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Once seen never forgotten - Unusual neurological syndromes

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Introduction
Clinical neurology is a very structured discipline. Neuroanatomy dictates that lesions in particular locations will produce a very specific set of neurological signs. Thus, following rules of localization enables everyone to potentially locate problems within the central nervous system (NCS) and from there, to establish a list of differential diagnoses and diagnostic and therapeutic plans. However, there are neurological disorders that are either extremely difficult to localize, or have been poorly described in the literature, or have a classic constellation of signs that are pathognomonic for the condition. The aim of this presentation is to review some of these conditions with the hope that these will be easily recognizable to the participants in the future.

Scotty Cramp and other Hypertonicity Syndromes
There are several disorders recognized in specific breeds of dog, most notably the Scottish and Cairn terrier (Scottie Cramp) and the Cavalier King Charles Spaniel (Episodic Falling)3. Scottie cramp is a hereditary disorder believed to be transmitted by an autosomal recessive mode. Signs manifest within the first few months of age and are triggered by excitement or exercise. With exercise, the dog’s hind limbs develop a stilted gait and become progressively more spastic, causing the lumbar spine to arch and the pelvic limbs to appear to ‘catch up’ to the thoracic limbs. The dog tends to develop a praying posture with severe spasticity of all musculature. The pupils are often miotic, although the dog does still appear to be conscious. The episodes last a few minutes and the dog is normal between episodes. The signs are usually responsive to a benzodiazapine. The pathogenesis has been linked to serotonin levels because drugs such as amphetamine (that antagonize serotonin) worsen signs, and drugs that increase serotonin levels (e.g. tryptophan) can improve signs. Episodic falling, or hypertonicity of Cavalier King Charles Spaniels is somewhat similar in nature. Signs usually develop in the first 6 months of life and are triggered by excitement, stress and anxiety. Signs manifest in the pelvic limbs first and again they appear to develop severe spasticity. Benzodiazepines, in particular clonazepam, will control the clinical signs.

Exercise Induced Collapse in Labrador Retrievers
This is another autosomal recessive disease that has been enigmatic to the specialty until recent genetic studies revealed the underlying cause as a mutation in the dynamin 1 gene1. Signs typically develop in the first two years of life and manifest as weakness and collapse after heavy exercise. The dogs develop a weak and ataxic gait in the pelvic limbs first and this rapidly progresses to recumbency. Many dogs appear to have no control over their torso or neck and will have a marked truncal sway and head bob if they attempt to walk. They are usually panting very heavily and signs seem worse in warmer weather. The disease can be fatal but usually the dog will recover completely within 15 – 30 minutes. Dynamin 1 is a GTPase that regulates endocytic vesicle formation. It is proposed that this mutation affects the efficiency of this process, leading to collapse under conditions of high intensity exercise. Similar conditions have been seen in Border Collies and Australian Shepherds. However, they do not appear to have the same mutation.

Dancing Doberman Disease
This aptly named disease affects adult Doberman Pinschers causing a classic shifting of weight from hind limb to hind limb. Signs develop between 6 months and as late as 7 years of age. The dogs typically start to hold one hind limb in flexion when standing. The signs progress to involve both hind limbs, causing the shifting of weight that leads to the appearance of dancing. Other signs noted include a reluctance to sit, causing the animal to raise and lower its hind end several times and ultimately rest gingerly on one hip, and atrophy of the gastrocnemius muscles. Reflexes are intact. Although the animals do not appear to be painful on palpation, observation of their movements and behavior is suggestive of abnormal and unpleasant sensations. Histological evaluation of nerve and muscle has produced a variety of observations, but changes in the nerves, (sensorimotor) are present. There is no known treatment, and signs tend to progress insidiously but do not usually result in paralysis or overt pain. The affected dogs therefore live out their life as a functional pet.

Generalized Tremor Syndrome – or Little White Shakers
This is a dramatic condition that classically affects small white breeds of dog such as the Maltese and West Highland White terrier.4 However, it also affects other breeds such as Dachshunds, Boston Terriers, Lhasa Apso’s and so is not restricted to dogs of a specific coat color. Signs usually come on rapidly in young adult dogs and consist of severe generalized tremors that can be present at rest in severe cases.
es but are definitely exacerbated by intent. The gait is often hypermetric, and opsoclonus and vertical nystagmus may be present. Many dogs also lose their menace response although they are visual (ie a sign of cerebellar involvement). Diagnostic workup typically shows mild inflammatory changes in the cerebrospinal fluid. Infectious testing is negative and dogs respond to a combination of immunosuppression and benzodiazepine. In severe cases, additional drugs such as Phenobarbital and carbamazepine may be needed to eliminate the tremors. This disease is presumptively believed to be an autoimmune disease of the central nervous system, and primarily of the cerebellum; in general the prognosis is good.

**Idiopathic Head Tremor Syndrome**

This is a syndrome that affects the English bulldog most commonly, but is also seen in other breeds. In my experience, beagles are also over-represented. Signs present in young dogs – they typically develop a vertical nodding of the head, although we have seen some that shake their head from side to side.

When the episodes first start, they tend to be short lived, usually come on when the animal is at rest and the animal appears otherwise normal. They are completely unresponsive to anti-epileptic drugs. Over the next few weeks, the episodes often become more frequent and often last for longer. The most effective way to stop the episode is to get the patient to fix their attention on something – a food treat is ideal. There is no specific treatment and with age, the problem appears to become less severe. The etiology of this movement is unknown.

**Idiopathic Vertigo**

There is almost nothing written about episodic vertigo in dogs. We are all familiar with idiopathic or geriatric canine vestibular syndrome, a condition that acutely produces severe vestibular signs that improve with time. Episodic vertigo is distinct from affected dogs show signs for a few minutes to hours and are completely normal in between. This can occur in dogs of any age, and owners report that they usually become extremely anxious, pant heavily, they may salivate or vomit, they develop nystagmus and are ataxic if they attempt to walk. They may have a head tilt, but this is not always present.

The signs resolve within 10-15 minutes or may persist for a few hours. These patients merit a full diagnostic workup and in some cases an etiology is found. For example, I have seen this associated with severe hypotension (signs resolved with treatment of hypotension), and in a dog that had a history of severe otitis externa/medial that had been treated surgically. In this case, the dog developed a head tilt to the side that had received surgery. In most cases, the diagnostic workup is unrewarding. Some dogs respond to treatment with diazepam and meclizine, others do not. The cause is unknown, but in human medicine peripheral vestibular disorders that can cause paroxysmal vertigo (e.g. Menieres disease and Benign Paroxysmal Positional Vertigo) are well described. Positional Vertigo is believed to be due to debris in the semicircular canals and can be treated by performing a series of head movements.

**Bilateral Peripheral Vestibular Disease**

This is simply a manifestation of idiopathic vestibular disease that can occur in cats. The only reason to mention it here is that the classic sign of vestibular disease, a head tilt, is not present and therefore the syndrome is frequently not recognized. Affected cats adopt a posture low to the ground and have extreme ataxia when they attempt to move. Their head executes wide swings from side to side, and this is the classic sign that can help with identification of the syndrome. Eye movements are abnormal and physiologic nystagmus cannot usually be elicited, but spontaneous nystagmus is not necessarily present.

**Muscle Contractures – Semimembranosus/Semitendinosus and Infraspinatus**

While not neurological syndromes, these disorders frequently cause bizarre gaits that can be interpreted as neurologic in nature. The muscles involved become replaced with fibrous tissue, preventing the limb from describing a normal range of motion. The most common disorder is contracture of the semi-membranosus, semitendinosus, or gracilis muscles in German Shepherd dogs. Signs usually develop in the first 5 years of life; as the hind limb is protracted, the arc described by the paw is abruptly curtailed and the paw strikes back and down to the ground. Presence of the contracture can be confirmed by palpation of the tight fibrous band on the medial aspect of the thigh. The condition is usually bilateral but asymmetrical. Surgical intervention, even removal of the entire affected muscle, is frequently unsuccessful in the long term, as more muscles become involved. However, resection combined with rehabilitation ahs been reported to be successful by some groups. Overall, this is simply a mechanical, non-painful gait disorder. Contracture of the infraspinatus muscle causes the thoracic limb to circumduct while walking and when the elbow is flexed.

**Cushings Myopathy**

A very low percentage of dogs will develop an unusual myopathy that has been described a pseudomyotonia secondary to uncontrolled Cushings disease. These dogs develop an unusual gait characterized by stiffness, (a failure to flex the joints) and usually, associated with clear signs of Cushings Disease such as calcinosis cutis, or poor skin elasticity. The disorder can be confirmed by confirmation of Cushings Disease, EMG (which demonstrates complex repetitive discharges) and muscle biopsy. Appropriate treatment of Cushings Disease will control the signs of myopathy somewhat, although a complete remission may not occur.

**References**