CEREBELLAR CORTICAL ABIOTROPHY IN A LABRADOR RETRIEVER: CASE REPORT

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Abstract: A 12 weeks old male Labrador Retriever dog was diagnosed with cerebellar cortical abiotrophy and a possible concurrent immunodeficiency.

INTRODUCTION

Cerebellar abiotrophy, also referred to as the cerebellar cortical abiotrophy, which is a genetic neurological disease in animals best known to affect certain breeds of horses and dogs. It develops when the Purkinje cells, located in the cerebellum, begin to die off. These cells affect balance and coordination. They have a critical role to play in the brain. The Purkinje layer allows communication between the granular and molecular cortical layers in the cerebellum.

Abiotrophy means the loss of a vital nutritive factor. The exact cause of cerebellar abiotrophy is not known, but it is thought to be due to an intrinsic metabolic defect (5).

In most cases, the Purkinje neurons begin to die off shortly after the animal is born and the condition is noticeable when the animal is less than six months old, though sometimes the onset of symptoms is gradual and the animal is much older before the owner or caretaker notices a problem.

The condition in Kerry Blue Terriers is sometimes called progressive neuronal abiotrophy. Other terms used to describe the condition in dogs include cerebellar cortical atrophy and postnatal cerebellar cortical degeneration (1).

Cerebellar abiotrophy in horses was originally thought to be a form of cerebellar hypoplasia and was described as such in older research literature. However, it was discovered that in horses, the death of Purkinje cells began after the animal was born, rather than occurring in utero. Cerebellar hypoplasia is particularly common in cats and has similar symptoms.

There are other diseases that lead to cerebellar degeneration but the loss of Purkinje cells is a clear way to diagnose cerebellar abiotrophy, and the combination of symptoms is sufficiently unique in that cerebellar abiotrophy can easily be distinguished from other conditions, even in a living animal.

Cerebellar cortical abiotrophy in Labrador Retrievers is a degenerative condition and clinical signs occur at about 12 weeks of age including pelvic limb ataxia, hypermetria, truncal ataxia, and wide-based stance. Rapidly, signs progress to thoracic limb involvement, falling, and inability to walk. Positional nystagmus and reduced menace response may be
noted. Results of laboratory diagnostic testing for toxoplasmosis, canine distemper virus, Cryptococcus capsular antigen, and electroencephalography were normal (4).

The genetic status of this condition remains to be confirmed. Grossly, the cerebellum is smaller than normal. There is loss of Purkinje cells and granule cells, granule layer thinning, and folia white matter gliosis in all regions of the cerebellar cortices, especially in central and lingula lobules of the rostral vermis. The culmen and declive lobules are affected later in the disease. Retrograde degeneration of brainstem nuclei is not observed (3).

A similar disorder has been seen in a puppy of a predominantly Labrador Retriever breed (2). The clinical signs were similar to those described above; however, the progression was very slow and the dog required no assistance to walk, even at one year of age. Also, abnormal nystagmus was not seen. This dog exhibited seizure-like episodes, characterized by progressing episodes of recumbency with opisthotonus and muscle rigidity. Microscopic findings were neuronal loss and/or axonal swelling in the olivary and vestibular nuclei, along with swollen axons in the vestibular nerve in the region of the cochlear nucleus. Axonal spheroids were seen in the cerebellar nuclei. These changes were considered to represent transsynaptic degeneration as a consequence of Purkinje cell loss. Increased numbers of Bergmann’s astrocytes were seen around Purkinje cells and there were increased numbers of astrocytes in the cerebellar white matter.

CASE REPORT

Case history. A 12 weeks-old male Labrador Retriever dog (consanguine family), which manifested a recurrent digestive and respiratory infection, was referred for investigation with one-month history of disease.

Clinical manifestations. Physical examination showed anemia, rachitism, fever, but no anorexia. In addition, the patient manifested incoordination of the left pelvic leg.

Respiratory system. The physical findings were tachypnea, cough, crackles, the accentuation of bronchial breath sounds, and the presence of the secretions in the airways.

Gastrointestinal system. The puppy manifested chronic, recurrent, diarrhea, persisting for many days, which was responsive to antibiotics.

Nervous system. Incoordination, first appeared in the left pelvic leg and after few days of progression, typical cerebellar ataxia.

The patient died in 3 days after the developing of ataxia.

Anatomopathology. Grossly, the cerebellum was normal. Other macroscopic findings were anemia, rachitism and pneumonia.

The histopathologic findings included the cerebellar cortical abiotrophy – the loss of Purkinje cells (necrosis) in some areas; lymphohistiocytic pneumonia – congestion/necrosis and desquamation of the pneumocytes, interstitial lymphohistiocytic infiltrate and focal moderate neutrophilic exudate.

CONCLUSIONS

- The recurrent infection was generated by a possible immunologic defect.
- The cerebellum was grossly normal because of rapid progress of infection which stopped the abiotrophy.
- In this case, other neural symptoms (positional nystagmus, opisthotonus, muscle rigidity) could not be seen due to rapid evolution of the disease.
- Cerebellar abiotrophy cannot be prevented, other than by selective breeding to avoid the gene, and it cannot be cured.
BIBLIOGRAPHY

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