Cervical syringohydromyelia secondary to a brainstem tumor in a dog

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- Syringohydromyelia is rare, and although it is considered to be associated with congenital diseases, improved imaging techniques have revealed that it can also develop as an acquired disorder; thus, diagnosis of syringohydromyelia in an adult dog indicates a need to search for an underlying disease condition.
- Acquired syringohydromyelia can be the result of tumors of the caudal fossa, especially meningiomas; cerebellar herniation is fundamental for its formation and progression.
- In dogs that may have space-occupying lesions of the caudal fossa, the cervical portion of the spinal cord should be examined via magnetic resonance imaging or other imaging techniques to investigate whether syringohydromyelia is present.

An 11-year-old sexually intact male Pekingese was evaluated at the **Veterinary Teaching Hospital of the Ontario Veterinary College** (**VTH-OVC**) because of a head tilt. Five months prior to this evaluation, the dog had been examined by the referring veterinarian because of a head tilt to the right. At that time, a presumed diagnosis of geriatric vestibular disease was made. The head tilt had resolved according to the owner; however, 3 months after the examination by the referring veterinarian, an examination was performed at the VTH-OVC for other health problems (coughing and vomiting) during which a right-sided head tilt, ataxia, and scoliosis with concave deviation of the vertebral column to the right were observed. No further neurologic diagnostic testing was performed at that time.

On evaluation at the VTH-OVC (2 months after the evaluation for coughing and vomiting), the dog was alert and responsive but would fall asleep when not stimulated. Other neurologic abnormalities detected included a head tilt to the right, marked ataxia, scoliosis of the cervicothoracic portion of the vertebral column with concave deviation to the right, rotatory nystagmus with the fast phase to the left, and proprioceptive positioning deficits in all limbs (more pronounced in the fore- and hind limbs on the right side). The extent of the scoliosis had remained unchanged over the past 2 months. The signs of involvement of the vestibular nerve or nuclei, somnolence (presumed to be a consequence of the involvement of the ascending reticular activating system), and predominantly right-sided proprioceptive deficits suggested that damage was localized to the brainstem (more specifically at the level of the right rostral medulla).

The results of a CBC and serum biochemistry profile were within reference ranges. Findings of a brainstem auditory evoked response test of the left ear were unremarkable, whereas a flat trace was recorded during testing of the right ear, which suggested peripheral impairment of the auditory function at the level of the cochlear receptors.

A **magnetic resonance imaging** (**MRI**) scan of the brain was recommended and performed[a] at a private neuroimaging facility. Assessment of the images revealed a large 2 × 1.5-cm extra-axial, broad-based mass on the floor of the brainstem, mainly on the right side, that extended from the caudal portion of the midbrain to the caudal aspect of the medulla. The mass appeared hypointense on T1-weighted images and hyperintense on T2-weighted images. It was well demarcated and surrounded by moderate edema. After an IV injection of contrast medium (gadopentetate dimeglumine),[b] the lesion became greatly enhanced, except for a central area (**Figure 1**). A cerebellar herniation was detected on the T1-weighted sagittal images. The lateral and third ventricles were enlarged, whereas the fourth ventricle was compressed and displaced to the left side. An enlargement of the central canal was observed in the first and part of the second cervical spinal cord segments on the T1-weighted sagittal images. Unfortunately, other cervical segments of the spinal cord were not included in the initial MRI scan. The brainstem lesion detected via MRI was highly suggestive of a neoplastic process. Because of the extra-axial location of the mass and its enhancement after IV injection of the contrast agent, the main differential diagnosis was meningioma. Other tumors such as astrocytoma or oligodendroglioma were also considered because contrast enhancement of the mass was unequal in the central region. Other less likely diagnoses included abscess and granuloma formation.

The dog was given an IV injection of dexamethasone (0.25 mg/kg [0.11 mg/lb]), followed by oral administration of prednisone (0.5 mg/kg [0.23 mg/lb]) every 24 hours. Surgical treatment was considered impractical because of the tumor location. Radiation therapy administered with a cobalt 60 unit was initiated 2 weeks after diagnosis. The dog received 10 fractions of 2.25 Gy, followed by 8 fractions of 2.5 Gy to a total dose of 42.5 Gy over a 4-week period. Administration of prednisone (0.5 mg/kg, PO, q 24 h) was continued throughout radiation treatment and the

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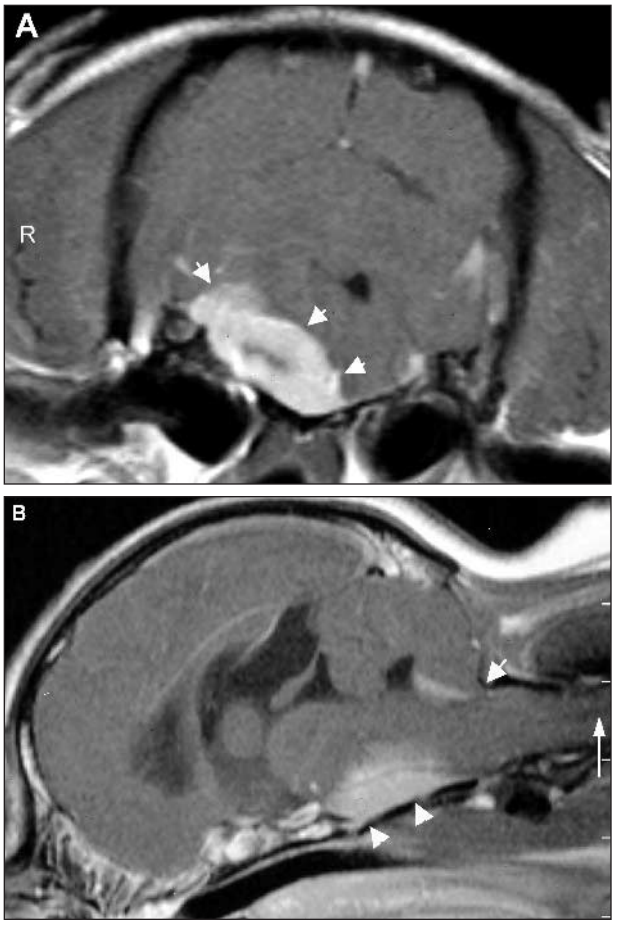

Figure 1—Transverse (A) and sagittal (B) T1-weighted images (obtained after IV administration of contrast medium [gadopentetate dimeglumine]) of the head at the level of the rostral portion of the medulla of a dog with a head tilt to the right, marked ataxia, scoliosis of the cervicothoracic portion of the vertebral column (concave deviation to the right), rotatory nystagmus (fast phase to the left), and predominantly right-sided proprioceptive positioning deficits. In panel A, an extra-axial, broad-based enhanced mass is present on the right, extending to the left (arrows). A nonenhanced central area can be observed in the center of the mass. The fourth ventricle is compressed and displaced to the left. R = Right side of the dog. In panel B, the mass extends from the midbrain to the caudal medulla (arrowheads). A cerebellar herniation (arrow) and a dilatation of the central canal (long arrow) are detected.

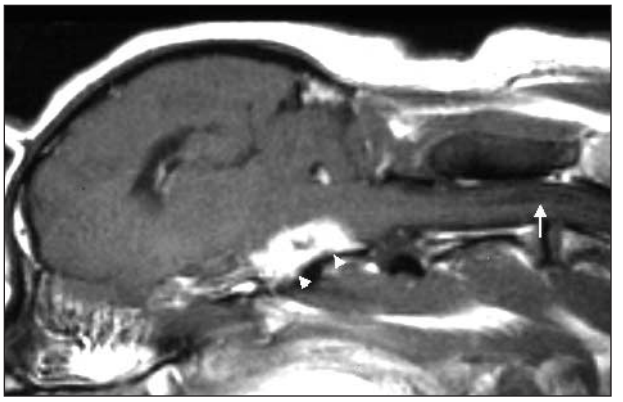

Figure 2—Sagittal T1-weighted image of the dog in Figure 1 after IV administration of gadopentetate dimeglumine, performed 9 weeks after radiation treatment. A dilatation of the central canal is detectable from the first to the third cervical spinal cord segment. This dilatation reaches its maximum size at the caudal aspect of the second cervical vertebra (arrow). The brainstem mass is also observed (arrowheads).

Figure 3—Transverse T1-weighted image of the dog in Figure 1 at the level of the caudal end of the second cervical vertebra. Notice an eccentric central canal dilatation that is suggestive of syringohydromyelia. The syringohydromyelia extends dorsally to the left side (arrow). R = Right side of dog.

following 2 weeks. No adverse effects of radiation treatment were observed other than mild dermatitis at the irradiated site and within the right ear canal. A neurologic reevaluation was performed 1 and 4 weeks after the end of radiation therapy. One week after treatment, mild ataxia, head tilt, and slightly delayed proprioceptive positioning deficits were observed, but nystagmus was no longer present. By the fourth week, the head tilt, scoliosis, and proprioceptive positioning deficits were undetectable, but mild ataxia persisted. The animal was neurologically normal 2 months after radiation therapy.

A second MRI evaluation was performed 9 weeks after radiation treatment. The cervical portion of the spinal cord was included in the scan. The reduction of the mass size was estimated at 30% to 40%. The dilatation of the ventricular system rostral to the lesion was still present. The cerebellar herniation had improved but not totally resolved. On the T1-weighted sagittal images, a linear hypointense area (centrally located within the spinal cord) that extended from the first to the third cervical spinal cord segment was detected (**Figure 2**); this was considered to be a dilatation of the central canal that reached maximum enlargement at the transition of the second and third cervical spinal cord segments. The T1-weighted transverse spinal cord images confirmed the central location of the enlargement in the cervical portion of the spinal cord, which extended dorsally to the left at the level of the second cervical spinal cord segment (**Figure 3**). This lesion was suggestive of a syrinx formation. These MRI findings supported a diagnosis of cervical syringohydromyelia.

Six months after the end of radiation treatment and 4 months after the second MRI scan, a third MRI examination was performed. At that time, the dog continued to be clinically normal. Compared with the results of the second MRI scan, the findings of the third

MRI examination indicated a slight reduction of the tumor size and resolution of the cerebellar herniation. The syringomyelia detected on the previous images could no longer be seen. A mild dilatation of the central canal remained, but the eccentric cavity could no longer be detected (**Figure 4**).

Syringohydromyelia is characterized by a longitudinal cavity within the spinal cord that extends over several vertebral segments.[1] Typically, the syrinx contains fluid that is highly similar in composition to the CSF and extracellular fluid. On histologic examination, the cavity may be a dilatation of the central canal and lined with ependymal cells (characteristic of hydromyelia), or it may lie within the parenchymal substance and be lined with glial cells (characteristic of syringomyelia).[2] Although hydromyelia can be differentiated from syringomyelia on the basis of these histologic features, in some instances, when a hydromyelic cavity expands it can disrupt the ependymal layer and may be lined with glial tissue consistent with a syringomyelic cavity.[3] Because of the difficulty in distinguishing syringomyelia from hydromyelia clinically, a combination of terms such as syringohydromyelia has been proposed.[2] A careful clinical evaluation of patients with syringomyelia will, in almost all instances, reveal an associated disorder characterized by either obstruction of CSF flow, tethering of the spinal cord, or spinal tumor.[2] The reported causes of syringomyelia can be classified as diseases of the craniocervical junction or diseases of the spinal cord. An association between syringomyelia and brain tumors, although rare, has been identified in humans.[4-6] In humans, several brain tumors including astrocytomas,[7] gliomas,[8,9] medulloblastomas,[4,7] epidermoid tumors,[6,10] and most commonly menigiomas[4,5,7,11-14] have been described as causes of syringomyelia. To the authors' knowledge, there is no report of intracranial tumors and associated syringomyelia or syringohydromyelia in the veterinary medical literature. In fact, there are only a few reports of syringohydromyelia in dogs and cats.[15-22]

The clinical signs of syringohydromyelia reflect spinal cord dysfunction and may include ataxia, paresis, signs of spinal pain, and scoliosis.[3] In some reports[16,18] of syringohydromyelia in dogs, it has been suggested that scoliosis is the major clinical sign; however, in most reports,[15,17,20-22] ataxia and paresis with no signs of scoliosis are identified as the main clinical findings. Signs of neck and ear pain (with persistent scratching of the shoulder region) have also been reported in dogs with syringohydromyelia.[22] In humans with infratentorial neoplasms, development of syringohydromyelia is considered incidental to the cause of the neurologic deficit; patients have clinical signs attributable to the tumor only and none attributable to syringohydromyelia.[5,7,8,11-13] Only a few reports[6,14] describe syringohydromyelia with associated clinical signs secondary to brain tumors. In a study[23] involving 105 humans with syringohydromyelia (all causes), an absence of symptoms was reported for 22% of the individuals. In the dog of this report, vestibular signs were the main reason for consultation. The scoliosis was more likely caused by the vestibular dysfunction than by the syringohydromyelia. Although a syrinx was still present 9 weeks after radiation therapy, the dog had no scoliosis, head tilt, or ataxia at that time. The presence of vestibular signs in association with somnolence and proprioceptive deficits indicated a brainstem lesion; therefore, a syrinx in the cervical portion of the spinal cord was not suspected initially. As observed in most humans with syringomyelia secondary to brain tumors,[5,7,8,11-13] clinical signs associated with the brain mass were predominant, and there were no clinical signs of syringohydromyelia in the dog of this report. It was initially questioned whether the syrinx was associated with the brainstem mass, or whether it was secondary to spinal cord disease, or a congenital disorder. The dog had no known history of trauma to the cervical portion of the vertebral column, and MRI performed after radiation treatment did not reveal any evidence of spinal cord disease located caudal to the syrinx. Moreover, the syrinx was not detected on the third MRI scan. Therefore, it is logical to assume that the brainstem mass caused the cerebellar herniation, resulting in the syrinx formation, and that its resolution was associated with shrinkage of the tumor and return of the cerebellum to a normal position.

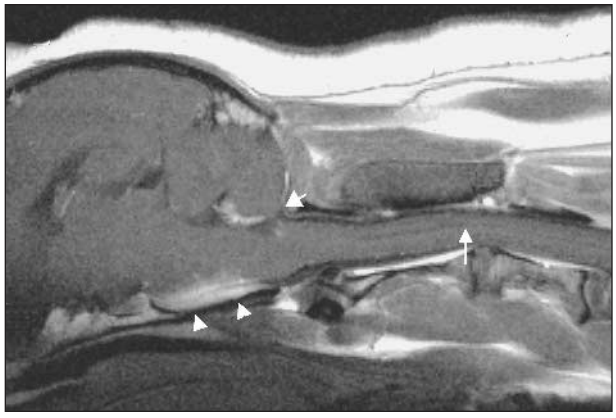

Figure 4—Sagittal T1-weighted image of the dog in Figure 1 after IV administration of gadopentetate dimeglumine, 6 months after radiation treatment. Cerebellar herniation is no longer detectable (arrow). The brainstem mass is also smaller, compared with equivalent images obtained previously (arrowheads). Compared with the image in Figure 2, minimal dilatation of the central canal is present (long arrow). The placement of the sagittal section is similar to that depicted in panel B of Figure 1.

It is important to note the association between syringohydromyelia and caudal fossa masses. A lesion in the brainstem or in the cervical portion of the spinal cord can cause both ataxia and proprioceptive deficits. In animals with space-occupying lesions in the brainstem, syringohydromyelia may remain undetected if the cervical portion of the spinal cord is not evaluated. Because the signs of cervical syringohydromyelia can also precede signs associated with a brainstem mass,[6] the finding of cervical syringohydromyelia should prompt investigation of possible caudal fossa disorders.

An association of hydrocephalus with syringohydromyelia has been reported in humans[23,24] and dogs,[19-21] and hydrocephalus was present in the dog of this report. However, in this dog, no clinical signs were attributed to hydrocephalus.

Among the MRI techniques, T1-weighted images are the most useful for assessment of morphologic and cystic changes within the spinal cord.[25] However, even with this advanced imaging technique, differentiation of

syringomyelia from hydromyelia is difficult. Eccentric cavity location within the spinal cord may be more suggestive of syringomyelia, but this is not pathognomonic.[25]

Recently, the pathogenesis of syringomyelia has been reevaluated.[2,26] In humans with Chiari type-I malformation[26] and with brain tumors,[6,7,11,12] cerebellar herniation results in an anatomic and physiologic block to the flow of CSF. Expulsion of CSF into the cervical subarachnoid space is the normal compensatory mechanism in response to brain expansion during cardiac systole. Brain expansion (during systole) forces the cerebellar tonsils into the partially enclosed subarachnoid space, which then act as a piston. Because the partially enclosed subarachnoid space has low compliance to abrupt changes in volume, tonsillar descent results in increased cervical subarachnoid pulse pressure waves that compress the spinal cord externally.[26] This forces the extracellular fluid through the wall of the spinal cord into the spinal cord parenchyma. The fluid migrates through the Virchow-Robin spaces (ie, there is free communication between the subarachnoid space and the perivascular space) at the pia mater, predominantly at the root entry zones.[2] The final consequence is a collection of extracellular fluid within the spinal cord, either in the parenchyma or in the central canal.[2] This recent theory suggests that a patent communication between the fourth ventricle and the central canal is not essential for syrinx formation as previously believed.[2] The more frequent formation of syringes in the cervical portion of the spinal cord has been explained as a consequence of higher CSF flow velocities in the cervical and thoracic portions of the spinal cord, compared with that in the lumbar region.[27,28]

There are several reports[5,6,11-13] of resolution of syringomyelia after surgical removal of the infratentorial tumor in humans. In the dog of this report, resolution of syringohydromyelia occurred after radiation treatment of the brainstem mass; it appears that the reduction of the mass size (by 30% to 40%) after radiation therapy was enough to allow resolution of the cerebellar herniation and promote better CSF flow. To the authors' knowledge, resolution of syringohydromyelia after radiation treatment of a brainstem tumor in animals or humans has not been previously reported.

The advent of MRI and its use in veterinary medicine has increased our awareness of the frequency of syringohydromyelia. In humans and animals, detection of syringohydromyelia often indicates an underlying pathologic process, and a primary cause should be sought. As the findings in the dog of this report highlight, syringohydromyelia is a possible complication of caudotentorial masses. It is recommended to include the cervical portion of the spinal cord in MRI scans when a patient is suspected of having a brainstem or cerebellar space-occupying lesion. Moreover, whenever syringohydromyelia is observed via MRI, a primary cause should be sought cranial or caudal to the affected region, justifying a complete neuroimaging evaluation.

[a]Magnetom Vision, Siemens Canada, Mississauga, ON.
[b]Magnevist, Berlex Canada Inc, Lachine, QC.

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