CASE REPORT

Cauda Equina Neuritis: A Chronic Idiopathic Polyneuritis in Two Horses

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Summary

Two cases of cauda equina neuritis are compared and contrasted. Neurological deficits of the tail and perineum were noted and functional deficits were seen in gait, urination, defecation and cranial nerve function. Lesions consisted of nonsuppurative inflammation of the nerve trunks and proliferation of the perineurium of the cauda equina. Cranial nerve involvement in one case supported a diagnosis of polyneuritis equi rather than cauda equina neuritis. The possible etiologies and pathogenesis of this disease are discussed.

Résumé

Névrite de la cauda equina, une polynévrite idiopathique chronique, chez deux chevaux

Les auteurs présentent les similarités et les différences relatives à deux cas de névrite de la cauda equina. Ils constatèrent un déficit neurologique de la queue et du périnée, ainsi que des irrégularités dans la démarche, la miction, la défécation et le fonctionnement des nerfs crâniens. Les lésions consistaient en une inflammation non suppurante des troncs nerveux et une prolifération du périnèvre de la cauda equina. L’atteinte de nerfs crâniens, dans un des deux cas, justifia un diagnostic de polynévrite équine plutôt que de névrite de la cauda equina. Les auteurs commentent les causes possibles et la pathogénèse de cette maladie.

Introduction

There have been a number of reports of tail and anal sphincter paralysis in horses. This problem is usually the result of chronic inflammation of the extradural root of the cauda equina, with corresponding loss of function in the sacrococcygeal region. Investigators have found inflammatory changes in the cranial and peripheral nerves and have suggested the name polyneuritis equi (1,2,3,4) for this syndrome.

Authors in Germany first described the pathology and clinical findings (5). The disease has since been reported in other parts of Europe (6,7) and more recently in the United States of America (8). The etiology is still unknown. The most favored explanations at present include an immunological response to a persisting herpesvirus infection (9), an allergic neuritis in response to antigen releases by trauma or infection (6), or a postinfectious allergic neuritis, comparable in its pathogenesis and lesions to the Guillain-Barre syndrome (GBS) and its laboratory model, experimental allergic neuritis (EAN) (4).

The following is a report of two cases of this syndrome seen at the Western College of Veterinary Medicine (WCVM), University of Saskatchewan.

Case 1

History — A seven year old crossbred pony mare, used for chuckwagon racing, was presented to the WCVM in the fall of 1980 with a one to two week history of ataxia and a droopy right ear. The mare had received Western and Eastern equine encephalomyelitis and tetanus vaccines six months previously and had undergone a Caslick’s operation two months previously. She had been one of a group of 20 ponies on cereal grain stubble and had been observed infrequently by the owner.

Clinical Findings — The mare exhibited a wide based stance with fore and hindlimb extension and a head tilt to the right. She was depressed, regurgitated water while drinking and ate with difficulty. Unilateral facial paralysis involved the right ear, upper and lower lips and eyelids. There was frequent urination and the bladder was dilated and atonic. The rectum was also flaccid and dilated. Loss of proprioception in conjunction with an abnormal gait, deficits in cranial nerve III, VI and VII were present. The anal sphincter reflex was absent as was perianal sensation and the tail was paralyzed.

A mild neutrophilia was noted on a complete blood count. Total protein and fibrinogen were elevated and serum phosphorus and magnesium concentrations were low. Cerebrospinal fluid (CSF), collected from the lumbosacral region, was yellow, cloudy, contained elevated protein and cell counts, with moderate numbers of neutrophils. Bacteria were not seen (Table 1). Urinalysis was normal.

Postmortem Examination — Mild coning of the cerebellum, opacity of the dura mater, foci of white material overlying the cerebral hemispheres, yellow gelatinous exudate in the third ventricle and edema of the cauda equina were the main gross lesions (Figure 1). Focal irregular areas of erosion of the urinary bladder and dilation of the bladder and rectum was also evident.

Histologically, the severest lesions were in the nerves of the cauda equina and branches of the trigeminal nerves. These were characterized by a non-suppurative inflammation of many nerve fibres with extensive perineurial fibrosis and Wallerian degeneration.
(Table II). The inflammatory infiltrate consisted mainly of lymphocytes with lesser numbers of macrophages, swollen glial cells and foci of neutrophils. Prominent myelinophagia, Wallerian degeneration, perineural, fibrosis and perivascular lymphocytic cuffs were seen in affected areas. Severely involved nerves were adjacent to normal nerves and nerves with small foci of inflammation. Gasserian ganglia were mildly involved as were a few scattered nerve roots in cervical, thoracic, lumbar and sacral cord areas. Calcification of spinal nerve roots and perineural fat was extensive. The reaction was intense but not diffuse and the spinal cord itself was spared.

No viral, bacterial, fungal, or protozoal agents were isolated from the cauda equina and brainstem.

**Case 2**

**History** — An eight year old gelded crossbred pony, used for chuckwagon racing, was presented to the WCVM in the late summer of 1982 with a history of difficult urination and defecation for about three weeks. The horses had been vaccinated for Western and Eastern equine encephalomyelitis in the spring.

**Clinical Findings** — The pony dribbled urine intermittently from the sheath. The anal sphincter, tail and rectum lacked motor tone and rectum was distended with feces. Rectal examination revealed an enlarged, atonic bladder. A neurological examination was performed. Lack of pain sensation around the tail and perineum, hindlimb weakness and a mild proprioceptive deficit were observed. Cranial nerve involvement was not detected.

The hemogram was normal, however the serum protein and fibrinogen was elevated. The cerebrospinal fluid collected from the lumbosacral region was clear and colorless, but had an elevated total protein and cell count suggestive of a mild, chronic inflammation (Table I). The pony was euthanized.

**Postmortem Examination** — Gross lesions primarily involved the spinal column where there was fibrosis, reddening, and enlargement of the cauda equina caudal to S1, and considerable peridural edema caudal to T3 (Figure 2). The lumbosacral joint had excessive movement but no bone lesion was

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**Gasserian ganglion**

**Femoral nerve** - +

**TABLE II**

**Distribution of Extradural Granulomatous Inflammatory Lesions in the Central and Peripheral Nervous System of Two Horses Affected by NCE**
detected. The bladder contained a large urolith and cloudy urine but no obstruction was present. Fibrous tags were present on the parietal surface of the liver and the cecum. There was verminous arteritis of the anterior mesenteric artery.

Microscopic lesions were primarily confined to the extra-dural nervous tissue, involving mainly the cauda equina (Figure 3), however, very mild lymphocytic infiltrations were also seen in some spinal root ganglia, the Gasserian ganglia and right femoral nerve (Table II). From the midlumbar region caudally, the reaction involving lymphocytes, macrophages and plasma cells became progressively more severe. Caudal to S1, the reaction appeared more chronic, granulomatous and sclerotic with predominance of fibroblasts, fibrocytes, macrophages, giant cells and plasma cells. Lymphocytes and neutrophils associated with areas of necrosis were less prominent. Vascular changes consisting of endothelial ballooning degeneration, medical hypertrophy and adventitial necrosis in association with lymphocytes were present. The reaction in the distal cauda equina contained mainly mature fibrous tissue, whorled around blood vessels (Figure 4). There were also few granulomata, the centers of which contained caseation necrosis and neutrophils and occasional gran cells. The lower spinal cord contained secondary areas of axonal loss and Wallerian degeneration.

No bacterium, virus, fungus, or protozoa were isolated or seen in Grams, Grocott Methenamine Silver, or Ziehl Neelson stained sections. The CSF did not contain antibodies to equine herpesvirus 1.

Discussion

Both of these horses had typical signs of the syndrome known as neuritis of the cauda equina (NCE) (4,10). However, cranial nerve involvement seen in the first case and some spinal root involvement in both cases lead us to support a diagnosis of polyneuritis equi, the name proposed by Sjolte (1). The clinical and pathological findings eliminate the differential diagnoses of spinal nematodiasis (11), sorghum toxicity (12), cervical vertebral stenotic myelopathy, equine degenerative myeloencephalopathy and equine protozoal myeloencephalitis (13).

Cauda equina neuritis does not appear to have any breed or sex predilection and the syndrome normally occurs as a single case (14). In the acute form of the disease, the horse may be hyperesthetic around the perineum and sometimes the head, which may be accompanied by a head tilt and ataxia. Later, as the hyperesthesia lessens, the horse becomes hypoesthetic or anesthetic in affected areas. The insidious form, seen in both of these cases, presents as a gradual onset of paralysis of
the tail, urinary bladder and sphincter, rectum and anal sphincter resulting in urinary and fecal incontinence. Constipation, inability to swallow, weak mastication and inability to stand may also occur (14).

Elevation of CSF protein was noted in both cases, hence CSF taps should be considered as a possible aid to clinical diagnosis in suspected cases. Experimental allergic encephalomyelitis, which is pathologically similar to NCE, is characterized by the appearance of IgG in the CSF early in the syndrome (15) and may account for the elevated protein levels in these cases. The elevated serum fibrinogen seen in both cases can be associated with the extensive inflammatory reaction.

The lesions described are consistent with the nonsuppurative inflammatory infiltration of the nerve trunks of the cauda equina, involving lymphocytes, macrophages and giant cells with occasional central areas of necrosis and fibrosis, as previously described (2,3,4,6,7,8,10). Vascular lesions consisting of arterial internal elastic lamina disruption and arterial thickening have been described (6). The well developed vascularity of the inflamed nerve trunks often appear to be the central focus of the inflammatory infiltrations within the nerves (2). This would be consistent with our findings in case 2. We consider the vascular wall changes to be a result of the inflammatory reaction rather than the cause. Rooney (16), on the other hand, described fibrinoid degeneration of small arteries associated with the cauda equina, which he attributed to a possible sequel to equine viral arteritis.

Speculation as to the etiology of cauda equina neuritis has been extensive, but the disease has not been reproduced. Early reported cases were often attributed to a traumatic lesion, either excessive mobility of the sacro-lumbar region or callus formation with subsequent irritation of the cauda equina and development of NCE (17). Both of our cases were chuckwagon ponies and may have suffered trauma while racing. Gross necropsy did reveal excessive movement of the lumbosacral joint in case 2. Initial trauma of the extradural nerve root may have released myelin into the blood stream with subsequent autoimmune reaction.

Viral and bacterial infections have been proposed causes of NCE (9). Routine diagnostic microbiological procedures failed to reveal the presence of viral, bacterial or protozoal agents in these two horses. However, previous subclinical infections must be considered as possible initiating causes. One possible explanation of this syndrome in horses is an immunogenic response to a persisting herpesvirus infection (9). However, equine herpes viral antibody or virus could not be detected in case 2 and studies in case 1 failed to isolate equine herpesvirus 1 on culture. Equine herpesvirus 1 has been isolated from horses with neurological deficits varying from mild ataxia to severe paralysis, with or without cranial nerve involvement and bladder paralysis. Unlike NCE, usually more than one horse is affected, often following a previous upper respiratory tract infection or abortion. Mildly affected horses generally recover (13). Herpesvirus infections in other species produce Marek’s disease in chickens (18), herpes Zoster in man (19) and possibly Gullain-Barre syndrome (19), all of which bear some pathological similarity to NCE.

Coonhound paralysis (20), experimental allergic encephalomyelitis (21), chronic canine polyneuritis (22), Guillain-Barre syndrome (4), Marek’s disease (18) and cauda equina neuritis (4) exhibit primary demyelination. Primary demyelination is characterized by segmental myelin damage without primary changes in the axon, whereas in secondary demyelination both the axoplasm and myelin undergo degeneration (23). In primary demyelination, myelin damage either occurs in the absence of inflammatory cells, e.g. lead poisoning, or with the presence of mononuclear inflammatory cells, mainly lymphocytes and macrophages, e.g. experimental allergic neuritis (24).

Although NCE is thought to have an autoimmune pathogenesis basis (9), primary demyelination with mononuclear infiltration is not necessarily autoimmune. Wisniewski (25) proposed a “bystander mechanism” whereby any antigen that reaches the peripheral nervous system and attracts and activates lymphocytes and macrophages, may be a nonspecific cause of a primary and secondary demyelination. Infectious diseases, trauma and aberrant strongyle migration may act in this way to produce NCE.

Most authors note that NCE is extradural in nature. This may, in part, be explained by the difference in peripheral and central nervous myelin.
Analysis of human peripheral nervous myelin reveals less galactosphingolipid and more sphingomyelin than central nervous myelin and the basic protein is different in each case. The basic protein difference gives the different types of myelin different antigenic properties. It is the antigen responsible for initiating EAN (26) and we believe it is probably the antigen that initiates NCE.

Resolution of the disorder may be impossible as the horse may produce an overabundant amount of granulation tissue (27) potentiating, instead of controlling, the initial inflammatory reaction. If the initial reaction to the antigen could be depressed, the degree of neurological damage would be limited and may be reversible, providing fibrosis hasn’t occurred.

These cases demonstrate the necessity to examine thoroughly the cranial and peripheral nerves of suspect cases at necropsy. Cerebrospinal fluid examination may also aid in an antemortem diagnosis.

Acknowledgments

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References

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