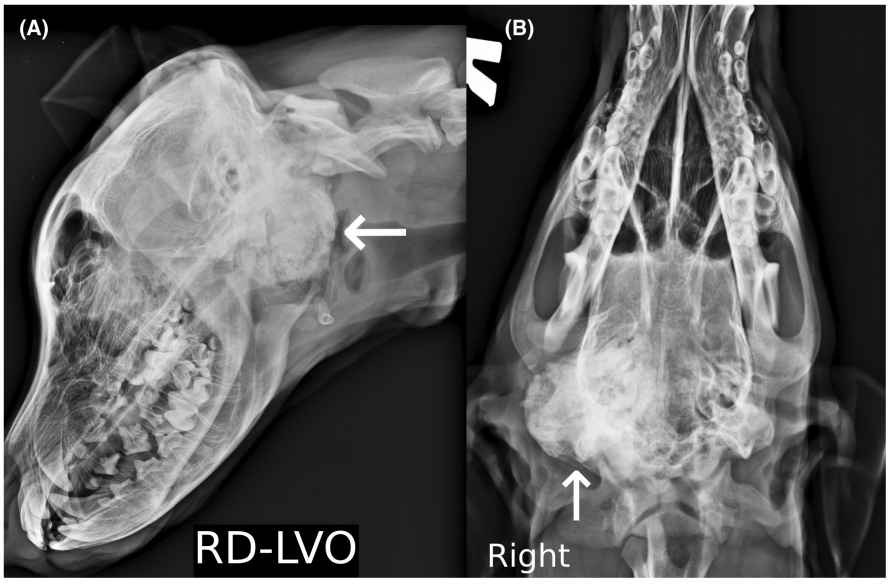

FIGURE 1 DV (A) and oblique (B) radiographs of the skull, showing the rounded mass (white arrows) of mineral opacity, located at the region of the right tympanic bulla.

A full-body CT study was performed (skull and extremities with a slice thickness of 1.25 mm, thorax and abdomen with a slice thickness of 2.5 mm), before and after administration of intravenous iodinated contrast medium. In the skull, the presence of the previously described mass was observed (Figures 2, 5A), affecting the right temporomandibular region, extending from the level of the cranial part of C2 until joining the temporal bone in the region of the middle ear and caudal to the zygomatic process. The mass showed mineral attenuation with multiple hypoattenuating areas, having a "cauliflower-like" appearance, and with irregular margins in its most caudal portion, and was slightly heterogeneous in its area of attachment to the temporal bone. In the junction area, a marked thickening and increased attenuation of the skull bones in the region (temporal and occipital) were observed, until reaching approximately the midline and extending toward the left side, where the same type of proliferative lesion of the parietal and temporal bones was observed up to the region of the middle ear. In the left side, changes were markedly less severe when compared to the right. Bilaterally, the temporomandibular joints and mandibles were unremarkable. Secondary to the presence of the mass, the right external ear canal was decreased in diameter; a slight amount of fluid was present in the tympanic cavity, whose lumen was markedly reduced with complete loss of the normal anatomy associated with the osteoproliferative lesion. The regional lymph nodes (mandibular and retropharyngeal) were slightly increased in size, although maintaining normal morphology and attenuation. In the rest of the regions included in the study (thorax, abdomen, and extremities) no significant alterations were detected. Considering this CT findings, CHS, or an atypical CMO was considered most likely; other differentials, such as osteochondroma, osteochondromatosis, or other neoplasia were considered less likely.

Biopsy samples were taken from the mineralized mass and right parietal bone, using Jamshidi needles. Nine incisional samples of 3–5 mm depth were obtained and submitted for histological evaluation. Microscopic examination revealed irregular and thick trabecular bone composed by woven bone, lamellar bone, and immature cartilage. The trabeculae showed small medullary spaces lined by activated osteoblasts and scant osteoclasts with mild fibroblastic proliferation and scant neutrophilic infiltrate. Irregular resting lines were also seen at the periphery of the trabeculae (Figure 3). Changes were consistent with bone-cartilage proliferative lesion, probably reactive or less probable a benign neoplasm.

Based on clinical evolution and further diagnostics findings, a presumptive diagnosis of CHS was established.

The dog was treated continuously with NSAIDS (meloxicam 0.05–0.1 mg/kg SID orally) and/or metamizole (25 mg/kg BID orally) to control pain. Monthly follow-ups were performed, with progressive improvement of the clinical signs and normalization of the

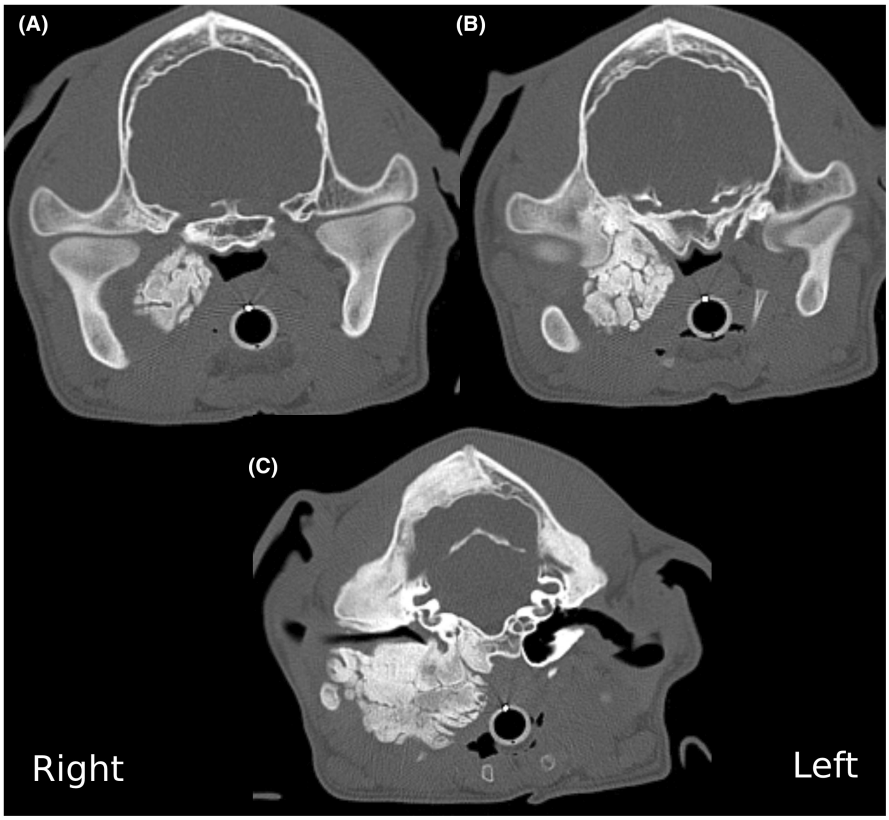

FIGURE 2 CT study of the skull in bone algorithm from rostral to caudal (A–C), showing the mass-like lesion at the right temporomandibular region, with increased bilateral thickness and hyperattenuation of the temporal, parietal, and occipital bones. Both temporomandibular joints are within normal limits.

physical exam. Six months later the dog was rechecked for elective castration; he showed no pain, and was not given any medication for 2 months. A skull CT scan was proposed and accepted by the owners taking advantage of the need for general anesthesia. On physical exam, the mass was smaller compared to previous physical exams, with persistence of a very slight right-sided atrophy of the temporal muscles. No other abnormalities were noted in the physical exam. In the control CT, the lesions affecting the temporal and parietal bones completely resolved. The only changes that persisted were the mild regional lymphadenopathy and the mass, which had a more compacted appearance compared to the previous study, being slightly smaller. The temporomandibular joints and mandibles still showed no abnormalities. (Figures 4, 5B).

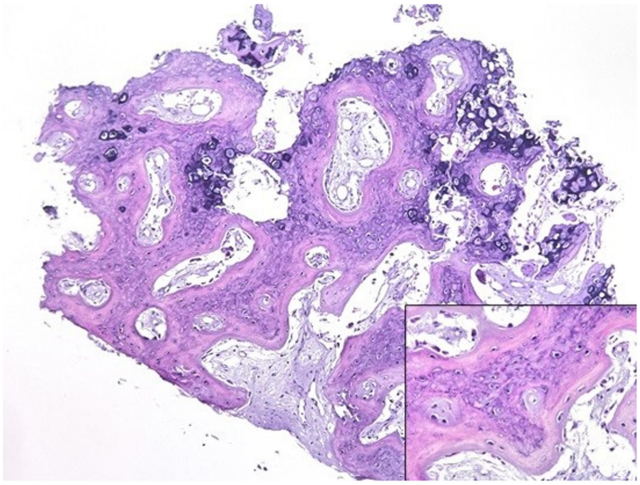

FIGURE 3 Histopathologic (HE) image showed trabeculae composed by two components; woven bone, and cartilage with irregular resting lines. Trabeculae show small medullary spaces lined by normal osteoblasts and few osteoclasts.

3 | DISCUSSION

Calvarial hyperostosis syndrome is a poorly described disease seen in young canine patients of multiple breeds. It is a nonneoplastic osteoproliferative disease, characterized by exacerbated growth of the cortical or periosteal bone of the skull, which can be painful.[1–6] Other associated clinical signs include lymphadenopathy, eosinophilia and fever, with other less frequent signs such as hydrocephalus, lameness, or purulent osteomyelitis described in some case reports.[1,2] It is mostly described in Bullmastiffs, although it has also been reported in

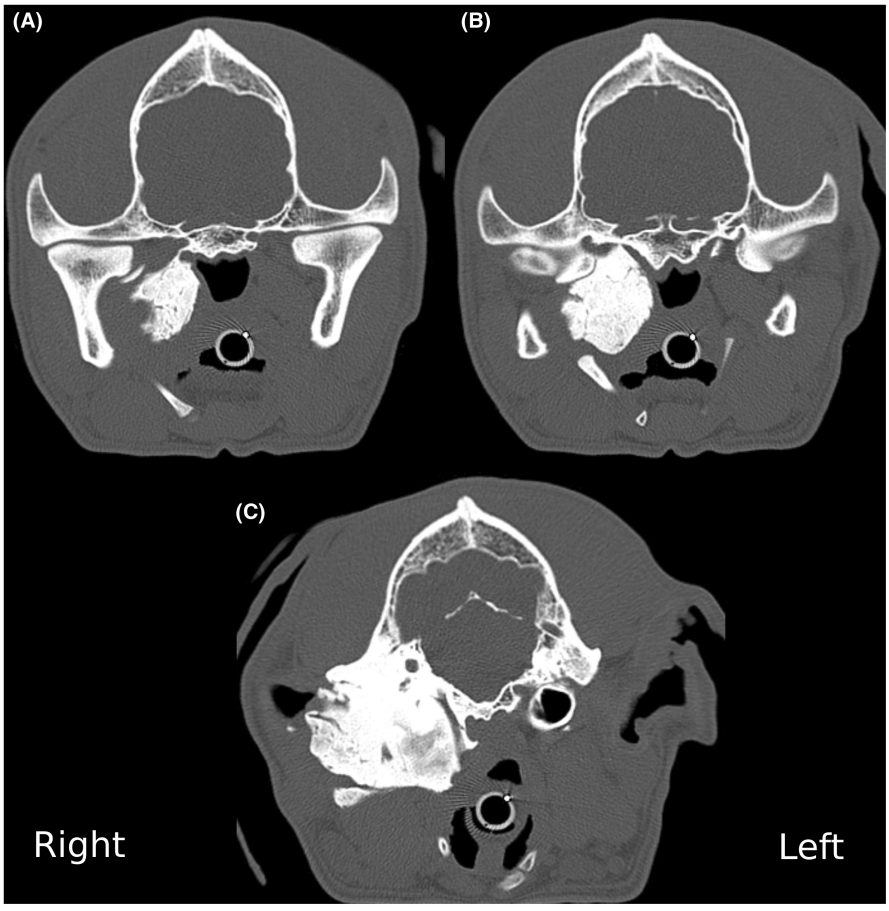

FIGURE 4 Skull CT 6 months after diagnosis, at the same level of the images of Figure 1. Absence of the previously described thickening and hyperattenuation of the temporal, parietal, and occipital bones. The mass-like lesion at the region of the temporomandibular joint has a more compacted appearance.

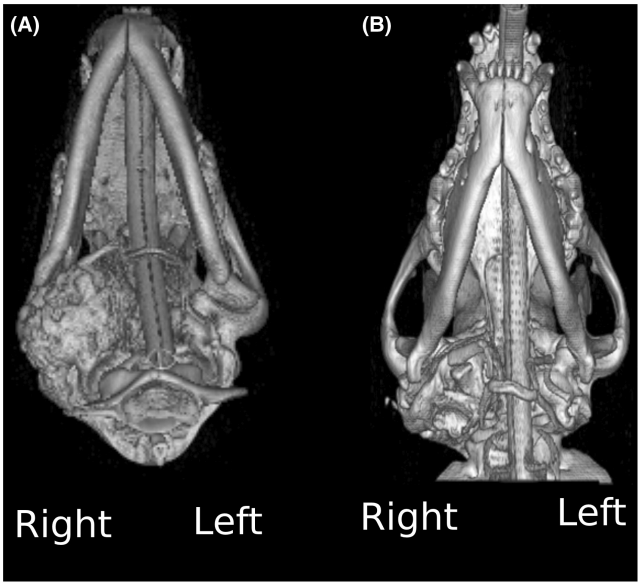

FIGURE 5 Comparative 3D reconstructions in VD position of the initial study (A) and 6 months after diagnosis (B). A moderate decrease of the mass volume is observed.

Pit Bull Terriers, Weimaraners, and English Springer Spaniels.[1–6] A possible male predisposition was observed but has not been confirmed. In the case described in the present article, the patient presented painful inflammation of the skull bones together with lymphadenopathy. This is the first report of a Dalmatian dog in which the disease has been described.

Calvarial hyperostosis syndrome is a disease that presents clinical, radiological, and histopathological features similar to those of craniomandibular osteopathy (CMO) in dogs or infantile cortical hyperostosis in children (ICH), although with small differences at the histopathological level and in imaging caracteristics.[1–4,6] In both CHS and CMO, nonneoplastic, painful, and self-limiting bone proliferation is observed affecting the skull bones of young (4–10 months of age) dogs. In both diseases, regression of the changes with persistence of mild changes is expected when dog reaches skeletal maturity, even so a few cases have been described with complete disappearance of lesions.[1–6] Even so, differences are observed between the two, with CHS having a normally unilateral and asymmetric distribution, without involvement of the mandible and with a predilection for flat bones, in which a thickening of the cortical bone is observed radiologically.[1,4] In some cases, bilateral involvement of the flat bones of the skull has been described, although it is an extension of a unilateral lesion, not a pure bilateral involvement.

Infantile cortical hyperostosis is a hereditary disease observed in children between 10 weeks and 5 months of age, characterized by subperiosteal, bilateral, and symmetric bone neoformation, mainly affecting the mandible and the appendicular skeleton, also tending to be self-limiting.[2]

Histologically, both CMO and CHS show a nonneoplastic bone proliferation with various degrees of inflammation, although this depends on the time the sample is obtained and the treatment previously received.[1–4,6] Even so, there are differences at the histological level. CMO consists of interconnected trabecular bone, endosteal neoformation, and abundant osteoclasts on the bone surface.[4] The medullary cavities are filled with connective tissue and dense foci of inflammatory cells. CHS consists of fibrovascular tissue in the intertrabecular space, sometimes with an inflammatory infiltrate, which will depend on the stage of the disease at which the biopsies are taken. In our dog, the histopathology showed bone-cartilage proliferation without involvement of the medullary cavity, consistent with CHS.[4]

Long bone involvement has also been described in several cases of CHS, with evidence of osteomyelitis and response to antibiotics, supporting a possible bacterial origin in some cases.[2,3,6] In our case, no bone alteration was observed in any of the limbs or other bones, even though the patient presented a specific episode of lameness.

Aside from CMO, other differentials for these injuries in our case are osteomyelitis or neoplasia.

The etiology of CHS remains unknown, although it does not appear to be associated with traumatic, neoplastic, or degenerative abnormalities.[2,3,6] Possible originating factors are infectious, nutritional, metabolic, or genetic. In the case of ICH, a possible role of prostaglandins E1 and E2 has been suggested,[1] which in some studies with children affected by the disease were observed to be increased in serum. In the same studies, a role of collagen disorders was suggested.

Most cases of CHS are self-limiting, with lesions regressing once bone tissue maturity is reached.[1–6] The treatments proposed consist of anti-inflammatories (NSAIDs and steroids) together with analgesics, to alleviate the discomfort and pain of the patients during the active phases of the disease.[1,3,4] Because in some cases the images can be mistaken with foci of osteomyelitis, antibiotic treatment has also been administered. Even so, no treatment has been described that prevents, reduces or reverses the lesions, acting only as palliative. In the case described, the patient presented a good response to anti-inflammatory treatment with NSAIDs, and these could be de-escalated until their withdrawal at a time close to the patient's skeletal maturity.

The diagnosis of CHS is based on the clinical presentation, symptoms, disease progression, radiologic findings, and histopathology.[1–6] Although radiography has been used as the imaging technique in most of the cases, in two

cases MRI was performed,[1,3] and CT has several advantages over both. CT allows a multiplanar study with better evaluation and detail of the multiple layers of the bone, allowing a better delineation of the involvement and its degree in the different bones. Although it requires general anesthesia, it is a faster technique than MRI, allowing a better resolution and assessment of the skeletal system. In the present study, two CT studies were performed, one at the time of diagnosis and the second 6 months later. In this study, the diagnosis of CHS was established based on the premise that it was a young large breed dog, with a painful, mainly unilateral, asymmetric thickening of several skull bones without temporomandibular joint or mandibular involvement, presence of concurrent lymphadenopathy, histopathological lesions compatible with nonneoplastic bone proliferation and self-limiting character with partial regression after several months. In conclusion, CHS should be considered in the differential diagnosis of young Dalmatian dogs presenting with this clinical and radiographic signs.

AUTHOR CONTRIBUTIONS

Carles Planas: Conceptualization; investigation; writing – original draft. **Rosa Novellas:** Supervision; validation; visualization; writing – review and editing. **Yvonne Espada:** Supervision; validation; visualization; writing – review and editing. **Albert Lloret:** Conceptualization; supervision; validation; visualization; writing – review and editing. **Jaume Alomar:** Resources; validation.

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CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest or financial disclosures.

DATA AVAILABILITY STATEMENT

Patient-specific identifying information was omitted, and no patient information was altered or falsified in an attempt to maintain anonymity.

CONSENT

Written informed consent for research and publication is obtained by the institution from the patient's owner upon patient registration in accordance with the journal's patient consent policy. Owner consent to perform the follow-up CT under the same anesthetic event for routine castration was obtained in advance of the procedure.

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