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What Is Your Neurologic Diagnosis?

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A 7-month-old 20.4-kg sexually intact male Hungarian Vizsla was referred for evaluation because of a 1-month history of nonprogressive ataxia affecting all 4 limbs with no signs of pain or discomfort. General physical examination findings were unremarkable apart from marked scuffing of the claws of all 4 limbs.

Neurologic examination

Observation

Mental	Alert	X	Depressed		Disoriented		Stupor		Coma		
Posture	Normal	X	Head tilt		Tremor		Falling		Other		
Gait	Normal		Ataxia	X	Pelvic limbs		All 4		Circling		
Paresis	Pelvic limbs		Tetra	X	Hemi		Mono				
Other	Mild hypermetria of the thoracic limbs										

Key: 4 = Exaggerated, clonus; 3 = Exaggerated; 2 = Normal; 1 = Diminished; 0 = None; NE = Not evaluated.

Postural reactions	Left forelimb	Right forelimb	Left hind limb	Right hind limb
Wheelbarrow	NE	NE		
Hopping	1	1	1	1
Extensor postural thrust			NE	NE
Proprioceptive positioning	2	2	2	2
Hemistand/walk	NE	NE	NE	NE
Placing–tactile	NE	NE		
Placing–visual	NE	NE		

Spinal reflexes	Left forelimb	Right forelimb	Left hind limb	Right hind limb
Quadriceps			2	2
Extensor carpi	2	2		
Flexion	2	2	2	2
Crossed extensor	2	2	2	2
Perineal			2	2

Cranial nerves

	L	R
II, VII–Vision menace	2	2
II, III–Pupils resting	2	2
Stim L	2	2
Stim R	2	2
II–Fundus	2	2
III, IV, VI–Strabismus, resting	2	2
III, IV, VI, VIII–Strabismus, position	2	2

	L	R
VIII–Nystagmus, resting	2	2
VIII–Nystagmus, change	2	2
V–Sensation	2	2
VII–Facial mm	2	2
V, VII–Palpebral flex	2	2
IX, X–Gag	2	2
XII–Tongue	2	2

Comments
No abnormalities

Sensation (Locate and describe any abnormality)

Hyperesthesia	0	
Superficial pain	2	
Cutaneous reflex	2	
Deep pain	NE	

Make your assessment, then continue reading.

Assessment

Anatomic diagnosis

For the proprioceptive ataxia of all 4 limbs and ambulatory tetraparesis, a C1-C5 or C6-T2 myelopathy could be considered on the basis of gait evaluation findings. Given the normal muscle tone and segmental spinal reflexes, a C6-T2 lesion was excluded. A brainstem lesion was deemed unlikely because of the dog's normal mentation and lack of cranial nerve deficits.

For thoracic limb hypermetria, a C1-C5 myelopathy or cerebellar lesion was considered. However, a cerebellar lesion was thought to be unlikely owing to a lack of other deficits suggestive of such a lesion, such as intention tremors or an abnormal menace response.

The decreased hopping in all 4 limbs could have been a result of C1-C5 or C6-T2 myelopathy. However, C6-T2 myelopathy and a brainstem lesion were excluded for the aforementioned reasons.

Likely location of a single lesion

The most likely location of a single lesion was the C1-C5 spinal cord segments.

Etiologic diagnosis

Primary differential diagnoses for insidious onset of C1-C5 myelopathy in a young dog included developmental (vertebral malformations or syringomyelia) and degenerative causes. Given the absence of signs of pain and lack of disease progression, traumatic, infectious (eg, protozoal infection or spinal epidural empyema), and inflammatory (eg, immune-mediated) causes were considered less likely. However, a lesion related to previous trauma (vertebral fracture) or an acute noncompressive nucleus pulposus extrusion or fibrocartilaginous embolus could not be ruled out. Considering the young age of the dog, neoplasia was considered unlikely but lymphoma, primary vertebral tumor (osteoma or osteosarcoma), and pseudoneoplastic lesions (eg, hamartoma) were considered as possible differential diagnoses.

Diagnostic Test Findings

Results of a CBC and serum biochemical analysis were unremarkable. Computed tomography and MRI of the cervical portion of the vertebral column were performed after the dog was anesthetized. The CT images revealed separation of the odontoid process from the body of C2 **(Figure 1)**. The odontoid process was centered over the ventral arch of C1. The vertebral canal at this level appeared to be 50% narrower than the vertebral canal at C2-3 level. Additionally, there was a fusion of the C2-C3 vertebrae (block vertebrae). Flexion of the neck resulted in dorsal displacement of C1 relative to C2, resulting in closure of the space between the dorsal aspects of C1 and C2 and further narrowing of the vertebral canal. The position of the odontoid process remained static in relation to the ventral aspect of C1. The CT diagnosis was separation of the odontoid process from the

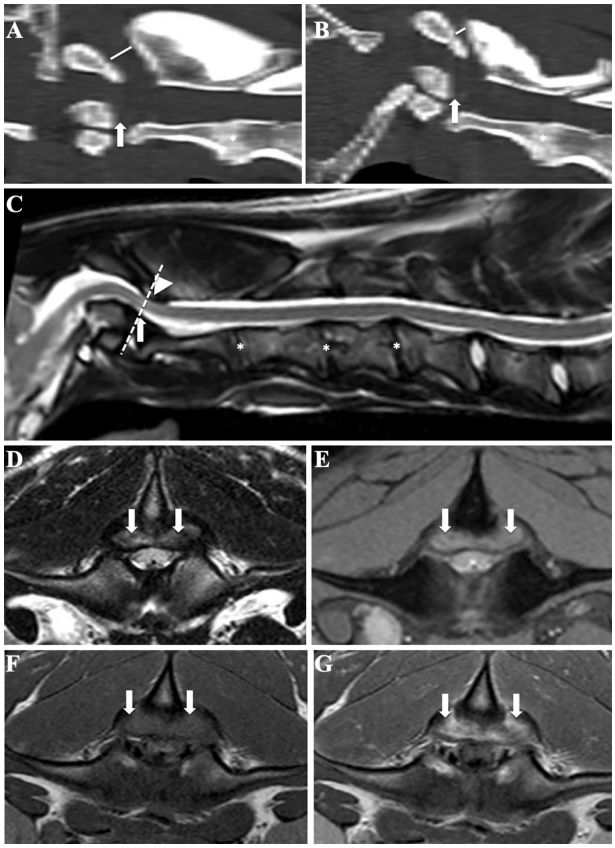

Figure 1—Computed tomographic and MRI images of the cervical portion of the vertebral column of a 7-month-old Hungarian Vizsla that had a 1-month history of nonprogressive ataxia affecting all 4 limbs. A and B—Sagittal CT reconstructions of the cranial cervical portion of the vertebral column at the level of the atlantoaxial junction obtained with the dog's neck in a neutral position (A) and in a flexed position (B). Notice the separation of the odontoid process from the body of C2 (arrows). Additionally, there is a fusion of the C2 and C3 vertebrae (asterisk). Flexion of the neck resulted in dorsal displacement of C1 relative to C2, leading to closure of the space between the dorsal aspects of C1 and C2 and further narrowing of the vertebral canal (white lines). C—Sagittal T2-weighted image of the cervical portion of the dog's vertebral column in the flexed position, confirming the separation of the odontoid process from the body of C2. There is a hypointense band connecting the odontoid process and C2 that extends over the dorsal aspect of the odontoid process and reaches the craniodorsal aspect of C2 (arrow). At this imaging level, the spinal cord has a sigmoid shape and is dorsoventrally flattened. There is a mild dorsal compression at the level of the dorsal articulation between C1 and C2 (arrowhead). Additionally, there is absence of an intervertebral disk signal between the C2 and C3 vertebrae, C3 and C4 vertebrae, and C4 and C5 vertebrae (asterisks). D and E—Transverse T2-weighted (D) and T2*-weighted (E) MRI images obtained at the level of the dotted line in panel C. Notice the dumbbell-shaped structure that has caused dorsal spinal cord compression (arrows). This structure appears to be heterogeneously hyperintense (compared with the surrounding muscles) with irregular iso- to hypointense margins. F and G—Transverse T1-weighted MRI images obtained before (F) and after (G) contrast administration; images were obtained at the level of the dotted line in panel C. The dumbbell-shaped structure is isointense (F) and has strong, patchy, heterogeneous contrast enhancement (G).

body of C2 (suspected os odontoideum [OO]) with secondary atlantoaxial (AA) instability and vertebral canal stenosis at the C1-C2 level.

Sagittal MRI (Figure 1) confirmed the separation of the odontoid process from the body of C2. The odontoid process was centered over the ventral arch of C1. There was a hypointense band that connected the odontoid process and C2 and extended over the dorsal aspect of the odontoid process to the craniodorsal aspect of C2. At this imaging level, the spinal cord had a sigmoid shape and was dorsoventrally flattened. There was loss of the CSF signal in the dorsal aspect of the spinal cord at the level of the AA junction; in the ventral aspect of the spinal cord at the level of the AA junction, there was attenuation of the CSF signal with associated focal, ill-defined, T2-weighted intramedullary hyperintensity. At the level of the dorsal articulation between C1 and C2, there was mild dorsal compression. On T2-weighted and T2*-weighted transverse images, there was a dumbbell-shaped structure at the level of the dorsal articulation between the caudal aspect of the dorsal arch of C1 and the cranial margin of the lamina of C2 (ie, in the anatomic location of the dorsal AA ligament). This structure appeared to be heterogeneously hyperintense, compared with the surrounding muscles, with irregular iso- to hypointense margins. On T1-weighted images, the structure appeared to be isointense and had strong, patchy, heterogeneous contrast enhancement. On T2-weighted images, the spinal cord appeared to be hyperintense, corresponding to the focal hyperintensity evident on the sagittal images. There was an absence of the odontoid process in its anatomic location on transverse images. Additionally, there was an absence of the intervertebral disk signal between the C2 and C3 vertebrae (block vertebrae).

Magnetic resonance imaging confirmed the separation of the odontoid process (suspected OO) and AA instability with associated intramedullary spinal cord changes compatible with spinal cord edema or gliosis and proliferative changes of the connective tissue at the level of the dorsal C1-C2 articulation and dorsal AA ligament.

The main differential diagnosis for OO is a fracture of the odontoid process. Os odontoideum can be distinguished by the smooth surface of the ossicle. An acute fracture has clear, sharp fracture lines, whereas cortical discontinuity and callus formation are associated with old fractures. Additionally, in an acute traumatic event, secondary pathologic changes, such as hemorrhage and soft tissue injury, are expected.

Comments

Os odontoideum is a rare anomaly of the odontoid process.[1] Radiographically, OO is defined as an independent ossicle of variable size with smooth circumferential cortical margins separated from C2.[2] In the veterinary medical literature, there is only 1 report[3] describing cervical radiographic and postmortem examination findings of OO in 2 small-breed dogs. However, OO may be underreported in the veterinary medical literature because dens separation is often cited as a cause of AA instability.[4]

Congenital AA instability is a common condition in toy-breed dogs but is less often encountered in large-breed dogs. In affected dogs, AA instability can be identified on survey cervical radiographic views.[4] Typical radiographic findings consistent with AA instability include dorsal displacement of C2 into the vertebral canal and an increased distance between the arch of C1 and the spinous process of C2. In general, underlying odontoid process anomalies can also be identified. Often, various craniocervical junction anomalies can be present concomitantly with AA instability.[4] Advanced imaging, such as CT and MRI, allows detailed evaluation of soft tissue and bony components of these complex conditions. The best imaging modality for assessing bones is CT with multiplanar reconstructions. A reformatted sagittal CT scan can show the precise location of the odontoid process and its position in relation to adjacent structures as well as the presence and degree of AA instability. Computed tomography is also useful for assessment of anatomic landmarks, and 3-D reconstruction of images enables surgical planning. Dynamic imaging, when performed in a safe manner, can provide an accurate diagnosis of AA instability.[5] Interestingly, for the dog of the present report, flexion of the neck (Figure 1) resulted in dorsal displacement of C1 relative to C2 and closed the space between the dorsal aspects of those vertebrae. In classic cases of AA instability, an increase in distance between the dorsal arch of C1 and the spinous process of C2 is expected. In humans with OO, AA instability is defined as a change in C1 translation from the neutral position, which occurred in the dog of the present report.[6] Moreover, flexion of the dog's neck resulted in the movement of the OO and atlas in the same direction with respect to C2, thereby causing further narrowing of the vertebral canal.

In contrast to CT, MRI has superior soft tissue resolution and therefore provides information not only about OO and AA instability, but also the degree of spinal cord compression and signal changes of the spinal cord in affected animals. Atlantoaxial instability is usually best appreciated on sagittal T2-weighted images. For the dog of the present report, apart from the dorsoventral flattening and intramedullary hyperintensity of the spinal cord (the latter most likely compatible with gliosis), MRI revealed proliferative changes at the level of the dorsal AA articulation, which could have represented proliferation of connective tissue secondary to chronic instability.

The pathogenesis of OO remains controversial, with theories regarding both congenital and acquired processes. Os odontoideum can be classified into 2 anatomic types: orthotopic and dystopic. Orthotopic OO is defined as an ossicle that moves with the anterior arch of C1, and dystopic OO is defined as an ossicle that is functionally fused to the midline of the ventral aspect of the foramen magnum.[1] The diagnostic imaging findings in the case described in

the present report were compatible with orthotropic OO and associated AA instability.

For the dog of the present report, treatment options included surgical decompression and AA stabilization or conservative management. The owners declined surgical treatment. The dog's neurologic status remained unchanged at the 6- and 15-month follow-up evaluations.

References

1. Jumah F, Alkhdour S, Mansour S, et al. Os odontoideum: comprehensive clinical and surgical review. *Cureus*. 2017;9(8):e1551. doi:10.7759/cureus.1551
2. Wang Q, Dong S, Wang F. Os odontoideum: diagnosis and role of imaging. *Surg Radiol Anat*. 2020;42(2):155–160.
3. Ladds P, Guffy M, Blauch B, Splitter G. Congenital odontoid process separation in two dogs. *J Small Anim Pract*. 1971;12(8):463–471.
4. Slanina MC. Atlantoaxial instability. *Vet Clin North Am Small Anim Pract*. 2016;46(2):265–275.
5. White DA, Renberg WC, Roush JK, Hallman MR, Mauler DA, Milliken GA. Flexed radiographic angles for determination of atlantoaxial instability in dogs. *Vet Surg*. 2019;48(8):1406–1415.
6. Klimo P Jr, Kan P, Rao G, Apfelbaum R, Brockmeyer D. Os odontoideum: presentation, diagnosis, and treatment in a series of 78 patients. *J Neurosurg Spine*. 2008;9(4):332–342.