A dropped face, a dropped jaw and a wonky eye: neurological disorders affecting the head and face

Steven De Decker
DVM, PhD, DipECVN

Royal Veterinary College
Hatfield, United Kingdom
SDeDecker@rvc.ac.uk

A large number of disorders can cause abnormalities of the head and face. The musculature of the head and face is innervated by 12 pair of cranial nerves (CNs). The nuclei of these CNs are located in the brainstem. The most important step when evaluating an animal with facial abnormalities is to determine which CNs are affected and if the problem is most likely located in the peripheral (eg, the cranial nerve itself) or the central nervous system (eg, the brainstem). Clinical signs can vary from pure cosmetic abnormalities, such as drooping of the face, to potentially life-threatening abnormalities, such as megaesophagus.

HEAD TILT, NYSTAGMUS, AND STRABISMUS

These clinical signs suggest dysfunction of the vestibulocochlear nerve (CN VIII), oculomotor nerve (CN III), trochlear nerve (CN IV), and abducent nerve (CN VI). These are most often seen in vestibular disease, which is discussed in *"Clinical reasoning in vestibular disease"*

ATROPHY OF THE MASTICATORY MUSCLES

Atrophy of the muscles of mastication can be unilateral or bilateral and can be neurological or non-neurological in nature. Although around 25% animals will not demonstrate any abnormalities on further diagnostics, a common cause of unilateral atrophy is a peripheral nerve sheath tumor of the trigeminal nerve (CN V) or other tumours affecting the brainstem. Unilateral atrophy of the masticatory muscles can therefore be caused by a variety of benign and malignant conditions. A proportion of affected animals will also demonstrate decreased sensation of the face.
.

Non-neurological causes of bilateral atrophy include cachexia, and primary and secondary hyperadrenocorticism. A common cause of bilateral masticatory atrophy is masticatory myositis. This is an immune mediated condition targeting type 2M myofibers, which are found exclusively in the muscles of mastication. The muscles of mastication include the masseter, temporalis, and pterygoid muscles. Large breed dogs are most often affected and the German Shepherd, Hungarian Vizsla, and Cavalier King Charles Spaniel seem to be predisposed.[1,2] Dogs can demonstrate painful swelling of the muscles of mastication, exopthalmos, and unwillingness to opening the mouth in the acute stage of the disease. This stage is not always recognized by the owner and can evolve rapidly to muscle atrophy, enopthalmos and an inability to open the mouth due to muscle atrophy, destruction and contracture. A definitive diagnosis is made by detection of antibodies against 2M muscle fibers. Treatment consists of long-term immunosuppressive drug administration (prednisolone or azathioprine). Most dogs treated aggressively in the early stages of disease respond favorably. Although muscle atrophy is often permanent, the primary goal of treatment is to maintain or regain the ability to open the mouth. Physiotherapy of the jaw muscles by encouraging chewing is advised. It should never be attempted to open the jaw by applying excessive force.

DROPPED JAW

Dropped jaw can be caused by non-neurological and neurological disorders. Non-neurological causes of dropped jaw include jaw fractures and bilateral luxation of the temporomandibular joint. Neurological causes of a dropped jaw include idiopathic trigeminal neuritis, lymphoma (especially in cats), generalized polyneuropathy (eg, Botulism), Neospora, idiopathic hypertrophic pachymeningitis, and rabies (in endemic regions). The most common cause of dropped jaw in dogs is idiopathic trigeminal neuritis. Golden retrievers are overrepresented[3] and most animals do not demonstrate other neurological deficits. A proportion of animals will also demonstrate a degree of decreased facial sensation and/or Horner's syndrome. Treatment is supportive by assisting the animal to eat. This can be done by offering soft food and keeping the mouth partially closed with a muzzle. A minority of cases will need a feeding tube placed. This disorder has a good prognosis and most animals will demonstrate a complete recovery in 3 weeks time. Physiotherapy can be offered by encouraging the animal to play with a tennis ball.[3]

ABNORMAL FACIAL EXPRESSION

Abnormal facial expression is typically caused by facial nerve (CN VII) impairment. This cranial nerve is largely responsible for motor function of the face and dysfunction will result in a 'drooped face'. The most common cause of facial nerve paralysis is idiopathic facial nerve paralysis (75% of dogs and 25% of cats with facial nerve paralysis). Other causes of facial nerve paralysis include otitis media/interna, neoplasia of the middle ear, myasthenia gravis, trauma, iatrogenic, brainstem disease, and generalized lower motor neuron disease. Idiopathic facial nerve paralysis occurs acute, can be unilateral or bilateral, and is typically not associated with any other neurological deficits. Because affected animals are unable to blink their eye, exposure keratitis can occur.

Treatment therefore consists of proper eye lubrication to avoid this complication. Although this is a rather benign condition, prognosis for recovery is guarded. Not all animals will recover normal facial expression and recovery is often incomplete. Recovery can take 3-6 week if it occurs.[4] Chronically affected cases can progress to opposite clinical signs. These animals can demonstrate contracture of the facial muscles, with a decreased palpebral fissure, elevated position of the ear, and retraction of the lip. This facial expression should be differentiated from tetanus and 'hemifacial spasms'. The latter is caused by irritation of the facial nerve.

REGURGITATION

Regurgitation is often associated with megaesophagus. This can occur as an isolated clinical sign or can be part of generalized lower motor neuron disease. The occurrence and clinical importance of megaesophagus in animals with generalized lower motor neuron disease is discussed in *"Diagnosis and management of neuromuscular disease"*. Other important causes are congenital megaesophagus, persistent right aortic arch, esophagitis, and brainstem disease. Treatment consists of management of the underlying disease if possible, feeding the animal in a vertical position, and feeding a diet of a gelatin-like consistency. Every animal with megaesophagus should be closely monitored for clinical signs of pneumonia.[5] Prognosis is guarded because of the high incidence of aspiration pneumonia. Even after correct treatment of the underlying disease, megaesophagus is not always reversible.

DYSPHONIA, DYSPAGIA, AND STRIDOR

These clinical signs are often associated with laryngeal dysfunction. The larynx is innervated by the recurrent laryngeal and cranial laryngeal nerve, which both originate from the vagus nerve (CN X). Inherited and acquired causes are recognized. In some breeds with inherited disease, laryngeal paralysis is part of generalized lower motor neuron disease.[6] Acquired disease occurs typically in middle aged and older large breed dogs, such as the Labrador Retriever. Although this disorder was previously considered to be idiopathic, it has more recently been considered to be part of a more generalized neuropathy.[7] Other causes are iatrogenic after cervical surgery, lead and organophosphate toxicity, infection and neoplasia in the cervical region. Diagnosis is usually based on visual inspection of movement of the vocal cords and treatment consists typically of surgical lateralization of the arytenoid cartilage. The consequences of concurrent progressive neuromuscular disease should be considered before surgery is performed.

TONGUE ABNORMALITIES

Bilateral weakness of the tongue is manifested by the inability to retain the tongue in the mouth. Unilateral abnormalities cause marked asymmetrical movement of the tongue. In chronically affected cases, unilateral atrophy and contracture of the tongue will cause deviation to the affected side. This is an uncommon presentation and is most often associated brainstem disease, trauma, or generalized neuromuscular disease (eg, Botulism).

MULTIPLE CRANIAL NERVE ABNORMALITIES

Various combinations of unilateral or bilateral CN deficits can occur. It is important to evaluate if cranial nerve deficits are unilateral or bilateral. The most common cause of *bilateral* multiple cranial nerve abnormalities is generalized lower motor neuron disease, such as idiopathic polyradiculoneuritis (also referred to as Coonhound paralysis), botulism, and feline dysautonomia. These animals will often demonstrate concurrent signs of generalized weakness. The most common cause of *unilateral* multiple cranial nerve abnormalities is brainstem disease. These animals will often demonstrate other brainstem signs, such as decreased mentation, tetraparesis and ataxia, or central vestibular disease. Bilateral multiple cranial nerve deficits are typically not associated with brainstem disease because a lesion within the brainstem large enough to cause bilateral deficits would most likely be fatal.

REFERENCES

1. Haley AC, Platt SR, Kent M, et al. Breed-specific polymyositis in Hungarian Vizsla dogs. J Vet Intern Med 2011;25:393-397.
2. Pitcher GD and Hahn CN. Atypical masticatory muscle myositis in three cavalier King Charles spaniel littermates. J Small Anim Pract 2007;48:226-228.
3. Mayhew PD, Bush WW, Glass EN. Trigeminal neuropathy in dogs: a retrospective study of 29 cases (1991-2000). J Am Vet Med Assoc 2002;38:262-270.
4. Varejão AS, Muñoz A, Lorenzo V. Magnetic resonance imaging of the intratemporal facial nerve in idiopathic facial paralysis in the dog. Vet Radiol Ultrasound 2006;47:328-333.
5. Mace S, Shelton GD, Eddlestone. Megaesophagus. Compend Contin Educ Vet. 2012 32:E1
6. Granger N. Canine inherited motor and sensory neuropathies: an updated classification in 22 breeds and a comparison to Charcot-Marie-Tooth disease. Vet J 2011;188:274-285.
7. Jeffery ND, Talbot CE, Smith PM, Bacon NJ. Acquired idiopathic laryngeal paralysis as a prominent feature of generalised neuromuscular disease in 39 dogs. Vet Rec 2006;158:17.