MULTIMODALITY FEATURES OF BECKER-LIKE MUSCULAR DYSTROPHY IN A YOUNG DOG

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Introduction

In humans, Becker muscular dystrophy (BMD) is an x-linked recessive genetic neuromuscular disorder due to mutations in the gene coding for dystrophin leading to severe muscular weakness and gait abnormalities in juvenile age.

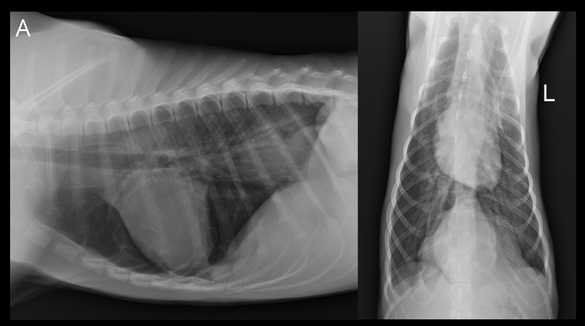

FIGURE 1 A. Thoracic radiographs (left lateral and dorsoventral views). B. Computed tomography images. There is asymmetry of the diaphragmatic profile with an undulating and irregular appearance associated with herniation of the gastric fundus into the caudal esophagus through the esophageal hiatus.

Materials and methods

An 8-month-old, male, mixed-breed dog was presented for macroglossia, decreased mandibular extension, ptyalism, dysphagia and regurgitation. Serum creatine kinase activity was severely elevated. Thoracic radiographs were performed to exclude aspiration pneumonia. The dog underwent magnetic resonance imaging (MRI) of the head to assess the suspected underlying myopathy. Whole body computed tomography (CT) was performed to rule out other musculoskeletal abnormalities. Tricipital and bicipital muscular biopsies were taken to characterize the myopathy.

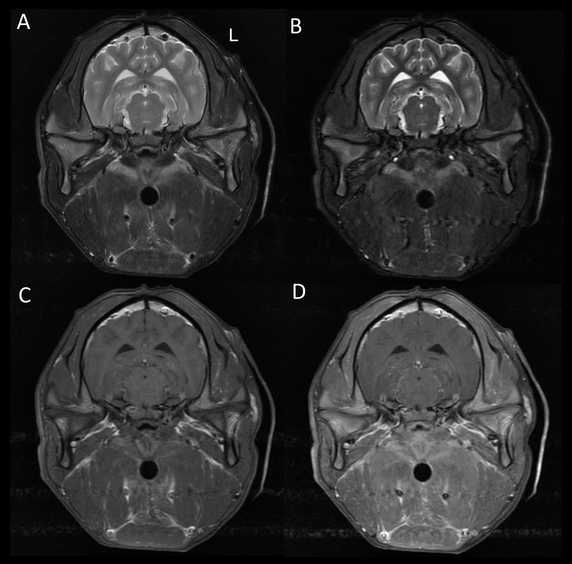

FIGURE 2 MRI of the head. A, Transverse T2w; B, STIR; C, T1w pre-contrast; D, T1w post-contrast images with a 3 mm slice thickness. There is bilateral and symmetric increase in volume of the geniohyoid and mylohyoid muscles and moderate bilateral hypotrophy of the masseter and temporal muscles. All the affected muscles are heterogeneously hyperintense on T2w and STIR sequences and heterogeneously enhancing in the T1w post-contrast sequence.

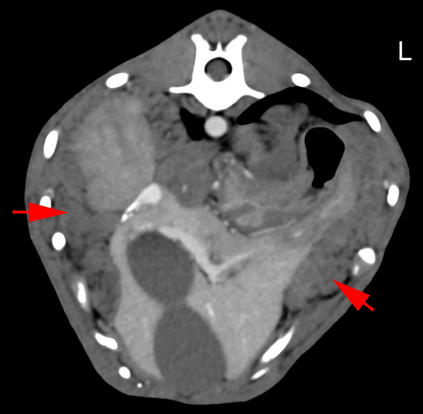

FIGURE 3 Computed tomography. The diaphragm is diffusely thickened, asymmetric with a marked undulating appearance and slight heterogeneous enhancement (arrows). The thickened diaphragm causes compressive impression on the liver parenchyma which preserves normal architecture.

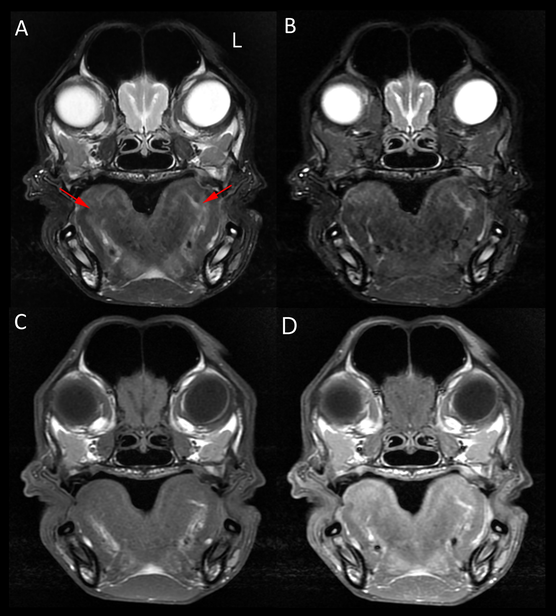

FIGURE 4 MRI of the head. A, Transverse T2w; B, STIR; C, T1w pre-contrast; D, T1w post-contrast images with a 3 mm slice thickness. The tongue is markedly and symmetrically enlarged obliterating most of the oral cavity, partially folded in a "'V-shape" (arrows).

Results

Thoracic radiographs showed a gastro-esophageal hiatal hernia, thickening and asymmetry of the diaphragm. MRI showed a severely and symmetrically enlarged tongue obliterating most of the oral cavity, bilateral and symmetric increase in volume of the geniohyoid and mylohyoid muscles and hypotrophy of the masticatory muscles. The affected muscles were heterogeneously hyperintense on T2-weighted and STIR sequences, enhancing in the T1-weighted post-contrast series. There was secondary bone remodeling of both the mandibular bodies and of the hyoid apparatus. No additional abnormalities were noted on CT. Muscular histopathology was consistent with BMD.

Conclusion

Multimodality imaging has been helpful for a complete characterization of the musculoskeletal abnormalities secondary to BMD. In young patients with dysphagia, mastication and swallowing difficulties, muscular dystrophy should be considered as a differential diagnosis.

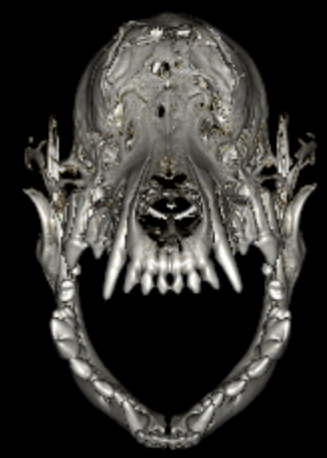

FIGURE 5 Three-dimensional reformatted computed tomography. There is secondary bone remodeling of the mandibular bodies causing severe widening of the intermandibular space as a possible consequence of an abnormal growth of the tongue and extra-lingual muscles.