CASE REPORT

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Dropped-head syndrome in a dog secondary to myopathy of the cervical extensor muscles

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Abstract

A 4-year-old, male, entire Newfoundland was presented for investigation of an acute onset cervical ventroflexion and pyrexia, 4 days following a hip replacement surgery. Neurological examination confirmed prominent cervical ventroflexion characterised by an inability to raise the head, without evidence of cervical hyperaesthesia or associated neurological deficits. Magnetic resonance imaging identified focal, bilaterally symmetrical changes to the semispinalis cervicis musculature, responsible for elevation of the head. The presentation and diagnostic imaging findings appeared analogous to droppedhead syndrome in humans. *Toxoplasma* IgG antibodies were found to be mildly elevated, and therefore treatment with clindamycin was initiated. The dog responded well to treatment and was reported to be clinically normal 2 months thereafter.

BACKGROUND

Dropped-head syndrome (DHS) is a rare condition in humans.¹ This term was introduced to describe the clinical features associated with a myopathy involving the cervical extensor muscle.² DHS is characterised by weakness of the cervical extensor muscles, particularly the semispinalis cervicis muscles, resulting in an inability to elevate the head.^{3,4} The condition can occur as an isolated clinical finding or as part of a generalised neuromuscular disorder.⁵ DHS has been reported to occur secondary to a wide range of conditions, including myasthenia gravis,^{6,7} polymyositis,^{3,8} cervical myelopathies⁹ and myopathies involving the neck extensor muscles.^{10–12}

Reluctance to elevate the head due to cervical hyperaesthesia is a common presentation in dogs^{13,14}; however, cervical ventroflexion secondary to an inability to elevate the head is scarcely reported.¹⁵ Cervical ventroflexion is commonly encountered in cats associated with neuromuscular disorders¹⁶ and electrolyte imbalances.¹⁷ This species difference is thought to relate to the biomechanical properties of the nuchal ligament in the dog.¹⁸ Inflammatory myopathies affecting the cervical region, associated with tetraparesis have been previously reported.^{19,20} To the authors' knowledge, a focal myopathy of the cervical extensor muscles has not be previously documented in the veterinary literature. We describe a case of cervical ventroflexion in a dog associated with a focal myopathy of the cervical extensor muscles, analogous to presentations of DHS in humans.

CASE PRESENTATION

A 4-year-old, male, entire Newfoundland was presented for acute onset cervical ventroflexion without associated hyperaesthesia. Four days before presentation, the dog had undergone a left-sided total hip replacement surgery with mesenchymal stem cell and platelet-rich plasma administration, for the treatment of hip dysplasia at another referral centre. Before surgery, the dog was reported to be in good health, apart from a chronic grade 3/10 left pelvic limb lameness treated with ongoing firocoxib (Previcox 3.4 mg/kg orally [PO] once daily). The general anaesthetic and surgical procedure were reported to be routine, and the dog was discharged following 72-hour hospitalisation. During the subsequent 24 hours, the dog was noted to be lethargic and inappetent, with evidence of low head carriage.

On presentation, general physical examination revealed evidence of tachycardia (144 beats per minute) and marked pyrexia (41.1°C), but was otherwise unremarkable. Prominent cervical ventroflexion with an inability to raise the head was noted (Figure 1), without evidence of cervical hyperaesthesia or associated deficits on neurological examination. The neurological localisation was therefore focal neuromuscular system. Potential differential diagnoses included focal neuropathy, myopathy or myositis, focal myasthenia gravis, electrolyte abnormality, including hypokalaemia or hypocalcaemia, and less likely toxicity such as organophosphate.

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INVESTIGATIONS

Haematology revealed a mildly decreased haematocrit (33.1%, reference interval: 37%–55%), and biochemistry revealed mild hypoalbuminaemia (25.9 g/L, reference interval: 26.3–38.2 g/L), mild hypocalcaemia (1.82 mmol/L, reference interval: 2.18–2.79 mmol/L), mild hyperbilirubinaemia (5.4 μ mol/L, reference interval: 0.1–4.2 μ mol/L), mildly increased alanine aminotransferase activity (155.7 mmol/L, reference interval: 19.8–124 mmol/L), moderately increased creatine kinase activity (8343 U/L, reference interval: 67–446 U/L) and mildly increased C-reactive protein activity (50.8 mg/L, reference interval: <10 mg/L).

Point-of-care ultrasound examination of the thorax and abdomen was unremarkable. General anaesthesia was performed for advanced imaging to investigate for an underlying cause of the pyrexia. Computed tomography of the cervical region, thorax and abdomen (Canon Aquilion ONE Genesis Edition, Canon Medical Systems, UK) was performed before and after administration of iohexol (600 mgI/kg intravenously [IV]). The dog was premedicated with methadone (Synthadon; Animalcare, UK; 0.1 mg/kg IV). Anaesthesia was induced with propofol IV (PropoFlo Plus; Abbott, UK) administered until the trachea was intubated with a 14mm internal diameter cuffed endotracheal tube (Flexicare Medical, UK). Anaesthesia was maintained with sevoflurane (SevoFlo; Zoetis, UK) to an end-tidal of 1.5%-1.8%, carried in oxygen through a circle breathing system (Intersurgical, UK). Computed tomography revealed an incidental peritoneopericardial diaphragmatic hernia, with involvement of the greater omentum and changes consistent with the recent total hip replacement surgery. No cause of cervical ventroflexion was evident.

Blood and urine cultures were performed, revealing no bacterial isolates. Repeat haematology and biochemistry, 3 days following the onset of symptomatic treatment, documented improving levels of creatine kinase (1085 U/L, reference interval: 67–446 U/L) and C-reactive protein (23.3 mg/L, reference interval: <10 U/L).

Five days following initial presentation, a magnetic resonance imaging (MRI) examination of the cervical area was performed under general anaesthesia using a 1.5T Philips Achieva MRI unit with a dedicated spinal coil. The dog was premedicated with butorphanol (Dolorex; MSD Animal Health, UK; 0.2 mg/kg IV) and otherwise the anaesthetic protocol used was analogous to that used previously. Sequences obtained: sagittal T2-weighted (T2W), short T1 inversion recovery (STIR), T1-weighted (T1W) and contrast-enhanced T1W; dorsal T2W and contrast-enhanced T1W, as well as transverse T2W, T1W, contrast-enhanced T1W and a 3D balanced turbo gradient echo (Philips' BaltGRAD) sequence. The multiple sequences demonstrated a bilateral, marked and relatively homogeneous increase in T2W and STIR signal of the semispinalis cervicis musculature over the lamina of C3 and extending caudally over the cranial half of C4 (Figures 2-4, arrows). The changes were predominantly bilaterally symmetrical, but extended further caudally to the level of C5 on the left side (Figure 2, caudal arrow). The volume of the muscles was not altered. No signal change was visible on T1W images,

LEARNING POINTS/TAKE-HOME MESSAGES

- Focal myopathy of the cervical extensor musculature, particularly the semispinalis cervicis muscle, can result in cervical ventroflexion with inability to elevate the head.
- The presentation and diagnostic imaging findings can appear analogous to dropped-head syndrome in humans.
- Toxoplasmosis was suspected on the basis of elevated *Toxoplasma* antibodies and a clinical response to clindamycin treatment; however, idiopathic or non-infectious inflammatory causes cannot be excluded.

but the T2W and STIR changes were matched by marked contrast enhancement (Figures 2 and 3). Other findings included degeneration of the nucleus of the C4–C5 intervertebral disc and a marked generalised cervical oesophageal distension with gas. On the basis of the MRI findings, serology for infectious diseases was performed. *Neospora* serology was negative with IgG titres less than 100; however, *Toxoplasma* IgG antibodies were positive at 1/64. Due to miscommunication with the diagnostic laboratory, concurrent IgM antibodies were not tested on this sample.

DIFFERENTIAL DIAGNOSIS

In light of a suspected inflammatory myopathy on MRI, in addition to the presence of pyrexia, infectious causes were considered including *Toxoplasma* and *Neospora*. A non-infectious inflammatory myopathy, similar to isolated neck extensor myopathy (INEM) in humans, was not discounted. An ischaemic myopathy, secondary to positioning during general anaesthesia, was considered less likely given the bilateral symmetrical nature of the lesion and marked pyrexia.

TREATMENT

Symptomatic treatment was initiated during hospitalisation, while urine and blood culture results were pending, including cephalexin (Cephacare 15 mg/kg PO twice daily), paracetamol (10 mg/kg PO twice daily) and firocoxib (Previcox 3.4 mg/kg PO once daily). Despite resolution of pyrexia, the dog's cervical ventroflexion remained unchanged. Given the elevated *Toxoplasma* IgG antibodies, clindamycin (Antirobe 18.1 mg/kg PO twice daily) treatment was commenced, which correlated with a marked improvement of the clinical signs over the subsequent days. The option of performing surgical biopsy for histopathology was discussed, but declined by the owners in light of the clinical improvement with the addition of clindamycin treatment. The dog was discharged subsequently with ongoing courses of clindamycin, paracetamol and firocoxib.



FIGURE 1 Photographs demonstrating the posture of the patient on presentation (left image) compared to re-examination at 4 weeks (right image).

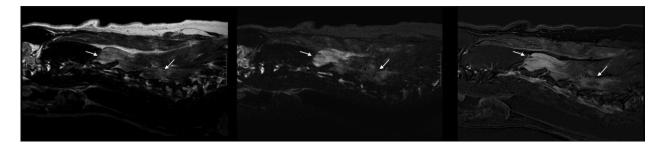


FIGURE 2 Matching sagittal (to the left of midline) T2W, STIR and post-processing subtraction (left central and right panel, respectively) images of the cervical region, demonstrating the cranial and caudal muscular signal changes (arrows).

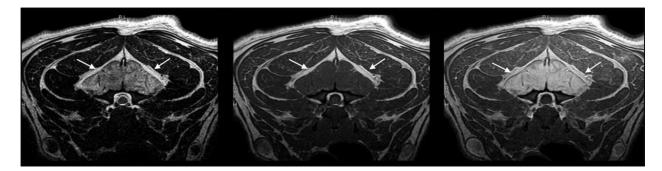


FIGURE 3 Transverse T2W, T1W and post-contrast T1W (left, central and right panel) images at the level of C3–C4 intervertebral disc space, demonstrating the bilateral distribution of the signal changes (arrows).

OUTCOME AND FOLLOW-UP

On re-examination 4 weeks later, the owners reported progressive improvements in the dog's posture, with no evidence of cervical ventroflexion. Neurological examination revealed no deficits with normal head carriage but a mild lameness of the operated leg (Figure 1). *Toxoplasma* IgG antibody levels remained positive at 1/64, but given the clinical improvement, treatment was discontinued. A telephone update 2 months following discontinuation of treatment, confirmed no recurrence of the clinical signs.

DISCUSSION

This case report highlights a focal bilateral myopathy of the semispinalis cervicis musculature in a dog, resulting in marked ventroflexion and inability to raise the head. Inter-

estingly, this clinical presentation shares marked similarities to DHS described in humans. This syndrome is characterised by weakness of the cervical extensor muscles, leading to a failure to elevate the head and a chin-on-chest posture.³ The semispinalis cervicis muscle is a major cervical extensor muscle, and therefore dysfunction of this muscle, as seen in the current case, is attributed as the major cause of DHS in humans.⁴ In a systematic review of presentations of DHS, almost one third of presentations were the result of an INEM as seen in the current case.²¹ The term INEM was developed to describe such focal neck extensor weakness in the absence of generalised neuromuscular disorders.¹² The aetiology of INEM is idiopathic; however, reports of response to immunosuppressive treatment challenge the acceptance that the condition is a non-inflammatory myopathy.²² INEM is frequently a slowly progressive condition; however, presentations with an acute onset and spontaneous remission are reported.²³

3 of 5



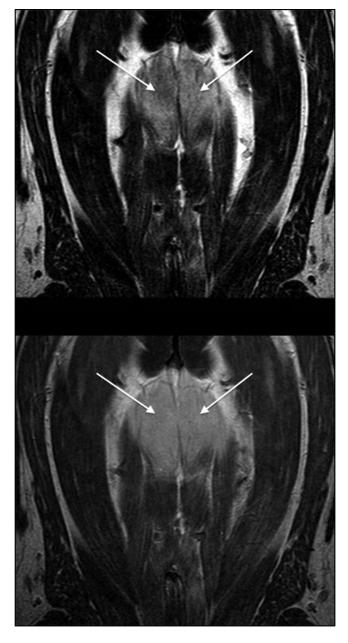


FIGURE 4 Dorsal T2W and post-contrast T1W (top and bottom panel) images at the mid cervical level, demonstrating the bilateral symmetric distribution (arrows) of the cranial muscle changes with T2W signal change and contrast enhancement.

On MRI, the muscular lesions were interpreted as most likely associated with inflammation, given that processes such as denervation injury would not have been expected to cause signal alteration during such a short time frame.²⁰ Ischaemic muscle damage secondary to surgical positioning, comparable to post-anaesthetic myopathy in horses or compartment syndrome in humans, could not be excluded given the size of the dog. The surgical anaesthetic was stable with no significant fluctuations in blood pressure that could predispose to such injury.²⁴ The dog was maintained in lateral recumbency for the surgical procedure, and therefore a focal, bilateral ischaemic injury to the cervical extensor muscles without involvement of the triceps and gluteal muscles, as is common in horses, was considered less likely.²⁵

In light of a suspected inflammatory myopathy on MRI, in addition to the presence of pyrexia, infectious causes were investigated. Infectious disease testing revealed evidence

of elevated Toxoplasma IgG antibodies, with the tentative diagnosis supported by a clinical response to clindamycin treatment.²⁶ Repeat IgG antibodies at 4 weeks demonstrated an unchanged titre. A four-fold increasing titre over 2-5 weeks is generally accepted as being suggestive of the diagnosis.²⁶ Due to miscommunication with the diagnostic laboratory, IgM antibodies were not performed on the initial blood sample. The absence of IgM antibody and PCR testing unfortunately limits the ability to confirm or refute toxoplasmosis as the underlying cause. Given the clinical response to clindamycin treatment, the owner declined surgical biopsy for histopathology to be performed, which could have yielded further diagnostic information. Electrodiagnostic testing could have been considered in this patient. Evidence of focal spontaneous electrical activity of the cervical extensor muscles on electromyography, in the absence of generalised muscle changes, could further support the hypothesis of INEM. Despite clinical response to treatment, it is challenging to know whether the antibiosis or anti-inflammatory treatment and time led to resolution of the clinical signs.

This case report describes the imaging features of a focal, presumed inflammatory myopathy of the cervical extensor muscles with a clinical presentation akin to DHS in humans. This presentation is, to the authors' knowledge, previously unreported in the veterinary literature.

AUTHOR CONTRIBUTION STATEMENT

Nicholas Grapes was the primary clinician, supervised by Steven De Decker and Alberta De Stefani. Francisco Llabres-Diaz was involved in the review and reporting of the diagnostic imaging studies. All authors were involved in the writing and review of the manuscript.

CONFLICT OF INTEREST STATEMENT

The authors declare they have no conflicts of interest.

ETHICS STATEMENT

This work involved a client-owned animal, and high standards of individual veterinary clinical patient care were followed. Informed client consent was obtained for treating the patient. Ethical approval from a committee was therefore not specifically required for publication.

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