Spongiform leucoencephalomyelopathy in border terriers: clinical, electrophysiological and imaging features

R. Gutierrez-Quintana, M. McLaughlin, L. Grau Roma, G. Hammond, A. Gray, M. Lowrie

Introduction
A novel spongiform leucoencephalomyelopathy causing a shaking puppy phenotype was reported in border terrier puppies in 2012, but no information regarding clinical progression, diagnostic imaging or electrophysiological findings is currently available. Therefore, the aim of this study was to describe the clinical, electrophysiological and MRI features of this canine leucoencephalomyelopathy and compare them with human white matter disorders.

Case presentation
Seven border terrier puppies (six males and one female) from four different and unrelated litters were presented to two referral hospitals. In all cases, the rest of the littersmates and both parents were reported to be completely normal.

The owners reported generalised coarse body tremors that started at around three weeks of age. The tremors continued progressing, and all the puppies had to be helped to eat. All affected puppies were presented for examination at around six weeks of age, and physical and neurological examinations were performed.

MRI revealed bilateral and symmetrical T2-weighted hyperintensities affecting the brainstem and cerebellar white matter (Fig 1). Histological examination of the brain and spinal cord revealed decreased myelin content and spongiform changes affecting the white matter of the cerebellum, brainstem and spinal cord.

Interpretation
Although the clinical, brainstem auditory evoked response and MRI findings were consistent in all cases, the results of this study should be interpreted with caution due to the small number of cases.

Results
Four of the puppies were euthanased. The remaining three puppies showed gradual deterioration until four months of age, and then slowly started improving over several months. Brainstem auditory evoked response revealed a normal wave I, a reduction in the amplitude of wave II and an absence of waves III to VII, which is compatible with a brainstem disorder (Fig 1).

The leucoencephalomyelopathy affecting border terriers has a pathognomonic clinical presentation with defining MRI and electrophysiological characteristics.

Some dogs can show gradual improvement of the clinical signs after four months of age.

Significance of findings
The findings of this study show that some border terriers with a shaking puppy phenotype can show gradual improvement of the clinical signs. They also demonstrate that MRI and brainstem auditory evoked response can be useful to further characterise white matter disorders in dogs.

R. Gutierrez-Quintana, M. McLaughlin, G. Hammond, A. Gray, School of Veterinary Medicine, University of Glasgow, Glasgow, UK
L. Grau Roma, School of Veterinary Medicine and Science, University of Nottingham, Loughborough, UK
M. Lowrie, Neurology Service, Dovecote Veterinary Hospital, Derby, UK
email: Rodrigo.gutierrezquintana@glasgow.ac.uk
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Fig 1: (a) Brainstem auditory evoked response of an affected border terrier puppy and (c) a normal puppy of the same age. (b) Sagittal T2-weighted MRI image of an affected border terrier puppy and (d) a normal puppy of a similar age – note the T2-weighted hyperintensities in the brainstem and cerebellar white matter (white arrows) and the thinning of the caudal part of the corpus callosum (black arrows)