Case Report

Bilateral angular carpal deformity in a dog with craniomandibular osteopathy

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Summary
A four-month-old West Highland White Terrier was presented to the Small Animal Teaching Hospital at the University of Liverpool with the complaint of a bilateral angular carpal deformity. A 20° valgus deformity was present in both thoracic limbs, centred on the distal radiophyses. Both distal ulnas were grossly thickened and there was concomitant thickening of the rostral mandible and calvarium. The dog exhibited signs of resentment on palpation of the mandible and signs of pain were elicited on flexion and extension of both elbow joints. No signs of pain were evident on palpation of the ulnas or calvarium. Radiographic images of both ulnas showed marked amorphous periosteal new bone formation. The distal ulnar physes were closed centrally and both elbow joints had humeroulnar subluxation. Radiographic changes to the calvarium and mandibular rami were consistent with a diagnosis of craniomandibular osteopathy. A bilateral ulna ostectomy was performed to correct the angular limb deformity and elbow subluxations. Histology of the ostectomised pieces showed changes consistent with craniomandibular osteopathy.

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Introduction
Craniomandibular osteopathy is a proliferative bone disease of young dogs characterized by irregular new bone formation and was first reported in West Highland White Terriers in 1958 (1). Primarily the bones of the skull are affected although occasionally the condition extends to the metaphyses of long bones, in particular the femur, radius and ulna (2–4). Although it is reported in a number of breeds, the West Highland White Terrier is overrepresented, and in this breed an autosomal recessive mode of inheritance has been suggested (4, 5). The occurrence of the disease in a number of non-terrier breeds, however, may indicate that other factors may be involved such as bacterial (Escherichia coli) or viral (canine distemper virus) infections (2). In a recent study of canine leukocyte adhesion deficiency syndrome in Irish Setters, seven out of 12 dogs had radiographic signs consistent with craniomandibular osteopathy (6).

As the condition is diagnosed primarily in the West Highland White Terrier and because of the clinical appearance of affected dogs, it is commonly referred to as ‘Westie disease’ or ‘lion jaw’. Swelling of the jaw, inappetence, pyrexia and lethargy are common manifestations of this condition (3). Craniomandibular osteopathy is characterized by a non-neoplastic, bilaterally symmetrical irregular proliferation of the affected bones. The disease affects skeletally immature dogs between three to eight months of age and cases are usually presented with signs of dysphagia, recurrent fever or lethargy (2). Signs of pain may be very severe when the mouth is fully opened. The disease is usually self-limiting and the irregular bony proliferations remodel with time. However in severe cases, ankylosis of the temporomandibular joint may occur (3).

Craniomandibular osteopathy has been compared to infantile cortical hyperostosis (Caffey’s disease) in humans and familial infantile cortical hyperostosis in rhesus monkeys (7, 8). The histopathological changes consist of a series of distinct processes (3). Initially there is osteoclastic resorption of the lamellar bone followed by replacement with primitive coarse bone that extends beyond the periosteal boundaries. The mosaic pattern of this new bone formation is consistent with sporadic and rapid resorption and deposition of new bone. The normal bone marrow spaces are replaced with a highly vascular fibrous stroma. Inflammatory cells, mainly lymphocytes, neutrophils and plasma cells, invade at the periphery of the new bone deposition. These changes may sometimes resemble forms of osteosarcoma, hyperparathyroidism or callus formation. Osteitis deformans (Paget’s disease) in humans and fibrous dysplasia have a similar histological
appearance although the location of lesions and age predisposition differ from craniomandibular osteopathy (3, 4).

There are a few reports of long bone involvement in dogs with craniomandibular osteopathy in the veterinary literature but it has not, to the authors’ knowledge, been reported as a cause of angular limb deformity (3, 4, 9, 10). Angular limb deformities in the thoracic limb are usually caused by premature closure or restriction of growth of a physis, most commonly the distal ulna physis, and are characterized by shortening of the limb, deviation of the paw and deformities of the carpus and elbow (11). This report describes a case of craniomandibular osteopathy with concurrent ulna involvement that resulted in a bilateral carpal valgus deformity.

**Case history**

A four-month-old entire female West Highland White Terrier was presented to the Small Animal Teaching Hospital at the University of Liverpool for investigation into the complaint of a shifting bilateral thoracic limb lameness of three weeks duration. The lameness was progressive and had deteriorated after exercise or long periods of rest. The owner also reported that the shape of the dog’s head was becoming more domed and the dog was lethargic. A reduced appetite over the previous two weeks was also reported. Treatment with non-steroidal anti-inflammatory drugs was halted as it had resulted in haematochezia. Physical examination revealed a quiet, alert and responsive dog. The lameness was graded at 4/10 on the left thoracic limb at a walk (12). There was thickening of the rostral mandibular rami and the calvarium had a more domed appearance than normal. Signs of pain were elicited by palpation of the mandible and on opening of the mouth. There was a bilateral carpal valgus with the left thoracic limb more affected (Fig. 1). The ulnas were grossly thickened distally and signs of pain were elicited by flexion and extension of both elbows.

Radiographs of the left thoracic limb demonstrated marked expansion and increased opacity of the distal ulna diaphysis and a patchy, ill-defined increase in opacity of the metaphysis (Fig. 2 and 3). Well-defined paraperiosteal new bone extended along the caudal aspect of the metaphysis and physis to 4 mm distal to the distal ulnar growth plate. Paraperiosteal new bone was also evident along the cranial aspect of the metaphysis (Fig. 2). The medial aspect of the distal ulna metaphysis formed a smooth bulge dorso-medially, which appeared to bridge the medial aspect of the growth plate. The central and medial part of the distal ulna physis appeared closed and the radius was bowed cranially and laterally. Radiographs of the right limb showed similar, but less pronounced changes. There was not any evidence of distal ulna growth plate bridging on any projections of this limb, but again, the central part of the distal ulna physis appeared closed and there was cranial bowing of the radius. In both limbs, the distal radial physes were narrowed at the lateral aspect and slightly flared medially. Bilateral carpal valgus was evident, which originated from this level; both limbs were deviated laterally.

**Fig. 1** Bilateral carpal valgus due to bridging of the distal ulna growth plate.

**Fig. 2** Mediolateral radiograph of the left antebrachium demonstrating a profuse periosteal reaction consistent with craniomandibular osteopathy. Paraperiosteal new bone was also evident along the cranial aspect of the metaphysis (arrow).

**Fig. 3** Craniocaudal radiograph of the left antebrachium.
by approximately 20° (13). Humeroulnar subluxation was also evident bilaterally (Fig. 4).

The bones of the calvarium (Fig. 5) and the mandibular rami (Fig. 6) were thickened, irregular and increased in opacity. The temporomandibular joints and tympanic bullae were normal.

A bilateral 2.5 cm ulna ostectomy was performed via an approach to the distal ulna (14). The adjacent periosteum was removed along with the ostecotomized section. A free autogenous fat graft was not placed. Postoperative radiographs demonstrated a bilateral radiohumeral subluxation, with cranial displacement of the radial head with respect to the humeral condyles (Fig. 7). The proximal point of the ostectomy was 5 mm distal to the medial coronoid process with the distal osteotomy 5 mm proximal to the distal ulna physis.

The limbs were placed in support dressings for four days and analgesia was provided by the administration of a 0.2 mg/kg morphine sulphatea - lidocaineb - ketaminec infusion. The dog was discharged to the owners after four days with instructions to administer carprofend at a dose rate of 2 mg/kg twice daily.

Histological examination of the ostecotomized ulnae showed irregular numerous peripherally-radiating bony trabeculae composed predominantly of woven bone. Trabeculae displayed surface scalloping overlain by osteoclasts. A mosaic pattern of cement lines (reversal lines) were present in the trabeculae. The medullary spaces contained moderate numbers of loosely packed, disorganized fibroblasts, neovascular blood vessels and small fragments of necrotic bone. Occasional lymphocytes, plasma cells and neutrophils were present in areas of active osteogenesis. Evidence of an infectious or neoplastic process was not found (Fig. 8). These findings, in conjunction with the osseous changes of the skull, were most compatible with a diagnosis of craniomandibular osteopathy (15).

At suture removal ten days later the dog was able to ambulate without any apparent difficulty. Radiographic images taken four weeks postoperatively demonstrated bilateral reduction of the radiohumeral subluxation. The proximal ulna had rotated cranially so that the anconeous was cranially displaced and the coronoid was positioned at the level of mid-humeral condyle in both elbow joints. The distal ulnar sections however appeared unchanged from the previous radiographs with marked expansion, increased opacity and parosteal new bone. The left distal ulna growth plate still appeared ‘bridged’ as previously, and the distal ulna growth plates still appeared closed centrally on both limbs. Carpal valgus remained bilaterally.

At four months post-surgery, radiographs demonstrated a bilateral reduction in the cranial rotation of the proximal ulna, so that in both limbs, the coronoid processes were now only 2 mm proximal to the radial head. Faint mineral opacities were evident in the soft tissues distal to this region. Both distal ulna sections had become markedly radiolucent, with thin, faint cortices and absent trabecular pattern. The right distal ulna growth plate remained closed, but the left appeared to have re-

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a Morphine: CP Pharmaceuticals, Wrexham UK
b Lignocaine: Hameln Pharmaceutical Ltd, Gloucester UK
c Vetalar: Pfizer, Sandwich, UK
d Rimadyl: Pfizer, Sandwich, UK
lateral radial osteotomies were performed and the distal limbs were inwardly rotated through 24°. The degree of rotation was assessed visually by sequentially rotating the distal part of the frame until the elbow and carpi were in the same axial plane. The osteotomies were stabilized with circular fixators. Postoperative radiographs showed a satisfactory correction of the valgus although a mild valgus, centred at the carpal level remained. Radiographic evaluation six weeks later confirmed satisfactory healing of the osteotomy and the circular fixators were removed. Radiographs of the skull were not repeated, but there was complete resolution of the mandibular pain. A telephone follow-up call performed 14 months after surgery revealed a satisfactory clinical outcome. Stiffness after long exercise was noted by the owner as well as low grade lameness after exercise of one hour duration or more. A mild carpal valgus remained. According to the owners, the domed appearance to the head remained unchanged but the dog’s demeanour and appetite had returned to normal.

Discussion

Lameness and limb swelling have been reported in West Highland White Terriers with craniomandibular osteopathy with extraosseous bone adjacent to the radius and ulna preceding mandibular changes by up to 60 days (3). Changes to the tympanic bullae are often noted in cases of craniomandibular osteopathy but were absent in this case. In this case, the changes to the mandible were relatively minor compared to those of the antebrachium implying that the limb changes may have occurred first. Although long bone involvement is reported in cases of craniomandibular osteopathy, to the authors’ knowledge this is the first report of craniomandibular osteopathy causing an angular limb deformity (3, 4, 9, 10).

Treatment options for craniomandibular osteopathy are usually supportive and symptomatic. Anti-inflammatory drugs are the treatment of choice, with prednisolone being the most effective (16). It may be necessary to liquidize the food in order to reduce the pain associated with mastication. Although the appearance of clinical signs may be cyclical and vary in severity, they usually resolve by about 12 months of age and the prognosis for the disease is usually good unless there is involvement of the temporomandibular joint. In such cases, the prognosis is guarded and excision arthroplasty of the affected temporomandibular joint(s) may be indicated.

Growth deformities of the antebrachium frequently occur because of the paired bone system and the unique shape of the distal ulna physis. The conical shape of this physis predisposes it to premature closure (17). Any lateral force at the level of this physis is converted to a compressive force and may result in a Salter Harris type V injury because of its unique shape (18, 19). As 85% of the ulna growth is from this physis, premature closure results in restriction of longitudinal growth of the radius (bow string effect). The result is radial shortening with cranial bowing, external rotation of the paw, valgus deformity and elbow subluxation (17, 19–21). Surgery is aimed at correcting the subluxation and removing the restriction on the radius to allow asymmetric physeal growth to correct the deformity (22). Following a partial ulnar ostectomy, the ulna may quickly heal and further deformity will occur if the animal continues to grow (23, 24). Although autogenous fat grafts may retard or inhibit ulna healing, removal of the periosteum...
has the greater retarding effect on ulna healing (24, 25). In one study of twelve dogs, removal or suturing of the periosteum over the ends of the ostectomy site prevented healing of the ulna and resulted in correction of the radius curvus deformity (26). Abnormal loading of the carpal bones during their development may lead to an angular limb deformity that is not susceptible to correction by realignment of the antebrachium (27). In this case the proximal and distal joints of the antebrachium were approximately parallel although a mild valgus was still present due to deformities within the carpus.

The histopathological diagnosis was consistent with exostosis, osteoma or a proliferative bone production process secondary to periosteal proliferation and lifting. Although this assessment was not pathognomonic for craniomandibular osteopathy, it was considered to be the most likely cause given the signalment, history, and clinical findings in this case. In cases of craniomandibular osteopathy, histopathological examination reveals that normal lamellar bone is replaced by an enlarged, coarse fibre (woven) bone. The bone marrow is replaced by a fibrous-type stroma with foci of inflammatory cells (3). Bone is relatively limited in its range of responses to injury and different diseases resulting in similar radiographic and histological changes (3). Although it is possible that the angular limb deformities may be due to reasons unconnected to the craniomandibular osteopathic changes, this was considered to be unlikely given the bilateral involvement and the apparent lack of trauma. Having identified bridging on radiographs of the left limb, it is possible that similar bridging was present on the right limb at some point previously that temporally closed the growth plate and brought about the deformity.

The reason for the postoperative radiohumeral subluxation is not clear. Stretching of the ligamentous support to the elbow, in particular the anular ligament, is a possibility as is transection of the anular ligament at the time of the osteotomy. The reduction of the subluxation in the four week postoperative radiographs with caudal rotation of the proximal ulna section supports the former. The proximal position of the cut was also relatively proximal and that may have contributed to the incongruency although resolution of the incongruency despite a lack of healing of the osteotomy does not support this. The position of the cut was however as described by some authors at approximately the level of the radial head and finishing above the interosseous ligament but others suggest aiming about 1 cm distal to the medial conoid process with a 30° caudoproximal craniodistal direction of cut (28, 29).

Choice of treatment of angular limb deformities is dependent upon the remaining potential for growth. Ulnar osteotomy alone is a valid technique in young dogs that still have a relatively long period of growth remaining (30). In this case, this failed to address the deformity so a further surgery performed. Acute corrections are reported in the literature using circular fixators, compression plates and external fixators, whereas the choice of technique depends upon clinical findings and surgeon preference (13, 21, 31, 32). The clinical evidence in this case suggested that most of the remaining deformity was due to an external rotation of the manus rather than a deformity within the long bones. Due to the shape and size of the distal radius and nature of the correction required it was decided that a circular fixator was the most appropriate method of correction in this case.

Conflict of interest
None declared.

References


