Wobbling off the Hedge

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Introduction

Wobbly Hedgehog Syndrome (WHS), also known as progressive paralysis, is a progressive degenerative neurological disease that affects primarily African hedgehogs and occasionally European hedgehogs as well.[4] The etiology of this disease is unknown but pedigree analysis indicates a familial tendency to the disease.[4] Clinical signs usually begin with mild ataxia that progresses to more severe neurological signs and ultimately leads to complete paralysis. The onset commonly occurs around two years of age but can happen at any age.[4] Wobbly Hedgehog Syndrome can only be definitely diagnosed by a post-mortem examination of the tissues from the central nervous system.[1] Characteristic histopathology of the disease includes vacuolization of the white matter of the brain and spinal cord and associated neurogenic muscle atrophy.[2] Numerous treatments have been attempted with little to no success leaving supportive care as the main focus.[4] The prognosis is grave with the majority of hedgehogs becoming completely paralyzed by fifteen months after the onset of their clinical signs and ultimately succumbing to severe muscle atrophy and starvation.[4]

History and Presentation

Baymax, a 3-year-old male African Pygmy Hedgehog, presented to Mississippi State University College of Veterinary Medicine Community Veterinary Services department on January 7, 2021 for wobbling when walking and a possible lump under his skin near his spine. Baymax began dragging his hindlimbs approximately two weeks before Thanksgiving which progressed into falling over on his right side and being unable to right himself. His environment consisted of two heat lamps that were above his cage with a temperature normally set around seventy-four degrees Fahrenheit and he was fed a diet of one tablespoon of Purina Pro Plan 7+

cat food per day. While his appetite remained good, he did not run around as much anymore nor use his wheel as often.

Upon presentation, Baymax was bright, alert, and responsive. His hydration status was within normal limits but he had an emaciated body condition. He weighed 296 grams which is nearly 200-300 grams underweight for an adult male African Pygmy Hedgehog. Chest, heart, and lungs were not evaluated. He had a respiratory rate of 64 breaths per minute. A soft mass caudal to his prepuce on his abdomen was appreciated along with a mass on his caudal back to the left of his spine. Both of his hindlimbs were extremely weak with his right hindlimb being the worst. Baymax was unable to retract his right hindlimb all the way when he curled into a ball and he appeared to not use it very much when he was walking around. The remainder of his physical exam was unremarkable.

Diagnostic Approach/Considerations

Wobbly Hedgehog Syndrome is presumptively diagnosed based off a set of typical clinical signs and history. One of the earliest indicators of WHS in hedgehogs is the inability to close their hood fully.[4] During Baymax's first visit on January 7, 2021, he demonstrated the inability to retract his right hindlimb when attempting to close his hood. Other common clinical signs include: falling consistently to one side, tremors, seizures, scoliosis, exophthalmos, muscle atrophy, self-mutilation, and difficulty regulating body temperature.[3] Throughout Baymax's visits, he displayed several of those clinical signs such as consistently falling on his right side, exophthalmos of his right eye, and significant muscle atrophy.

Diagnosis is usually based on exclusion through ruling out other differentials that have similar clinical signs since definitive diagnosis can only be confirmed post-mortem for WHS.

Other reported causes of progressive paralysis in hedgehogs are various brain tumors, intervertebral disc disease, and hepatic encephalopathy.[4] Whole body radiographs were taken on Baymax during his first visit. Findings showed severely decreased abdominal serosal detail most likely due to emaciation and decreased intra-abdominal fat with peritoneal effusion. There were two smoothly marginated, irregularly shaped, mineral opaque structures superimposed over the retroperitoneal space ventral to L3 and L4 on the lateral projections that measured 1.8 X 1.1mm. This may be due to nephrolithiasis, dystrophic mineralization, nonobstructive intestinal foreign material, or external debris on the patient. There was also an ovoid, smoothly marginated, mineral opaque structure associated with the subcutaneous tissues dorsal and to the left of the thoracolumbar spine which measured 6.5 X 4.5mm resulting in a convex deformation of the overlying quills and soft tissues. This may be due to a mineralized granuloma, neoplasia such as osteosarcoma, squamous cell carcinoma, adenoma, or osteochondroma. Based on Baymax's radiographic findings, lesser consideration was given to brain tumors and intervertebral disc disease as differentials for progressive paralysis in this case.

Additional conditions that can present like WHS initially include inner ear issues, nutritional deficiencies, skeletal problems such as broken legs or overgrown nails making it difficult to walk and semi-hibernation as a result of the temperature dropping below 70 degrees Fahrenheit.[6] These conditions were quickly ruled-out through obtaining a good history on Baymax's diet, environment, and husbandry practices along with performing a thorough physical exam.

Pathophysiology

The definitive etiology behind Wobbly Hedgehog Syndrome at this time remains unknown although there are several theories of infectious, nutritional, toxic, autoimmune, or

genetic causes.[4] There was one report in an European hedgehog with WHS that had mild inflammation which suggests an infectious etiology. Further virus isolation and testing indicated that the lesions more closely resembled canine distemper than WHS. Because of the lack of evidence of infectious, nutritional, and toxic etiologies, a genetic basis is the most plausible.[2]

In 1991, the United States banned the importation of African Pygmy Hedgehogs leading to subsequent inbreeding that in turn led to a lack of genetic variability. Research also shows that WHS has not been reported in wild African Pygmy Hedgehogs.[2] Although, the prevalence of WHS occurs in approximately 10% of pet African Hedgehogs in North America.[4] Additional studies have demonstrated that there is a lack of transmission of WHS between unrelated hedgehogs and there is a tendency of WHS to occur in family lines strongly suggesting an inherited component to the disease.[2] Breeding of hedgehogs with presumed WHS or those closely related to hedgehogs diagnosed with WHS is therefore not recommended.

Initially, the hedgehogs present with mild ataxia and balance issues and the clinical signs are usually relapsing and remitting.[5] Over several months, the signs become progressively more severe and include consistently falling to one side, tremors, muscle atrophy, and complications with regulating body temperature. As this disease further progresses, the hedgehogs experience severe weight loss. However, in most cases, there is no apparent loss of appetite until the terminal stages of the disease when most hedgehogs become dysphagic. The end result is tetraplegia with muscle atrophy.[4]

An in-depth study was performed on twelve African pygmy hedgehogs that were submitted for necropsy to the Department of Veterinary Pathobiology at Texas A&M University between 2010 and 2016 to study the pathophysiology of WHS. The study described WHS as a degenerative spongiotic disease of the myelin with widespread CNS involvement that results in

damage to the protective coating of the nerve cells.[2] There appears to be a loss of myelin first, then secondary degeneration, and loss of the axon followed by neuronal degeneration.[2]

The primary histologic lesion is vacuolization of the white matter tracts of the cerebrum, cerebellum, and brainstem.[8] The lesion also occurs in the white matter tracts throughout the spinal cord. The primary lesion in skeletal muscle is neurogenic atrophy of myocytes without inflammation. These myocytes are eosinophilic and triangular.[8]

WHS is often incorrectly compared to multiple sclerosis in humans.[4] However, autoimmune inflammation is not observed in the central nervous system tissues of the hedgehogs with WHS. WHS shares many features with most spongy myelinopathies in other species in having a predilection for the white matter and splitting of the myelin sheath at the intraperiod line.[2]

Treatment & Management Options

Numerous treatments for Wobbly Hedgehog Syndrome have been attempted with little to no success. Although, with hand-feeding and additional supportive care measures, hedgehogs with this disease can survive for many more months.[4]

Supportive care focuses on catering to the specific challenges the animal is experiencing and usually varies case by case. Some common examples include: using towels to help keep them upright, increasing accessibility to food and water dishes, and cleaning them frequently when they become soiled.[1] Keeping them warm is also highly important as hedgehogs with WHS can have issues maintaining warmth due to decreased mobility and the inability to seek out warm spots when heating pads are used.[1]

The most commonly reported therapies for hedgehogs with WHS include supplementation with Vitamin E, Selenium, Vitamin B, and calcionate syrup. Oral prednisone has often been prescribed as well. Antibiotics including trimethoprim sulfa, amoxi-drop liquid, and baytril sometimes have been prescribed for the treatment of concurrent infections.[4]

After Baymax's first visit on 1/7/21, he was prescribed Meloxicam which was administered at 0.02ml orally every 24 hours for seven days. This non-steroidal anti-inflammatory drug is commonly used to treat acute or chronic pain and inflammation. He was also given Hill's a/d Urgent Care canned food samples to offer ½ a teaspoon into his diet every day with the rest of his food which provides highly digestible protein and fat. This diet also has high levels of antioxidants to support the immune system and offset oxidative stress. Additionally, a recommendation was made to increase the temperature on his heat lamps from 74 degrees Fahrenheit to the higher end of the range around 80 degrees or to place heating pads under his cage to add warmth.

During his next visit seven days later, he was restarted on Meloxicam which had previously been discontinued after he was experiencing diarrhea. Baymax was also re-weighed and had a 20 gram increase since his last visit. However, he was unable to right himself after falling to his right side so a plan was made to build him a prototype cart for better mobility so he could access his food and water bowls.

On 1/28/21, Baymax came in for a weight re-check which was down 30 grams from his last visit. His overall appearance was weak with very minimal right hindlimb usage. Subcutaneous fluids of Lactated Ringer's Solution was administered along with Baytril.

Expected Outcomes & Prognosis

Baymax was humanely euthanized on 1/29/21 due to his declining health. He was no longer able to eat on his own nor could he get around even with his cart. He was submitted for necropsy where a definitive diagnosis of Wobbly Hedgehog Syndrome and osteosarcoma was made based on histopathologic examination of his CNS tissues.

The brain and spinal cord was described as having myelin degeneration and spongiosis that was bilaterally symmetrical, chronic with gliosis, and contained perivascular edema. The mass on the ribs was diagnosed as osteosarcoma that was focal, chronic, and had central necrosis.

The prognosis for Wobbly Hedgehog Syndrome is grave as this is a progressive, incurable neurodegenerative disease. Most hedgehogs are humanely euthanized or die in less than 2 years of diagnosis due to starvation.[7] In the advanced stages of the disease, the paralysis progresses from the rear of its body to the front ultimately paralyzing its tongue leaving it unable to swallow food.[3]

There have been reports of Vitamin E supplementation temporarily improving signs and slowing the progression of the disease; however, no treatment has been shown to prevent the progression of paralysis.[7]

Conclusion

Wobbly Hedgehog Syndrome is a condition characterized as progressive paralysis or degenerative myelopathy that has been observed in pet African hedgehogs and is predicted to have a genetic predisposition.[4] A definitive diagnosis can only be made post-mortem via histological examination of the spinal cord and brain tissues.[8] However, a presumptive diagnosis is usually made based on history and clinical signs that include an initial presentation of mild

ataxia, wobbling, and muscle atrophy that turns into tetraplegia during the terminal stage. This is an incurable disease and supportive care is the mainstay of therapy which focuses on providing a high quality diet, adequate warmth, and access to food and water bowls.[6] It is important to educate owners about the progression of this disease and that humane euthanasia will ultimately need to be performed when the pet's quality of life declines.[9]

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