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Epidemiology of equine motor neuron disease

HO Mohammed *, JF Cummings, TJ Divers, R de la Rua-Domenech, A de Lahunter

College of Veterinary Medicine, Cornell University, Ithaca, NY 14853, USA

Summary — Equine motor neuron disease (EMND) is a newly recognized neurodegenerative disorder of bulbospinal motor neurons in the horse. We conducted a case-control study to identify intrinsic factors associated with the risk of this disorder. Seventy-four cases and 160 controls were assembled. Controls included horses diagnosed with equine protozoal myelitis, equine degenerative myeloencephalopathy, and cervical stenotic myelopathy during the same time period as the cases. Logistic regression analysis was used to evaluate the association of each hypothesized factor while simultaneously controlling for the effect of other factors. Factors found to be significantly associated with the risk of the disease were breed and age of the horse. Quarterhorses are at a higher risk in comparison to other breeds of horse.

INTRODUCTION

Equine motor neuron disease (EMND) is a newly recognized neurodegenerative disorder of horses (Cummings et al, 1990). EMND is characterized by weight loss (despite a good appetite), muscle atrophy, and a generalized weakness. Affected horses had clinical findings of a short-strided gait, lowered head and neck, frequent shifting of the back limbs, excessive recumbency, muscle fasciculations and trembling (Divers et al, 1992). The weakness, weight

* Correspondence and reprints
loss, and muscle wasting develop progressively over several months in these afebrile horses who characteristically stand with their limbs drawn under their body. Affected horses are incapable of sustained exercise, which intensifies signs of weakness (Divers et al, 1992). The progression of weakness and wasting at some point has arrested or abated in several of the affected horses (Cummings et al, 1990; Divers et al, 1992).

Serum creatine kinase (CK) and aspartate aminotransferase (AST) levels are mildly to moderately elevated in EMND horses. Cerebrospinal fluid (CSF) samples often contain protein levels that are increased slightly with evidence of intrathecal IgG production (Divers et al, 1992). Electromyography typically reveals widespread denervation potentials. Positive-sharp waves are more frequent than fibrillations and pseudomyotonic bursts are infrequent.

Histopathologic studies reveal degeneration and loss of motor neurons in the spinal ventral horns and certain brain stem motor nuclei (ie trigeminal, facial, hypoglossal and nucleus ambiguus). Degenerating neurons are swollen, chromatolytic, and often contain distorted karyolytic nuclei (Cummings et al, 1990). These ghost-like neurons contain increased neurofilaments which are prematurely phosphorylated. Eosinophilic punctate inclusions which simulate the appearance of Bunina bodies often appear in degenerating perikarya (Cummings et al, 1990; 1993). Glial scars consisting of astrocytes, lipofuscin-laden microglia and occasional oligodendrocytes serve as long-term markers at sites of neuron loss. Masses of neurofilaments may also be found in spheroidal swellings along the proximal course of motor axons. Degeneration of myelinated axons in the ventral roots and peripheral nerves occurs in the wake of cell body changes. Degenerative perikaryal changes also occur in the sensory ganglia, but are much less frequent. In those 'burnt-out' cases in which the progression has been arrested for up to 2 yr, small compacted Büngner's bands remain discernible in nerves and roots and lipopigment-laden glial scars persist in the ventral horns.

Angular atrophy of skeletal muscle fibers develops. Muscle fiber typing studies in EMND cases indicate that the antigravity muscles are preferentially affected as Type I fiber atrophy predominates. Because this equine disease is typified by sporadic incidence, progressive weakness and wasting, and degeneration of bulbospinal motor neurons, it has been compared with the human progressive muscular atrophy (Hirano and Kato, 1992).

EMND is distinctly different from other equine diseases known to affect the spinal cord. These diseases include equine degenerative myeloencephalopathy (EDM), cervical stenotic myelopathy (CSM), equine protozoal myeloencephalitis (EPM), equine viral encephalomyelitis, equine rhinopneumonitis, and rabies. Clinical EMND cases have been recently reported in England (Hahn and Mayhew, 1993) and post mortem studies have documented cases in Belgium (Sustronck et al, 1993).

The objectives of this study were: 1) to describe some of the epidemiologic features of EMND; and 2) to identify risk factors associated with the likelihood of the disease and to quantify their risk.

MATERIALS AND METHODS

A case-control study was conducted to identify risk factors associated with likelihood of EMND. Cases consisted of all horses diagnosed with EMND in the United States and Canada. Diagnosis of EMND cases is based on clinical and histopathologic criteria. The clinical criteria were marked weight loss (> 200 lb), distinct muscle atrophy, generalized weakness, and elevated serum AST and/or CK. The histopathological criteria included swollen chromatolytic lower motor neurons, perikaryal and proximal axonal accumulations of neurofilaments, cell body necrosis,
neuronophagia, glial scar formations, motor axon degenerations in the ventral horns and in the small fascicles traversing the ventral funiculus en route to the ventral roots, axonal degenerations and formations of Büngner's bands in the ventral roots and peripheral and cranial nerves (Cummings et al, 1990). In some cases, a biopsy of the spinal accessory nerve was sampled and examined for the axonal changes. Suspected cases that did not satisfy the above-mentioned criteria were excluded from the study.

Controls were selected from horses diagnosed at necropsy as having EPM, EDM, or CSM during the same time period when cases were diagnosed. The control study population was limited to these 3 neurologic diseases to avoid information bias as a result of inaccurate diagnosis.

Data on intrinsic risk factors (age, breed, sex) and geographic origin of all cases and controls were collected. The bivariate association between these hypothesized factors and the risk of EMND was evaluated using the chi-square or Fisher's exact tests in BMDP (Dixon et al, 1992). Odds ratios and 95% confidence intervals were computed. Logistic regression analysis was used to evaluate the significance of each factor on the risk of EMND while simultaneously controlling for the effects of other factors.

RESULTS

Seventy-four cases were diagnosed with EMND since 1985. The majority of the cases appear to cluster in the north-eastern US. Cases usually originate at boarding and hack stables and with the exception of one operation, only one case was identified per stable. Table I shows the results of the bivariate association between breed and the risk of EMND. Quarterhorses, Appaloosa, and Standardbreds horses appear to be at a high risk of developing the disease when compared with the other breeds of horse combined. It also appeared that geldings were at a high risk of EMND when compared to mares.

The results of the logistic regression analysis are shown in table II. Factors that were significantly associated with the risk of EMND were breed and age. Quarterhorses were more likely to develop EMND in comparison to other breeds of horse. Thoroughbred horses retained the tendency of being at a high risk of EMND. No other breed was significantly associated with the likelihood of EMND. Age was significantly associated with the risk of EMND. The risk appeared to increase with age, reach the peak at around 16 yr of age and then decline. The sex of the horse was not associated with the risk of EMND.

DISCUSSION

The sporadic nature, the clinical signs, and the histopathologic findings in EMND bear a striking resemblance to these changes reported in humans affected with sporadic form of amyotrophic lateral sclerosis (ALS)

Table I. The bivariate association between breed of the horse and the risk of EMND.

<table>
<thead>
<tr>
<th>Breed of horse</th>
<th>Cases</th>
<th>Controls</th>
<th>Odds ratio</th>
<th>95% confidence interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Other breeds</td>
<td>15</td>
<td>78</td>
<td>1.0</td>
<td></td>
</tr>
<tr>
<td>Quarterhorse</td>
<td>29</td>
<td>20</td>
<td>7.5</td>
<td>3.2–18.1</td>
</tr>
<tr>
<td>Thoroughbred</td>
<td>18</td>
<td>49</td>
<td>1.9</td>
<td>0.8–4.4</td>
</tr>
<tr>
<td>Appaloosa</td>
<td>7</td>
<td>7</td>
<td>5.2</td>
<td>1.4–19.9</td>
</tr>
<tr>
<td>Arabian</td>
<td>1</td>
<td>8</td>
<td>0.7</td>
<td>0.1–5.9</td>
</tr>
<tr>
<td>Standardbred</td>
<td>4</td>
<td>1</td>
<td>20.8</td>
<td>1.9–157.5</td>
</tr>
</tbody>
</table>
(Hirano and Kato, 1992). The motor neuron changes in EMND appear to be progressive. However, in several cases the disease progressed slowly and then plateaued. These horses remain alive but are weak and wasted. To date, no full recovery in any of the surviving cases has been observed.

Since the initial report on EMND, the number of newly diagnosed cases has increased in the United States and Europe. We are not sure whether we are seeing the tip of the iceberg, where the identified EMND cases are only a small portion of those horses actually afflicted.

The significant association between the breed of the horse and the risk of EMND that we have observed should not be interpreted as a causal association. Despite the strength of association demonstrated in Quarterhorses, there was no familial relationship between any of the identified EMND cases. Furthermore, histocompatibility complex studies on some of the affected horse have not yet revealed any common antigenic markers.

**REFERENCES**


