ANIMAL MODEL FOR HUMAN DISEASE

Idiopathic Polyneuritis

Guillain-Barré Syndrome

Animal Model: Coonhound Paralysis, Idiopathic Polyradiculoneuritis of Coonhounds

Contributed by: J. F. Cummings, DVM, Department of Anatomy, New York State Veterinary College, Cornell University, Ithaca, NY, and D. C. Haas, MD, Department of Neurology, Upstate Medical Center, State University of New York, Syracuse, N Y.

Clinical Features

This syndrome, first described by Kingma and Catcott,[1] is primarily an occupational hazard for coonhounds (Fig 1) although it occurs in other dogs that encounter raccoons. A raccoon bite or scratch precedes the onset of signs by 7–14 days. The onset is marked by weakness and hyporeflexia in the hind limbs. Paralysis progresses rapidly, resulting in a flaccid symmetric quadriplegia. While recumbent, dogs remain alert and afebrile. Motor impairment is more pronounced than sensory changes. In severely affected animals, at the peak of their illness, one finds a complete absence of spinal reflexes, facial weakness and labored respiration. Electromyographic findings include fibrillations, positive-sharp waves and other evidence of denervation. Although 2 hounds in our care have died of respiratory failure, paralysis usually abates and recovery is to be expected (Fig 1), but the rate and extent of recovery is variable.

Pathologic Features

Pathologic changes are concentrated in the ventral roots and the spinal and peripheral nerves.[2] In keeping with clinical signs, ventral roots are affected more severly than dorsal roots (Fig 2). The lesions consist of segmental demyelination with axon preservation (Fig 3), Wallerian degeneration with disintegration of axons as well as myelin and, often, perivenular leukocytic infiltration. The constituents and prominence of the leukocytic infiltration appears to vary (Fig 4). Neurogenic muscle atrophy and retrograde chromatolysis in neurons of the ventral horn of the spinal cord occur secondary to axonal disruption.

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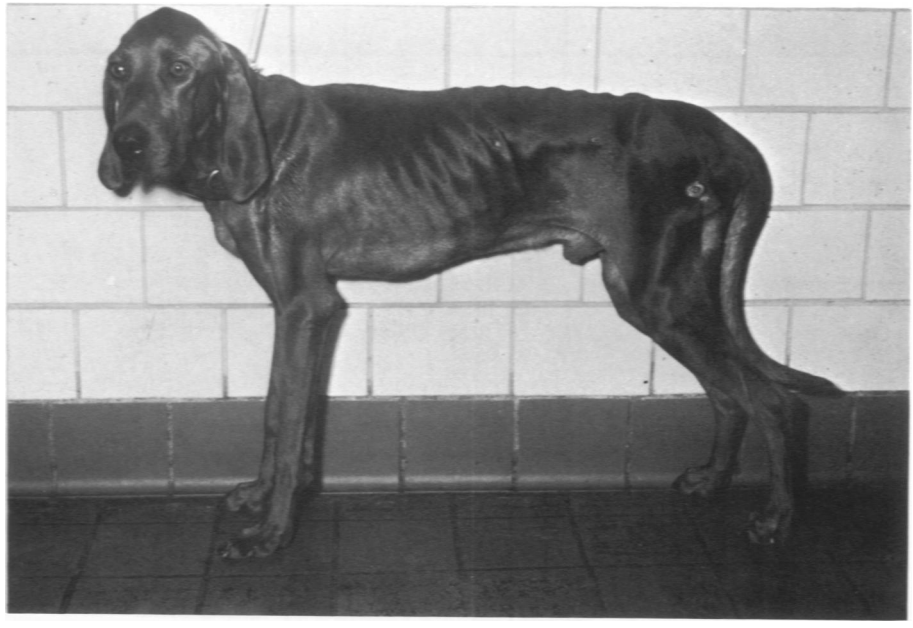

Fig 1—Red bone coonhound during recovery phase of paralysis. Note marked muscle atrophy and decubital ulcers that developed during the quadriplegic phase of illness. This hound subsequently sustained three more attacks of coonhound paralysis, each preceded by a raccoon bite.

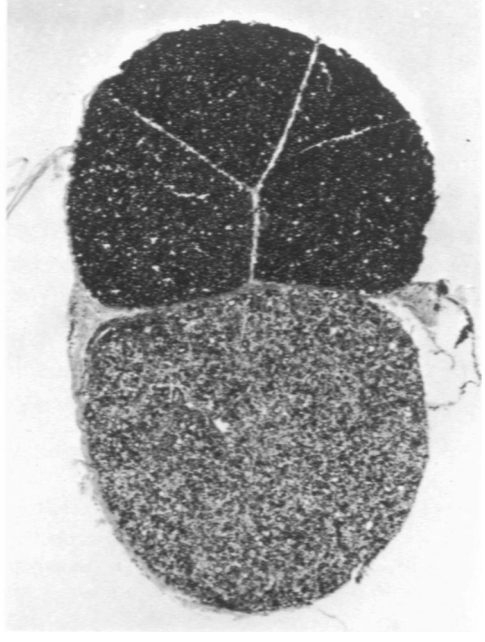

Fig 2—Transverse section of a lumbar spinal nerve. Note marked demyelination in the ventral root component. The dorsal root component above appears normal at this magnification (Luxol fast blue—cresyl echt violet, × 45).

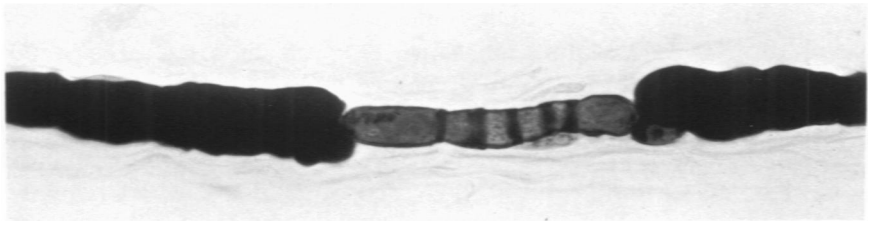

Fig 3—Single myelinated nerve fiber isolated from a lumbar ventral root of a dog that died 110 days after the onset of signs. Osmic acid stain demonstrates the restoration of myelin that occurs in areas of segmental demyelination. The short, partially remyelinated internodal length with prominent infundibuli appears thinner and paler than adjacent internodes (× 400).

Comparison with Guillain-Barré Syndrome

The clinical features of coonhound paralysis are very similar to those reported in the Guillain-Barré syndrome.[3] In man, however, the onset is preceded by an infection which often involves the upper respiratory tract. The initial symptom is usually weakness of the lower extremities which extends rapidly to the upper extremities and facial muscles.

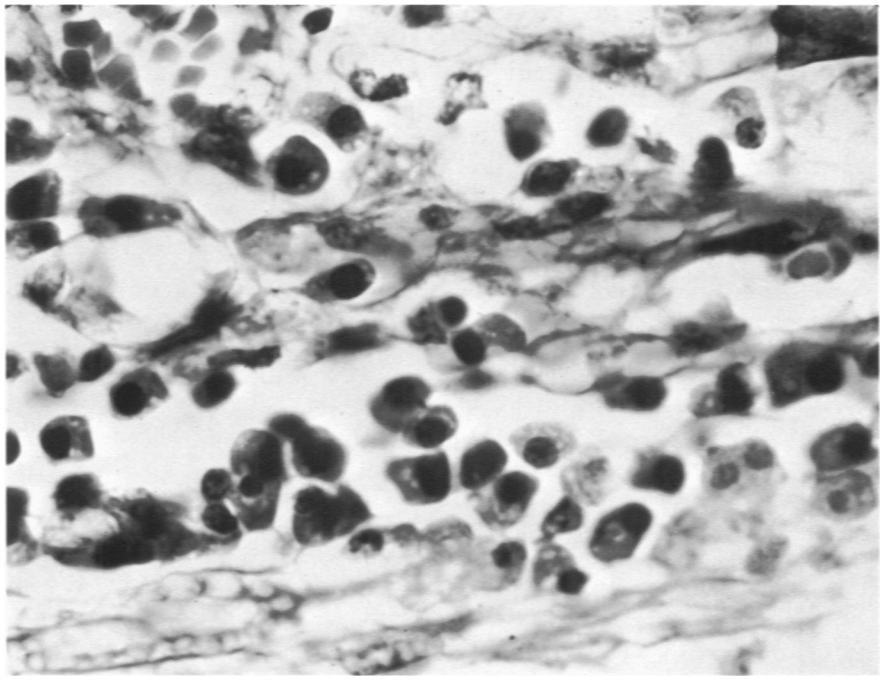

Fig 4—Section from a lumbar ventral root of a hound which died 20 days after the onset of signs. Note the perivenular infiltration of plasma cells (× 1000).

Flaccid quadriplegia is not uncommon and in about one-fourth of the cases a respirator is required because respirations are weak. Sensory changes are not usually prominent. The recovery rate varies from a few days or weeks to months.

Pathologic changes in coonhound paralysis resemble those found in the Guillain-Barré syndrome both in type and location.[4,5] Changes in roots and nerves include perivenular leukocytic infiltration; degeneration of myelin sheaths, both Wallerian and segmental types; swelling and fragmentation axis cylinders, and chromatolysis of ventral horn cells.

The cause of the Guillain-Barré syndrome is unknown. It has been postulated that autoimmune processes may be involved in the etiology.[6] To date, no infectious agent has been isolated from dogs afflicted with coonhound paralysis. Since the lesions in these dogs resemble the changes found in the Guillain-Barré syndrome and also, to some extent, those described in experimental allergic neuritis, an immune disturbance may be considered. However, the raccoon's role in initiating such a demyelinating neuritis remains enigmatic.

Availability

The clinic at Cornell rarely receives more than three or four cases of coonhound paralysis per year. Admissions usually occur during the raccoon-hunting season. The syndrome has not yet been reproduced experimentally, but if one wished to attempt this, it would seem advisable to use hounds that had previously sustained an attack of this paralysis.

References

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