Unilateral Facial Paresis Secondary to a Suspected Brainstem Arachnoid Cyst in a Maltese Dog

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ABSTRACT. An 8-year-old, intact female Maltese dog was presented with decreased tear production and unilateral loss of eye blinking. Neuro-ophthalmic examination and brain magnetic resonance imaging were performed to determine the origin of facial paresis. A cystic lesion in the left pontomedullary region which displayed equal intensity to cerebrospinal fluid was revealed. Hyposignality was noted on fluid attenuated inversion recovery sequences, and the lesion was suggestive of an arachnoid cyst. This report described unilateral lesion in the left pontomedullary region which displayed equal intensity to cerebrospinal fluid was revealed. Hyposignality was noted on fluid attenuated inversion recovery sequences, and the lesion was suggestive of an arachnoid cyst. This report described unilateral facial nerve dysfunction that resulted from a suspected brainstem arachnoid cyst in an unusual anatomic location.

KEY WORDS: arachnoid cysts, brainstem, canine, facial paresis.

Facial paresis and paralysis result in facial muscle dysfunction from cranial nerve diseases and injuries [26]. Intracranial and brainstem abnormalities including congenital and neoplastic space-occupying lesions can result in facial nerve dysfunction, which are classified as central in origin. Additionally, parasympathetic denervation to the lacrimal pathway of the facial nerve can also result in loss of tear production. Intracranial cysts are likely developmental in origin and can develop in cerebrum, cerebellum, and brainstem thus may cause cranial nerve dysfunction in human and dogs as well [5, 21, 29, 31]. However, intracranial cysts located in the brainstem are extremely rare in both human and dogs [8, 18, 21, 23, 28, 31, 32]. We described a case of unilateral facial paresis concurrent with lacrimal loss, which was strongly suspected to be caused by a pontomedullary cystic lesion.

An 8-year-old, intact female Maltese dog was admitted for evaluation of a unilateral blinking disorder. The patient had been diagnosed as keratoconjunctivitis sicca (KCS) by the referring animal hospital. Topical therapy for KCS including artificial tear, topical antibiotics and cyclosporine showed no improvement. Moreover, sudden onset of blepharoedema of the left eye (OS) was detected a few days before visiting our referral hospital.

The outer appearance of the face was symmetrical. However, the left nostril was drier and more narrowed than the right nostril (Fig. 1A). Marked blepharoedema and mild mucoid ocular discharge were observed bilaterally (Fig. 1B). Lacrimal function with the Schirmer tear test (STT (Tear Flo; Contacare Ophthalmics & Diagnostics, Gujarat, India)) demonstrated severely decreased tear production in the left eye (STT<5 mm/min, normal range, 15–25 mm/min) and mildly decreased production in the right eye (OD [STT 10 mm/min]). A neuro-ophthalmic examination revealed a loss of palpebral and corneal reflexes (OS) and the menace response (OS). Direct and consensual pupillary light responses (PLR) and the results of other bilateral cranial nerve tests were normal. The patient was tentatively diagnosed as unilateral facial paresis. As the course of clinical signs was slow and progressive, the presence of primary cause of facial paresis was strongly suspected rather than idiopathic facial paresis which typically showed acute onset. Thus laboratory tests were performed to search the cause of KSC and facial paresis. Otoscopic examination and skull radiography showed no evidence of otitis. The results of routine laboratory test were not remarkable. Reverse transcriptase-polymerase chain reaction (RT-PCR) to detect canine distemper virus antigen in both serum and ocular secretion showed negative results. The serum basal total thyroxine (T4) concentration was within normal range (basal total T4 2.9 μg/dl, reference range, 2.1–4 μg/dl). Based on the results, inflammation and infection of peripheral nerve and hypothyroid neuropathy were excluded. Therefore, central facial nerve dysfunction inducing neurogenic KCS and facial paresis was highly probable. Neurologic examination was performed due to the possible presence of intracranial or brainstem lesions that affect facial nerve. But postural reactions were all normal and no other cranial nerve abnormality was shown except facial nerve palsy. A magnetic resonance imaging (MRI) study of the brain was conducted with the 0.2 Tesla MRI scanner (E-scan; ESAOTE, Genova, Italy). A large, round, and cyst-like structure with clear demarcation was noted in the pontomedullary region. On the sagittal images, the cystic lesion...
mildly compressed fourth ventricles and cerebellum and a syrinx formation was detected in the cervical spinal cord segment. A small cyst was also identified in the quadrigeminal cistern. These cysts displayed hypointense signals on T1-weighted pre-contrast images (Fig. 2A and 2D). They were not enhanced after intravenous administration of gadolinium-diethylenetriaminepentacetic acid (Omniscan; Nycomed, Inc., Princeton, NJ, U.S.A.) (0.1 mmol/kg) (Fig. 2C and 2F). The cystic masses displayed strong hyperintense signals similar to those of cerebrospinal fluid (CSF) on T2-weighted images (Fig. 2B and 2E). Fluid attenuated inversion recovery (FLAIR) sequences demonstrated hypointense signals of the cystic lesions (Fig. 2G). Intracranial arachnoid cysts (IACs), cystic neoplasia, and brain abscesses were the primary rule-outs based on MRI findings. Results of CSF analysis were normal. CSF was collected by cerebellomedullary cisternal puncture method with 23 gauge spinal needle. Color and turbidity of CSF showed no remarkable findings. Total CSF protein determination was measured with a urinary reagent dipstick according to a previous method [9] and revealed normal findings in the present case (result; trace/ reference range; below 30 mg/dl). The number of total nucleated cells was 1 cells/μl (reference range; 0–5 cells/μl. Small numbers of monocyctoid cells and activated macrophages were detected on the cytological evaluation of CSF and those findings were normal according to the previous reports [1, 16]. To rule out viral infection, canine distemper virus antigen RT-PCR was tested, and result was negative for the CSF. In addition, bacterial and fungal cultures were performed on the CSF, and the results were all negative. Therefore, CSF analysis revealed no evidence of infectious, inflammatory, or neoplastic changes. In addition, the patient did not show fever or depression. The physiological conditions were all normal and the results of hemogram and serum chemistry were uneventful. The patient was tentatively diagnosed with IACs in the pontomedulla and quadrigeminal cistern.

Syringomyelia designates a fluid filled cavity formation within the spinal cord and is associated with congenital abnormalities or may be complicated with variable disorders including trauma, inflammation, tumor, and hemorrhage [13]. Congenital abnormality of the brain is the more common cause of syringomyelia and especially in small breed dogs, caudal occipital malformation syndrome (COMS) is believed to lead to abnormal CSF flow and to induce cervical syrinx formation [5, 7, 13]. In this present case, the cervical syringomyelia may be formed by COMS in considering the breed of our patient, but the possibility of secondary syringomyelia is still existent. Space occupying lesion in the caudal fossa may induce cerebellar herniation thus can be the primary cause of acquired syringomyelia. There are two reports of acquired cervical syringomyelia induced by cerebellar herniation due to brainstem tumor [3, 13]. According to one previous report [3], the size of the mass was reduced and cerebellar herniation and syringomyelia were resolved after corticosteroids and radiation therapy of the mass. This indicated that cervical syringomyelia could be induced by space-occupying lesion in brainstem resulting to cerebellar herniation. In the presenting case, the
cystic lesion in the brainstem was large enough to compress the cerebellum thus cervical syringomyelia was supposed to be induced by the space occupying large brainstem cystic lesion. Moreover, this case had no history of trauma to the cervical portion of the vertebral column, and of any other neurological signs until presented to our hospital. This shows the probability that the brainstem cystic lesion may be associated with the cervical syringomyelia.

Medical treatment with oral steroid and diuretics was initiated. Supportive topical therapy for KCS included topical antibiotics, NSIADs, and artificial tears. Blepharitis disappeared at the 1-month recheck, however, facial paresis was unchanged and a corneal erosion (OD) had developed. Systemic steroid therapy was tapered and eventually discontinued. Intensive topical therapy for KCS and the corneal erosion including artificial tear and antibiotics was maintained. On the 3-month recheck, although the inflammation of the ocular surface was lessened due to intensive topical eye drop treatment and the results of STT showed slight increase (STT 6 mm/min), the tear production was still abnormal. The unilateral dry nose and blepharoptosis persisted, too. Facial nerve dysfunction resulted from brainstem lesion was provisionally concluded as the cause of KCS.

Idiopathic facial nerve paralysis in the dog accounts for approximately 75% of all acute facial paralysis cases, and is the most common cause of peripheral facial nerve paralysis in dogs in the absence of otitis media [14, 31]. There are few reports describing central facial paresis and paralysis in the absence of concurrent neurologic signs. Lesions of the facial nerve parasympathetic nucleus or within the facial nerve proximal to the genu facial canal may result in dry eye along with facial nerve motor neuron signs [15]. Facial nerve dysfunction can occur secondarily to intracranial cysts in the pons and medulla that result in facial nerve impingement. A pontomedullary cystic lesion was detected in the patient in this case report and CSF analysis demonstrated that the lesion was non-inflammatory and non-neoplastic. Intracranial and brainstem cysts which were not associated infection, inflammation, and neoplasia may be epidermoid, dermoid, or arachnoid cyst. Generally, these cysts display different MRI characteristics. Dermoid cysts are characterized by mixed signals on both T1- and T2-weighted images because of the presence of fat [29] thus can be distinguished. IACs and epidermoid cysts have similar intensities of low intensity signals on T1-weighted and high intensity signals on T2-weighted images equal to CSF intensities [21, 23, 25]. But FLAIR may assist in the differentiation of the type of the cyst due to the different signal according to the contents. IACs which contain CSF have significantly suppressed signals, whereas epidermoid cysts are filled with protein and keratin, and display higher signals than CSF on FLAIR images based on the results previously described [8, 9, 20, 25, 27].

IAC in both dogs and human is often an incidental finding and in those cases, specific treatment is not considered [7]. Moreover, the present patient had unilateral facial paresis without other neurologic abnormalities. However, the size of the cystic lesion was too large to ignore as an incidental finding. The location of the cystic lesion and the symptom of facial nerve paresis had the consistent direction. Isolated facial nerve palsy associated with posterior fossa arachnoid cysts was reported in human case [22] and they suggested the direct relationship between the facial nerve palsy and the cyst, but the specific mechanism was not identified. In patients with brainstem lesions, common presenting clinical signs of dysfunction are cranial nerve deficits, vestibular sign, gait disturbance, upper motor neuron paresis/plegia and circling [2, 12–14, 22, 25, 28, 29]. The present case had only facial palsy as a clinical sign and did not show any other neurological abnormalities. Previous two reports [12, 13] described brainstem tumor cases which located in similar part of the brainstem. The first case [12] showed circling with cranial nerve deficits and the second case [13] showed circling with ipsilateral hemiparesis and cranial nerve deficits. Although the cause of clinical differences between two cases was uncertain, it could support the theory that any cases with the same brainstem lesions could reveal some different neurological signs. One other previous report [22] describes two patients who developed a gait disturbance in addition to facial palsy and that an arachnoid cyst in the caudal fossa impinged only the facial nerve, although MRI findings indicated that the eighth and facial nerves are impinged. Furthermore, another report described a dermoid cyst located on the ventral surface of the brainstem and the patients had only slight headache but no neurological deficits were detected [2]. Although our patient had normal postural reaction, unilateral facial paresis could be an isolated clinical presentation of brainstem cystic lesion. There is no fundamental medical treatment which can reduce the size of the IAC. Instead, medical therapy showed some clinical improvement through reducing increased intracranial pressure and brain edema but the responses are variable [5]. Surgical intervention is considered in cases of symptomatic IAC due to dogs with IAC may respond to medical therapy temporarily [5, 19]. The patient in this case did not respond to the medical treatment. However no additional neurological abnormalities were detected and the physical and mental status of the patient were maintained without continuous medical treatment or surgery until 3 months after diagnosis.

IACs in dogs are sporadically reported and are usually located in the quadrigeminal cistern [5, 21, 32]. A canine report describing a medullary dermoid cyst was presented in 1998 [29]. To our knowledge, there are no reports describing canine brainstem epidermoid cysts or IAC at this time [20]. Although a definitive diagnosis of IAC is only possible with histopathology, IAC can be differentiated from dermoid and epidermoid cysts by MRI with FLAIR sequences.

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