What Is Your Diagnosis?

History

A 4-year-old 27-kg (59.5-lb) sexually intact female Labrador Retriever was referred for evaluation because of a 2-year history of tetraparesis and incoordination that had been progressing slowly over that period. The dog had been treated with several NSAIDs and tramadol, with no improvement in clinical signs. Two days prior to referral, the patient’s neurologic status acutely deteriorated.

Neurologic examination revealed severe ambulatory tetraparesis and generalized proprioceptive ataxia. Postural reactions were absent in all 4 limbs, and segmental spinal reflexes were normal. No cranial nerve deficits were detected, and there was no sign of pain on spinal manipulation. Neurologic diagnosis and neuroanatomical localization was myelopathy at C1 through C5.

No abnormalities were detected on CBC, serum biochemical analysis, and 3-view thoracic radiography. A CSF sample was obtained by lumbar puncture; CSF analysis revealed a total nucleated cell count that was within reference range (3 cells/µL; reference range, < 5 cells/µL) and a high total protein concentration (0.64 g/L; reference range, < 0.45 g/L). Findings on cytologic evaluation of CSF were unremarkable. Computed tomographic myelography of the cervical vertebral column was performed (Figure 1).

Determine whether additional imaging studies are required, or make your diagnosis from Figure 1—then turn the page →
Figure 2—Same CT myelograms as in Figure 1. A—A severe intramedullary pattern (arrow) and spinal arachnoid diverticulum (asterisk) are evident. B—Notice the dorsal bone remodeling of the laminae of C5 (arrows).

Diagnostic Imaging Findings and Interpretation

Notice the extensive intramedullary enlargement over the caudal portion of C2 and cranial portion of C3 with marked circumferential attenuation of the contrast columns around the spinal cord (Figure 2). In addition, the dorsal and dorsolateral aspects of the subarachnoid space are enlarged from the mid portion of C3 to the cranial aspect of C6. The inner cortices of the laminae and pedicles of C3, C4, and C5 are remodeled, presumably resulting from pressure atrophy. These findings are consistent with a large intramedullary lesion at the level of C2 and C3 and a spinal arachnoid diverticulum extending from C3 to C6.

For further evaluation, low-field MRI was performed (Figure 3). A fusiform, well-defined, intramedullary lesion extending from the caudal portion of C2 to the cranial aspect of C3 was evident. Compared with unaffected areas of the spinal cord, the lesion was slightly hyperintense on T1-weighted images and strongly hyperintense on T2-weighted images. Postcontrast images provided strong and homogeneous enhancement of the lesion. A dilation of the dorsal and dorsolateral subarachnoid space extending from the mid portion of C3 to the end of the field of view (ie, mid portion of C5) was also observed.

Figure 3—Magnetic resonance images of the cervical vertebral column of the dog in Figure 1. On the midsagittal precontrast T1-weighted (A), precontrast T2-weighted (B), and postcontrast T1-weighted (C) images of C2 through C5, there is a fusiform, well-defined, intramedullary lesion (arrow). The mass is strongly and homogeneously contrast enhanced in the postcontrast T1-weighted image (C). A spinal arachnoid diverticulum (asterisk) is also evident. On the transverse postcontrast T1-weighted image (D), there is strong contrast enhancement of the lesion at the level of C2.
The main differential diagnoses for these imaging findings included neoplastic disease (ependymoma, astrocytoma, oligodendroglioma, lymphoma, and metastatic neoplasia), vascular malformation (spinal hamartoma), and inflammatory or infectious diseases (myelitis), with an associated spinal arachnoid diverticulum.

Treatment and Outcome

Because of the severity of clinical signs and guarded to poor prognosis, the owner declined further treatment and elected to euthanize the dog. Only a postmortem examination of the cervical spinal cord was allowed.

Grossly, a large and strictly intramedullary, gelatinous, grayish mass of ill-defined margins was observed. Microscopically, the mass consisted of a neoplastic cell population growing densely in the gray matter. The neoplastic cells were small, were moderately pleomorphic, and had distended nuclei with diffuse chromatin and slightly eosinophilic cytoplasm. The mitotic index was low (0 to 1 mitotic figure/hpf). The adjacent white matter was severely compressed, and there was swelling, dilation, and segmentation of nerve fibers. Curiously, the neoplastic cells invaded the white matter growing around blood vessels. Immunohistochemistry results for laminin and vimentin were positive, and immunohistochemistry results for neuronal and glial cell markers were negative. All these findings, together with the absence of nerve root or spinal nerve involvement, were consistent with an intramedullary schwannoma.

Comments

Intraspinal schwannomas in humans and dogs are usually intradural-extradural or extradurally located.1 In humans, intramedullary schwannomas are rare and account for 0.3% of all intraspinal tumors and 1.1% of intraspinal schwannomas. A male predisposition is reported, and the most common location is the cervical spinal cord (58%), followed by the thoracic (32%) and lumbar spinal cord (10%).2 To our knowledge, this is the first description of the diagnostic imaging and histologic features of an intramedullary schwannoma in a dog.

Intramedullary schwannomas in humans are usually seen as hypointense masses on T1-weighted images, with or without an inner isointense area. On T2-weighted images, they are usually hyperintense.2 Other MRI features include strong contrast enhancement and sharply delineated borders.3

The MRI findings of the case described in the present report were similar to those reported for humans,2 except that the lesion was slightly hyperintense on precontrast T1-weighted images. However, some intramedullary schwannomas in humans can be hyperintense on T1-weighted images (ie, melanotic schwannomas).3 For the dog of the present report, the signal on the precontrast T1-weighted images remains unchanged because no melanotic deposits, hemorrhage, or other substances that could cause a T1-weighted hyperintense signal were observed on histologic evaluation.

The origin of intramedullary schwannomas remains controversial. The absence of Schwann cells within the spinal cord is widely accepted, but aberrant proliferation of nerve fibers and Schwann cells (ie, schwannosis) in the perivascular spaces of the spinal cord has been documented secondary to physical injury and diabetes mellitus.3 Other hypotheses about the origin of these tumors suggest that they develop from Schwann cells of the posterior nerve root at the root entry zone, the subpial extension of Schwann cells along the perivascular nerve plexus of the spinal cord vessels, the ectopic neural crest cells during embryogenesis, or Schwann cells that differentiated from multipotential mesenchymal elements of the CNS.2 In the case described in the present report, lack of involvement of the spinal nerve roots and localization of the tumor within the spinal cord gray matter mainly, with a particular perivascular growing pattern, may suggest a possible perivascular nerve plexus origin.

Spinal arachnoid diverticula are focal fluid-filled dilations of the subarachnoid space that can cause a progressive compressive myelopathy and are most common in young dogs in the absence of other lesions. The disease process is uncertain, but they may result from a developmental abnormality in the arachnoid architecture.4 Acquired spinal arachnoid diverticulum may also occur secondary to other disease processes, such as chronic spinal cord compression and inflammatory spinal cord disease.4 Intramedullary schwannoma is a slow-growing, benign tumor 2 that could have been the cause of the development of the arachnoid diverticulum in the dog of the present report. The bone remodeling observed in the CT myelograms in the laminae of C3, C4, and C5 supported the hypothesis of a chronic progressive compression caused by the diverticulum. In fact, bony wall changes resulting from chronic pressure caused by slowly expanding arachnoid brain cysts are well described in humans.5,6

The dog of the present report had a history of chronic ambulatory tetraparesis and acute worsening of clinical signs 2 days prior to referral. Because of the similarity in expected clinical signs of intramedullary schwannoma and spinal arachnoid diverticulum, it was impossible to know whether the spinal arachnoid diverticulum developed secondary to the neoplasia, the neoplasia developed secondary to the spinal arachnoid diverticulum, or the 2 lesions developed independently.

Schwannomas are histologically benign neoplasms and a good clinical outcomes after gross total resection or subtotal removal is reported for humans;2 thus, surgical excision might be considered in dogs as well. In conclusion, intramedullary schwannoma, although rare, should be considered as a differential diagnosis for intramedullary lesions in dogs.

References